INTRODUCTION

The disorders in this chapter involve mostly the connective tissue structures of the skin and may rather loosely be described as “degenerative.” Solar elastosis, for example, occurs as the result of chronic exposure to solar radiation and could therefore be regarded as a response to injury, rather than a degenerative condition. This process results in an increase in the amount of elastic tissue rather than a decrease as occurs in normal aging. Nevertheless, it seems natural to group these disorders together as they tend to present with alterations of the normal connective tissue architecture that result in loss of structure and often in functional or cosmetic abnormalities. In addition, many of them have the same overlapping differential diagnosis. Therapeutic management of these lesions also tends to overlap or to be similar among the various conditions, although in most cases, these therapeutic options have not been subjected to evidence-based scrutiny, and in many conditions therapeutic intervention is unnecessary except perhaps for cosmetic reasons or for patient reassurance.

SOLAR (ACTINIC) ELASTOSIS

Senescent changes in areas of the skin not regularly exposed to sunlight manifest themselves clinically only in thinning of the skin and a decrease in the amount of subcutaneous fat. In contrast, there are often pronounced changes in the appearance of the sun-exposed skin of elderly persons, especially those with fair complexions. These changes, however, are the result of chronic sun exposure (photaging) rather than intrinsic (chronologic) aging.

Clinical Summary. In exposed areas, especially on the face, the skin shows wrinkling, furrowing, and thinning. In addition, there may be an irregular distribution of pigment. Chronic sun exposure may also lead to more fragile skin and increased areas of purpuric ecchymoses.

Histopathology. In skin not regularly exposed to sunlight, there is a progressive loss of elastic tissue in the papillary dermis with age. Elastic fibers in the papillary dermis are composed of elaunin and oxytalan fibers. Elaunin fibers, which consist of microfibrils and a small amount of elastin, run parallel to the dermoepidermal junction. Overlying oxytalan fibers, composed solely of microfibrils, form a thin, superficial network perpendicular to the dermal–epidermal junction which ends at the basement membrane zone (Chapter 3). In middle age, the oxytalan fibers in the papillary dermis are split and fewer than at a young age, and in old age they may be absent (1). Mature elastic fibers in the reticular dermis also undergo changes due to intrinsic aging, becoming fragmented and porous (2).

In the skin of the face exposed to the sun, especially in persons with fair complexions, hyperplasia of the elastic tissue is usually evident on histologic examination, by the age of 30, even though clinically the skin may appear normal. No white person past 40 years of age has normal elastic tissue in the skin of the face (3). The elastic fibers of the reticular dermis are increased in number, and they are thicker, curled, and tangled.

In patients with clinically evident solar elastosis of the exposed skin, staining with hematoxylin–eosin reveals, in the upper dermis, basophilic degeneration of the collagen separated from a somewhat atrophic epidermis by a narrow band of normal collagen. In the areas of basophilic degeneration, the bundles of eosinophilic collagen have been replaced by amorphous basophilic granular material.

A grading scheme for solar elastosis (i.e., chronic solar damage, CSD) has been developed, validated, and illustrated, and the grades are defined as follows: CSD 0, absence of elastotic fibers discernible at 200× magnification; CSD 1 (mild solar elastosis), scattered elastotic fibers lying as individual fibers, not as bunches in the reticular dermis; CSD 2 (moderate), densely scattered elastotic fibers distributed predominantly as bunches; CSD 3 (severe), amorphous deposits of blue-gray material with lost fiber texture. CSD defined in this way correlates negatively with the frequency of BRAF mutations in melanomas and with the role of the MC1R or melanocortin receptor in relation to the risk of developing melanoma (4).

With elastic tissue stains, the areas of basophilic degeneration stain like elastic tissue and therefore are referred to as elastotic material. The elastic material
usually consists of aggregates of thick, interwoven bands in the upper dermis (Fig. 15-1A) (5); but in areas of severe solar degeneration, the elastotic material may have an amorphous rather than a fibrous appearance (Fig. 15-1B) and may extend into the lower portions of the dermis rather than being confined to the upper dermis (6).

On staining with silver nitrate, the distribution of melanin in the basal cell layer may appear irregular, in that areas of hyperpigmentation alternate with areas of hypopigmentation (6).

Pathogenesis. Electron microscopic examination of areas of solar elastosis shows elastotic material as the main component (EM 13). Even though this elastotic material resembles elastic tissue in its chemical composition, it differs significantly in appearance from aged elastic fibers in unexposed, aged skin. Instead of showing amorphous electron-lucent elastin and aggregates of electron-dense microfibrils (Chapter 3), the thick fibers of elastotic material show two structural components: a fine granular matrix of medium electron density and, within this matrix, homogeneous, electron-dense, irregularly shaped inclusions (7). Microfibrils such as those observed in normal or aged elastic fibers are absent. Immunoelectron microscopy shows that the elastotic material has retained its antigenicity for elastin but not for microfibrils (8). The number and size of elastotic fibers are greatly increased over the number and size of elastic fibers found in normal or aged skin. Extensive amorphous material can be seen around the elastotic fibers and also among the collagen fibrils. Collagen fibrils are diminished in number, with those present often showing a diminished electron density, a diminished contrast in cross striation, and a splitting up into filaments at their ends (9).

Elastotic material is not regarded as a degeneration product of preexisting elastic fibers. Most current findings indicate that elastotic material is composed primarily of elastic tissue, much of which may be newly formed as the result of an altered function of fibroblasts. Evidence of transcriptional activation of the elastin gene in biopsied tissue and fibroblast cultures from sun-damaged skin further supports this. Additional accumulation of elastotic material may be secondary to a disruption in the balance between synthesis and degradation of elastin in photo-damaged skin (10).

The elastotic material that histochemically stains like elastic tissue resembles elastic tissue in its chemical composition and its physical and enzymatic reactions. Thus, the amino acid composition of the elastotic tissue resembles that of elastin and differs significantly from that of collagen. In particular, the elastotic material, like elastic tissue, has a much lower content of hydroxyproline than collagen (11). Moreover, the elastotic material in unfixed sections shows the same brilliant autofluorescence as do elastic fibers on examination with the fluorescence microscope (12), and both the elastotic material and elastic tissue are susceptible to elastase digestion (13). The elastotic material contains a large amount of acid mucopolysaccharides, as indicated by staining with alcian blue. A significant portion of these acid mucopolysaccharides may be sulfated because prior incubation with hyaluronidase removes only 50% to 75% of the alcian blue-positive staining. The basophilia of the elastotic material, however, is not affected by incubation with hyaluronidase (14).

The irregular distribution of melanin in the epidermis observed in some patients with solar degeneration, when studied by electron microscopy, is found to be caused largely by an impairment of pigment transfer from melanocytes to keratinocytes. Although some keratinocytes contain many melanosomes, others contain few or no melanosomes. The latter are surrounded by dendrites laden with melanosomes (15).

Differential Diagnosis. For a discussion of differentiation of solar elastosis from pseudoxanthoma elasticum, see Chapter 3.
Principles of Management. Consistent, long-term use of sunscreens and/or photoprotective clothing reduces solar elastosis significantly (16). Dermabrasion and long-term application of retinoic acid can lead to thickening of the epidermis, which may reduce the clinical appearance of photoaging (17,18). On certain sites, select lasers may ameliorate some of the accumulated photodamage as well.

**LOCALIZED EXPRESSIONS OF SOLAR ELASTOSIS**

Several clinically distinct forms of localized solar elastosis have been described. In the nuchal region, the skin, after many years of exposure to the sun, may appear thickened and furrowed. This is referred to as cutis rhomboidalis nuchae. Elastotic nodules of the ears are localized papular and nodular forms of solar elastosis usually occurring on the antihelix (19–22). Severe solar elastosis may also occur as yellowish plaques associated with small cysts and comedones. Favre–Racouchot syndrome (nodular elastosis with cysts and comedones) is an example occurring on facial skin lateral to the eyes (23,24). A similar condition occurring on the arms has been termed actinic comedonal plaques (25–27). Two other types of circumscribed solar elastosis occurring on the upper extremities are solar elastotic bands of the forearm (5,28) and collagenous and elastotic marginal plaques of the hands (29–36).

**Elastic Nodules of the Ears**

**Clinical Summary.** Elastotic nodules are most often bilateral, small, asymptomatic, pale papules and nodules on the anterior crus of the antihelix and, occasionally, helix (21) of the ears (19,20,22).

**Histopathology.** Irregular elastic fibers and clumps of elastic material are seen in the background of marked dermal solar elastosis (Fig. 15-2A). The fibers and clumps can be highlighted with a Verhoeff-van Gieson elastic stain (Fig. 15-2B) (21).

**Differential Diagnosis.** Clinically, they may mimic basal cell carcinomas, amyloidosis, gouty tophi, or chondrodermatitis nodularis helicis (19–22).

**Favre–Racouchot Syndrome (Nodular Elastosis with Cysts and Comedones)**

**Clinical Summary.** Favre–Racouchot syndrome is characterized by yellow plaques with multiple open and cystically dilated comedones. The condition typically affects the skin lateral to the eyes in elderly males (23,24,37). However, a case has also been documented on the shoulder (38). Although the condition is usually bilateral, it may be unilateral (39–41).

**Histopathology.** Dilated pilosebaceous openings and large, round, cystlike spaces are lined by a flattened epithelium and represent greatly distended hair follicles (23,24). Both the dilated pilosebaceous openings and the cystlike spaces are filled with layered horny material. Vellus hair shafts and bacteria have been demonstrated within the spaces as well, suggesting the cystlike spaces may represent closed comedones rather than true infundibular cysts (42). The sebaceous glands are atrophic. Solar elastosis often is pronounced (23), but it may be slight or absent (43). Because the comedones are open, they do not tend to become inflamed (44) (see section on Acne Vulgaris, Chapter 18).

**Pathogenesis.** It is thought to be primarily secondary to prolonged solar exposure with the formation of comedones facilitated by an extracellular matrix of compromised structural integrity (2). Smoking may also be a contributing factor in its development (45).

**Figure 15-2 Elastotic nodule of the ear.** A: A dome-shaped papule with marked solar elastosis in the dermis and clumped and irregular eosinophilic material representing degenerated elastic fibers. B: The course, clumped material is highlighted with an elastic stain.
Actinic Comedonal Plaques

Clinical Summary. In actinic comedonal plaques, solitary nodular plaques with a cribriform appearance and comedone-like structures occur, often unilaterally, on the arms or forearms (25–27). The plaques are composed of confluent erythematous to bluish papules and nodules. The condition has been described in fair-skinned individuals with a history of chronic sun exposure. They can be found in association with Favre–Racouchot syndrome and may, in fact, represent an ectopic expression of this entity (26).

Histopathology. Dilated corneocyte-filled follicular lumina are present within areas of elastic, amorphous material. The overlying epidermis is usually dyskeratotic and atrophic. The histologic findings are quite similar to those seen in Favre–Racouchot syndrome (25,26).

Solar Elastic Bands of the Forearm

Clinical Summary. Solar elastic bands of the forearm consist of soft cordlike plaques across the flexor surface of the forearms (5,28). The bands occur in areas of actinic damage and usually with senile purpura.

Histopathology. Nodular collections of basophilic homogenous amorphous material underlying an atrophic epidermis are conspicuous features. Thickened degenerated elastic fibers within the homogenous material are also observed. Stellate fibroblasts and a perivascular infiltrate of lymphocytes and hemosiderin-laden macrophages are found in close apposition to the elastic fibers. The nodular collections and thickened elastic fibers stain positively with Verhoeff-van Gieson elastic stain (5).

Collagenous and Elastotic Marginal Plaques of the Hands

Collagenous and elastotic marginal plaques of the hands have been described by several names: elasto-collagenous plaques of the hands (29), degenerative collagenous plaques of the hands (30,32), and keratoelastoidosis marginalis (31).

Clinical Summary. This acquired, slowly progressive condition is usually seen in elderly males and consists of groups of linear confluent papules along the medial and lateral aspects of the hands at the juncture of the palmar and dorsal surfaces. The medial aspect of the thumb and radial aspect of the index finger are most commonly affected. The condition closely resembles the genodermatosis, acrokeratoelastoidosis (34). However, there is no familial predisposition or involvement of the plantar surfaces (31,46,47).

Histopathology. The reticular dermis displays an acellular zone of haphazardly arranged collagen with some bundles running perpendicular to the epidermis (32). The bundles of collagen are admixed with fragmented elastic fibers and distinctive angulated amorphous “basophilic elastic masses” in the upper dermis. These masses can be demonstrated to contain degenerating elastic fibers and calcium (33).

Pathogenesis. Actinic damage and chronic repetitive pressure or trauma has been implicated in its pathogenesis (31,46,47).

PERFORATING DISORDERS

The perforating disorders comprise a group of disorders sharing the common characteristic of transepidermal elimination (TEE). This phenomenon is characterized by the elimination or extrusion of altered dermal substances and, in some cases, by such material behaving as foreign material.

Traditionally, four diseases have been included in this group: Kyrle disease (hyperkeratosis follicularis et para-follicularis in cutem penetrans), perforating folliculitis, elastosis perforans serpiginosa (EPS), and reactive perforating collagenosis (RPC). A fifth entity, acquired perforating dermatosis (APD), which is usually associated with renal disease and/or diabetes mellitus, in which clinical and histologic findings may resemble any of these four diseases, has been added to this group (48). Although TEE is a prominent feature in all these conditions, it has also been described as a secondary phenomenon in other entities, including such inflammatory disorders as granuloma annulare, one variant of pseudoxanthoma elasticum, and chondrodermatitis nodularis helicis. Elastic fibers can be transepidermally eliminated overlying sites of healing wounds. Collagen may be eliminated through keratoacanthomas (49). Needless to say, there is a long list of other conditions that can exhibit TEE as an associated reaction pattern.

Kyrle Disease

Kyrle disease is a rare disorder, described by Kyrle in 1916 (50). There is controversy as to whether it represents a distinct entity, an exaggerated form of perforating folliculitis (50,51), or whether it actually comprises a group of disorders with similar epidermal–dermal reaction patterns associated with chronic renal failure, diabetes, prurigo nodularis, and even keratosis pilaris. Therefore, the discussion of Kyrle disease and perforating disorders seen in chronic renal disease and/or diabetes has a very broad overlap in terms of their clinical and pathologic features.

Clinical Features. This eruption presents with a large number of papules, some coalescing into plaques, numbering in the hundreds and often distributed on the extremities. Although some may appear to involve the follicular units, these lesions are more likely to be extrafollicular. The typical patient is young to middle-aged and often has a history of diabetes mellitus. The papules are dome shaped, 2 to 8 mm in diameter, with a central keratotic
plug. Excoriations often are found in the vicinity of these lesions. Linear lesions related to possible koebnerization have been described.

Histopathology. The essential histopathologic findings include (a) a follicular or extrafollicular cornified plug with focal parakeratosis embedded in an epidermal invagination, (b) basophilic degenerated material identified in small collections throughout the plug, with absence of demonstrable collagen and elastin, (c) abnormal vacuolated and/or dyskeratotic keratinization of the epithelial cells extending to the basal cell zone, (d) irregular epithelial hyperplasia, and (e) an inflammatory component that is typically granulomatous with small foci of suppuration (Fig. 15-3). In most instances, it is important to perform elastic tissue stains and even trichrome stains to exclude perforating elastic fibers, as in EPS, or collagen fibers, as in RPC (52).

Pathogenesis. The primary event is claimed to be a disturbance of epidermal keratinization characterized by the formation of dyskeratotic foci and acceleration of the process of keratinization. This leads to the formation of keratotic plugs with areas of parakeratosis (53–55). Because the rapid rate of differentiation and keratinization exceeds the rate of cell proliferation, the parakeratotic column gradually extends deeper into the abnormal epidermis, leading in most cases to perforation of the parakeratotic column into the dermis. Perforation is not the cause of Kyrle disease, as originally thought (50), but rather represents the consequence or final event of the abnormally sped-up keratinization. This rapid production of abnormal keratin forms a plug which acts as a foreign body, penetrating the epidermis and inciting a granulomatous inflammatory reaction. A certain similarity exists between the parakeratotic column in Kyrle disease and that observed in porokeratosis of Mibelli (54). In both conditions, a parakeratotic column forms as the result of rapid and faulty keratinization of dyskeratotic cells, but, whereas in Kyrle disease the dyskeratotic cells are often used up so that disruption of the epithelium occurs, the clone of dyskeratotic cells can maintain itself in porokeratosis of Mibelli by extending peripherally.

Differential Diagnosis. See Table 15-1.

Principles of Management. Patients may experience reduced pruritus and decreased lesion symptomatology with regular renal-replacement therapy when the condition occurs in the setting of end-stage renal disease. Other treatment modalities are generally supported by case reports and anecdotal evidence, but may include topical anti-inflammatory or antipruritic agents, including topical corticosteroids, or, in some cases, topical retinoids. Limited lesions may respond to local destructive methods, including cryotherapy.

Perforating Folliculitis

Perforating folliculitis is a perforating disorder that has many features overlapping with Kyrle disease and also comprises one of the clinical and histologic patterns seen in APD.

Clinical Summary. As described by Mehregan and Coskey (56), this is a relatively uncommon disorder usually observed in the second to fourth decades and is characterized by erythematous follicular papules with central keratotic plugs. The lesions are 2 to 8 mm in diameter and tend to be localized to the extensor surfaces of the extremities and the buttocks. The key to making this diagnosis is the clinical and histologic identification of a follicular unit as the primary site for the inflammatory process.

Histopathology. The main pathologic abnormalities consist of (a) a dilated follicular infundibulum filled with compact ortho- and parakeratotic cornified cells (Fig. 15-4A); (b) degenerated basophilic staining material, composed of granular nuclear debris from nuclear neutrophils, other inflammatory cells, and degenerated collagen bundles (Fig. 15-4B); (c) one or more perforations through the follicular epithelium; and (d) an associated perifollicular inflammatory cell infiltrate composed of lymphocytes, histiocytes, and neutrophils. Additionally, altered collagen and refractile cosinophilically altered elastic fibers are found adjacent to the sites of perforation. When serial sections through the specimen are examined, a remnant of the hair shaft can sometimes be found.

Pathogenesis. Perforating folliculitis is the end result of abnormal follicular keratinization most likely caused by
Table 15-1

Differential Diagnosis of Perforating Disorders

<table>
<thead>
<tr>
<th>Disease</th>
<th>Primary Defect</th>
<th>Distinctive Features</th>
<th>Histogenesis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kyrle disease</td>
<td>Focus of dyskeratotic rapidly proliferating cells in the epidermis</td>
<td>Follicular or extrafollicular cornified plug embedded in an epidermal invagination associated with epithelial hyperplasia and absence of demonstrable collagen and elastin</td>
<td>Disturbance of keratinization, forming a plug that acts as a foreign body penetrating the epidermis and inciting a granulomatous inflammatory reaction</td>
</tr>
<tr>
<td>Perforating folliculitis</td>
<td>Hyperkeratotic plug in follicular unit containing a retained hair</td>
<td>Compact ortho- and para-keratotic plug with degenerated collagen, altered elastin, and mixed inflammatory cell infiltrate, including neutrophils</td>
<td>A primary irritant causing follicular hyperkeratosis, resulting in a mechanical breakdown of the follicle wall by the hair shaft</td>
</tr>
<tr>
<td>Elastosis perforans serpiginosum</td>
<td>Formation of numerous coarse elastic fibers in the superficial dermis</td>
<td>Formation of narrow channel through an acanthotic epidermis with elimination of eosinophilic elastic fibers</td>
<td>Thickened elastic fibers act as mechanical irritants or “foreign bodies”</td>
</tr>
<tr>
<td>RPC</td>
<td>Subepidermal focus of altered collagen caused by trauma</td>
<td>Formation of cup-shaped vertically-oriented channel with TEE of degenerated collagen</td>
<td>Histochemically altered collagen acts as “foreign body”</td>
</tr>
<tr>
<td>Acquired perforating disorder</td>
<td>Chronic rubbing due to pruritus, resulting in hyperkeratosis, keratosis, and perforation, possibly in association with other factors such as accumulation of poorly dialyzable substances</td>
<td>A combination of features similar to any of the above disorders</td>
<td>Exogenously altered skin in pruritic diabetes and patients with chronic renal disease</td>
</tr>
</tbody>
</table>

irritation, either chemical or physical, and even chronic rubbing. A portion of a curled-up hair is often seen close to or within the area of perforation or even in the dermis, surrounded by a foreign-body granuloma (55).

**Differential Diagnosis.** In Kyrle disease, the keratinous plug may be extrafollicular, the perforation usually is present deep in the invagination at the bottom of the keratinous plug, and no eosinophilic degeneration of elastic fibers.

**Figure 15-4 Perforating folliculitis.** A: A widely dilated follicular unit contains a mixture of keratin, basophilic debris, inflammatory cells, and degenerated collagen fibers. B: An area of disrupted follicular epithelium with adjacent associated perifollicular inflammation and alteration of collagen and elastic fibers.
fibers is found. In addition, in Kyrle disease, epithelial hyperplasia is a significant feature. For a discussion of the differential diagnosis of perforating folliculitis from EPS, see the following section on Differential Diagnosis for Elastosis Perforans Serpiginosa. See also Table 15-1.

Principles of Management. Treatment of this condition is similar to that of Kyrle disease, discussed above.

**Elastosis Perforans Serpiginosa**

EPS is the most distinctive of the perforating disorders because it demonstrates the best example of TEE. In EPS, increased numbers of thickened elastic fibers are present in the upper dermis and altered elastic fibers are extruded through the epidermis.

Clinical Summary. EPS is a rare disorder that affects young individuals with a peak incidence in the second decade. Men are affected more often than women. It is primarily a papular eruption localized to one anatomic site and most commonly affecting the nape of the neck, the face, or the upper extremities. The papules are typically 2 to 5 mm in diameter. These papules are arranged in arcuate or serpiginous groups and may coalesce; there is often mild perilesional inflammation and erythema.

Of particular importance is the association of EPS with systemic diseases. These associations include Down syndrome, Ehlers–Danlos syndrome, osteogenesis imperfecta, pseudoxanthoma elasticum, and Marfan syndrome. In addition, on rare occasions EPS is observed in association with Rothmund–Thompson syndrome or other connective tissue disorders, and as a secondary complication of penicillamine administration.

Histopathology. The essential findings include a narrow transepidermal channel that may be straight, wavy, or of corkscrew shape, and thick, coarse elastic fibers in the channel admixed with granular basophilic staining debris (Fig. 15-5A, B). A mixed inflammatory cell infiltrate accompanies the fibers in the channel. Also observed are abnormal elastic fibers in the upper dermis in the vicinity of the channel. In this zone the elastic fibers are increased in size and number. As these fibers enter the lower portion of the channel, they maintain their normal staining characteristics, but as they approach the epidermal surface they may not stain as expected with elastic stains (57).

Pathogenesis. The cause of EPS is not known. Because the elastic fibers show no obvious abnormality within the dermis except hyperplasia, it is conceivable that the thickened elastic fibers act as mechanical irritants or “foreign bodies” and provoke an epidermal response in the form of hyperplasia. The epidermis then envelopes the irritating material and eliminates it through transepidermal channels. The degeneration of the elastic fibers within the channels probably is caused by proteolytic enzymes set free by degenerating inflammatory cells (57). The channel is formed as a reactive phenomenon through which the “foreign bodies” are extruded. Because copper metabolism is essential to the formation of elastin (58) and because the administration of penicillamine, a copper chelating agent, has been found to induce EPS (59), it may be suggested that the primary abnormality begins with a defect in the metabolism of this essential element.

Differential Diagnosis (Table 15-1). Both Kyrle disease and perforating folliculitis have in common with EPS a central keratotic plug and a perforation through which degenerated material is eliminated. In addition, perforating folliculitis, like EPS, shows the elimination of degenerated eosinophilic elastic fibers. However, neither of the two diseases shows the great increase in elastic tissue that is observed in EPS in the uppermost dermis and particularly in the dermal papillae on staining with elastic tissue stains.

Principles of Management. Treatment modalities include local destruction with cryotherapy (60), topical medications including retinoids, corticosteroids, or salicylic acid, with rare reports of imiquimod (61) benefiting select patients. Systemic treatment can include retinoids (62). Other locally destructive methods may be considered in select cases.

**Reactive Perforating Collagenosis**

RPC is a rare perforating disorder in which altered collagen is extruded by means of TEE. True, classic RPC is
Acquired Perforating Dermatosis

This term was suggested by Rapini et al. (48) to describe a pathologic process encompassing the various forms of TEE seen in patients with renal disease and/or diabetes mellitus. Differences in clinical and histologic features, such as the presence of koebnerization, or histologic evidence of follicular involvement with or without collagen fibers in the epidermis have variably led to diagnoses of Kyrle disease, “acquired” RPC, or perforating folliculitis. Other terms that have been used include perforating disorder secondary to chronic renal failure and/or diabetes mellitus, perforating folliculitis of hemodialysis, Kyrlelike lesions, and uremic follicular hyperkeratosis (70–72).

Clinical Summary. Lesions are frequently pruritic, and range from hyperkeratotic papules and nodules resembling Kyrle disease to RPC-like umbilicated papules, nodules, and plaques to erythematous, follicular papules and nodules mimicking perforating folliculitis (48,49,72). Annular plaques and erythematous pustules have also been described, with histologic features of RPC and perforating folliculitis, respectively (72). The most common location is the extensor surfaces of the extremities, especially lower, but the trunk and head can also be involved (48,72). APD has also been described in renal disease secondary to chronic nephritis, obstructive uropathy, anuria, and hypertension-related nephrosclerosis (49). Cases of APD have also been reported in patients with lymphoma, AIDS, hypothyroidism, hyperparathyroidism, and associated with pruritis secondary to liver disease, neurodermatitis, “atopic dermatitis” and malignant neoplasia (48,73).

Histopathology. As mentioned, the histologic features of APD are variable, even within different lesions from the same patient. When vertically-oriented, Masson-trichrome positive collagen bundles are present within a perforation, the findings are suggestive of RPC. When perforation is associated with a follicle, the findings resemble perforating...
Folliculitis. However, chronic rubbing can lead to superimposed features of prurigo nodularis, distorting the follicle and making it unrecognizable. TEE in the absence of follicular involvement, without demonstrable collagen or elastin, is reminiscent of Kyrle disease. Perforation associated with EVG-positive elastic fibers within a transdermal canal, as seen in EPS, has also been described (72,74). Patterson et al. (75) reported a patient who had multiple lesions biopsied which variably showed histologic features of RPC, perforating folliculitis, and Kyrle disease. Combined TEE of both elastic and collagen fibers have been observed in four patients with renal disease (48), a finding which has only rarely been described in Kyrle disease or RPC (70).

**Pathogenesis.** RPC, EPS, and perforating folliculitis exist as distinct perforating disorders when not associated with renal disease and diabetes. At least in the setting of APD, however, the presence of various histologic patterns suggests the possibility of a common underlying mechanism (48). A nearly ubiquitous clinical feature of APD is pruritis. Certainly, lesions are often distributed in areas accessible to scratching, and reducing pruritis can lead to clearance (73). Poor blood supply secondary to diabetic microangiopathy, combined with trauma, may lead to dermal necrosis, alteration of connective tissue, and TEE (70,73,76). Others have suggested that a consequence of dialysis is the underlying cause of APD (72). Possible etiologies include disruption of metabolism of vitamins A and/or D (77) or the accumulation of a poorly dialyzable substance in the dermis (78,79). Fibronectin, a glycoprotein of the extracellular matrix, accumulates in the serum of uremic and diabetic patients. It has been suggested that transcriptional induction of fibronectin, possibly by transforming growth factor beta (TGF-\(\beta\)) or platelet-derived growth factor (PDGF), increases epithelial migration and proliferation and leads to TEE (77). APD has been reported in 5% to 11% of patients with chronic renal failure undergoing hemodialysis (49,78), and clearance of APD following renal transplant has been reported. However, it has also occurred following transplant, in patients with normal renal function (72). In addition, some cases have occurred prior to the start of hemodialysis or in patients who never underwent dialysis (49). Patterson suggests that TEE may represent a “final common pathway” of a variety of dermal and epithelial processes acting alone or in concert (70).

**Differential Diagnosis.** See Table 15-1.

**Principles of Management.** A similar topical approach to previously discussed diseases may also be reasonable for limited disease or in some clinical scenarios. In conjunction with treatment of underlying systemic disease, allopurinol, doxycycline, systemic retinoids, and narrow-band ultraviolet B phototherapy have been effective in treatment of APD (80).

**Perforating Calcific Elastosis**

(Periumbilical Perforating Pseudoxanthoma Elasticum)

In perforating calcific elastosis, also referred to as periumbilical perforating pseudoxanthoma elasticum (PPPXE), a gradually enlarging, well-demarcated, hyperpigmented patch or plaque is usually seen in the periumbilical region in middle-aged, obese, multiparous women with hypertension (81).

**Clinical Summary.** Most patients described have been African American (82). The patch or plaque is in some instances atrophic with discrete keratotic papules at the periphery (83); in other instances, it has a verrucous border (84), and in still others it has a fissured, verrucous surface throughout (85). Lesions occurring on the breast have also been described (86–88). Perforating calcific elastosis was initially regarded as EPS coexisting with pseudoxanthoma elasticum (89–93). The disorder was later shown to be distinct from EPS (84).

Four patients with perforating calcific elastosis associated with renal failure have been described (82,87,94,95). One patient demonstrated regression of the lesions with hemodialysis (82).

**Histopathology.** Numerous altered elastic fibers are observed in the reticular dermis. They are short, thick, and curled, and are encrusted with calcium salts, as shown by a positive von Kossa stain. They are thus indistinguishable from the elastic fibers seen in pseudoxanthoma elasticum (81). As in pseudoxanthoma elasticum, the elastic fibers are visible even in sections stained with hematoxylin–eosin, owing to their basophilia (83). The altered elastic fibers in perforating calcific elastosis are extruded to the surface either through the epidermis in a wide channel (84), or through a tunnel in the hyperplastic epidermis that ends in a keratin-filled crater (83) (Fig. 15-7A, B).

**Pathogenesis.** Electron microscopic examination reveals electron-dense deposits of calcium primarily in the central core of elastic fibers. Calcification of collagen bundles is also seen (86). The etiologic nature of perforating calcific elastosis has been debated. Some hypothesize it is an acquired lesion developing as a consequence of local cutaneous trauma from such factors as obesity, multiple pregnancies, ascites, or multiple abdominal surgeries (81,83,86). They cite the characteristic clinical presentation, lack of systemic manifestations, and absence of familial predisposition in most cases. The occurrence of perforating calcific elastosis in patients with renal failure may suggest that conditions resulting in an abnormal calcium phosphate product may produce abnormal calcification of elastic fibers. The occurrence of pseudoxanthoma elasticum–like eruptions in patients exposed to saltpeter (calcium–ammonium–nitrate salts) (96), and in one patient with chronic idiopathic hyperphosphatasia (97), supports
HYPERKERATOSIS LENTICULARIS PERSTANS (FLEGEL DISEASE)

A rare dermatosis first described in 1958 (103), hyperkeratosis lenticularis perstans is a disorder that starts in late life and persists indefinitely. An autosomal dominant transmission has been noted in several instances (104–106).

**Clinical Summary.** Hyperkeratosis lenticularis perstans consists of asymptomatic, flat, hyperkeratotic papules from 1 to 5 mm in diameter, located predominantly on the dorsa of the feet and on the lower legs. Removal of the adherent, horny scale causes slight bleeding. In addition to the central horny scale, larger papules often have a peripherally attached collarette of fine scaling. In two reported instances, extensive papular lesions were present on the oral mucosa (107). Other reported sites include the thighs, upper extremities, and ears (108,109). Unilateral involvement may represent postzygotic mosaicism (110).

**Histopathology.** In some instances, the histologic picture is nonspecific, showing hyperkeratosis with occasional areas of parakeratosis, irregular acanthosis intermingled with areas of flattening of the stratum malpighii, and vascular dilation with a moderate amount of perivascular lymphocytic inflammation (111). It seems, however, that if the specimen is obtained from a well-developed, markedly hyperkeratotic lesion, a characteristic, although not necessarily diagnostic, histologic picture may be revealed. The lesion shows a greatly thickened, compact, strongly eosinophilic horny layer standing out in sharp contrast to the less heavily stained basket-weave keratin of the uninvolved epidermis (108). The underlying stratum malpighii appears flattened, with thinning or even absence of the granular layer (Fig. 15-8).

Acanthosis is observed at the periphery. In some instances, bordering on the central depression, the epidermis at the periphery forms a papillomatous elevation resembling a church spire (103,112,113). Vacuolar alteration and apoptotic cells in the basal layer have been seen in some
cases (108,114,115). The dermal infiltrate is composed largely of lymphoid cells and is located as a narrow band fairly close to the epidermis with a rather sharp demarcation at its lower border. Immunohistochemical studies have shown the infiltrate to be predominantly T cells (116–118).

Pathogenesis. Under electron microscopic examination, the absence of membrane-coating granules was noted in some cases and regarded as the primary lesion of hyperkeratosis lenticularis perstans (107,106).

A defect in the membrane-coating granules seems likely to play a role in Flegel disease, because in other cases in which membrane-coating granules were present, the granules lacked a lamellar internal structure and appeared vesicular (119,120). In one case, both lamellar and vesicular bodies were observed, with lamellar bodies greatly predominating in uninvolved epidermis and vesicular bodies in lesional epidermis (121). However, one study of lesional skin from four patients found no such abnormalities in lamellar bodies (122).

Other ultrastructural findings in Flegel disease that have been described in some cases include Sézary cell–like lymphocytes (123), rodlike intracytoplasmic inclusions (124), and wormlike bodies in histiocytes (121). The significance and role of these findings in the etiology of Flegel disease is unknown.

Principles of Management. Treatment of Flegel disease is often unsuccessful. It is resistant to corticosteroids and topical keratolytic agents. Dermal abrasion and shave excision of lesions have been used. Topical 5-fluorouracil and oral retinoids have provided some benefit, but long-term results are disappointing. Topical retinoids may be of some benefit (110).

Figure 15-8 Hyperkeratosis lenticularis perstans. Dense compact orthokeratosis surmounts an epidermis, with focal flattening of the stratum malpighii. A lymphocytic infiltrate is present in the upper dermis.

**STRIAE DISTENSAE**

Striae distensae, often referred to as stretch marks, are a common cutaneous disorder with no significant medical importance but can be quite disturbing to those who are afflicted (125).

Clinical Summary. Striae distensae occur most commonly on the breasts and abdomen, and on the thighs, buttocks, and in the inguinal region. They are caused by stretching of the skin that occurs with pregnancy, growth spurts, obesity, and weight lifting. Use of systemic or local potent topical steroids can contribute to striae development. They consist of bands of thin, wrinkled skin that at first are red, then purple, and finally white, with a degree of atrophy (125).

Histopathology. The epidermis is thin and flattened. There is a decrease in the thickness of the dermis. The upper portion of the dermis shows straight, thin collagen bundles arranged parallel to the skin surface and transverse to the direction of the striae. The elastic fibers are arranged similarly. Fine elastic fibers predominate in early lesions, whereas in older lesions they are thick (126). Within the striae, nuclei are scarce and sweat glands and hair follicles are absent (127).

Pathogenesis. The histologic findings support the view that striae distensae are scars. They occur in conditions associated with increased production of glucocorticoids by the adrenal glands. Among these conditions are pregnancy, obesity, adolescence, and especially Cushing disease (128). In obesity, the increased adrenocortical activity is a consequence of the obesity, and the production of glucocorticoids returns to normal when the body weight is reduced (129). Similarly, the occurrence of striae in nonobese adolescents, as noted in 35% of the girls and 15% of the boys examined, is associated with an increase in 17-kerosteroid excretion (130). Striae may also form in response to prolonged intake of corticosteroids or following the prolonged local application of corticosteroid creams to the skin. The action of the glucocorticoids consists of an antianabolic effect suppressing both fibroblastic and epidermal activity, as tissue culture studies have shown (131).

Principles of Management. The use of topical tretinoin 0.1% cream can improve the appearance of early striae. Several types of lasers have been used to treat early striae (125,132,133). Improvement in the leukoderma of striae (125,132,133). Improvement in the leukoderma of striae (125) is achieved with a 308-nm excimer laser (133). Treatment is generally of limited benefit.

**LINEAR FOCAL ELASTOSIS (ELASTOTIC STRiae)**

Clinical Summary. Linear focal elastosis (elastotic striae) is an uncommon, but more likely underdiagnosed or underreported, disorder presenting as palpable striaelike yellow linear bands (2). This disorder was initially described in the lumbosacral region of elderly males (134). Lesions on the thigh and legs have also been reported (135,136).
Occasionally, lesions arise in association with striae distensae (137–140). A number of cases have occurred in individuals under 30 years of age (141–143).

**Histopathology.** Abundant fragmented, clumped, and wavy elastic fibers are present between hypertrophic collagen bundles in the mid reticular dermis (12,140). Elongated elastic fibers with split ends resembling a “paintbrush” can be seen (144). A decrease in papillary dermal elastic fibers has been demonstrated in the elastotic striae from one patient with coexistent pseudoxanthoma-like papillary dermal elastolysis (136). Unlike striae distensae, there is no decrease in thickness of the dermis or atrophy of the overlying epidermis.

**Pathogenesis.** Electron microscopic examination reveals fragmented elastic tissue throughout the dermis (134). Widespread elastic fiber microfibrils, some occurring in continuity with intracytoplasmic filaments of fibroblasts, along with sequential maturation of elastic fibers, have been observed suggesting an elastogenic process (145). An increased number of elastic fibers in lesional skin have also been documented (144). It has been postulated that elastotic striae may represent a regenerative process of striae distensae (137,140). This view is supported by the presence of contiguous striae distensae and elastotic striae in several patients. Accumulations of thin elastic fibers have been observed in late stages of striae distensae (126,127,146). Thus, linear focal elastosis may represent an excessive “keloidal” repair process occurring in striae distensae (140).

**Principles of Management.** There are no known efficacious treatments for this condition.

**PSEUDOXANTHOMA ELASTICUM–LIKE PAPILLARY DERMAL ELASTOLYSIS**

**Clinical Summary.** Papillary dermal elastolysis, first described in 1992 (147), is a rare, acquired condition consisting of soft coalescent yellow-white papules mainly on the lateral neck and supraclavicular regions of elderly women (136,147–154, 155). The clinical appearance of the skin lesions closely resembles those of pseudoxanthoma elasticum (PXE). However, no other systemic features of PXE are seen. A similar and possibly related disorder, white fibrous papulosis of the neck, is characterized by paler, more discrete and firm papules (156–162). The term fibroelastolytic papulosis of the neck has been suggested to encompass the spectrum of the two disorders (162).

**Histopathology.** There is a marked decrease to absence of elastic fibers in the papillary dermis (147). Focal elastic changes in the subpapillary and mid-dermis have also been demonstrated (163). No calcification or fragmentation of elastic fibers is seen. A slight decrease in elastic fibers along with the presence of thickened collagen bundles in the papillary dermis are differentiating features in white fibrous papulosis of the neck (158,162).

**Pathogenesis.** Electron microscopic examination confirms the absence of elastic fibers in the papillary dermis along with the presence of immature elastic fibers in the upper reticular dermis. Fibroblasts with prominent rough endoplasmic reticulum, with numerous dilatations and elongated dendritic processes, are present (147,153). In one patient, formation of loose component fibrils of elastic fibers and elastophagocytosis were seen ultrastructurally (151). Papillary dermal elastolysis has been suggested to be a disorder of intrinsic skin aging due to its histologic similarity (147). However, immunohistochemical studies have shown a decrease in both fibrillin-1 and elastin in affected areas, whereas aged normal-appearing skin demonstrates only a decrease in fibrillin-1. A defect in elastogenesis may contribute to the pathogenesis of the disorder (147,154). The female predominance and the report of a familial occurrence may suggest possible genetic factors (153). Immunohistochemical studies, using an antibody to serum amyloid component P, which specifically marks the peripheral mantle of microfibrils of elastic fibers in adults, underscored the absence of elastic tissue in the affected papillary dermis (164).

**Principles of Management.** There are no known efficacious treatments for this condition (155).

**MID-DERMAL ELASTOLYSIS**

Mid-dermal elastolysis is a rare disorder, first described in 1977 (165), occurring predominantly on the arms and trunk of middle-aged women.

**Clinical Summary.** Three patterns are seen (166). The type 1 pattern consists of widespread, large areas of fine wrinkling along skin cleavage lines. The type 2 pattern consists of small, soft, papular lesions with tiny perifollicular protrusions, leaving the hair follicle itself as an indented center (167,168). These two patterns may coexist in the same patient. Type 3 presents with persistent reticulate erythema (166,169). Urticaria (165), erythematous patches (170), or granuloma annulare (171) may precede some of the lesions. These postinflammatory forms of mid-dermal elastolysis have some similarity to a disorder first described in young South African girls termed postinflammatory elastolysis and cutis laxa (172–174). In the latter disorder, widespread wrinkling followed an acute phase of firm, erythematous, infiltrated lesions. An overlap with another disorder described as disseminated nevus anelasticus is also likely (175).

**Histopathology.** In this condition there is a selective absence of elastic tissue strictly limited to the mid-dermis.
of involved areas (165). The perifollicular protrusions around indented hair follicles result from preservation of a thin layer of elastic tissue in the immediate vicinity of the follicles. This causes the hair follicles to appear retracted while the perifollicular skin protrudes (167). A mild peri-vascular inflammatory infiltrate of mononuclear cells with occasional interstitial multinucleated giant cells exhibiting elastophagocytosis may be seen (176–178). Electron microscopic examination also reveals fragments of normal appearing elastic fibers within macrophages (176,179).

Pathogenesis. The disorder likely represents a post-inflammatory process, although in some cases this may be subclinical, or remote. Immunohistochemical evidence of immune activation has been observed in lesional T lymphocytes and endothelial cells (180). The cytokines and elastases produced by inflammatory cells along with elastaphagocytosis by macrophages may contribute to the loss of elastic fibers (176). A possible autoimmune mechanism has been hypothesized based on its associations with rheumatoid arthritis (181), silicone mammoplasty, elevated autoantibodies, false positive Lyme titers (182), lupus erythematosus (183), and Hashimoto thyroiditis (184). Culture studies performed with dermal fibroblasts from lesional skin showed an 80-fold increase in elastin mRNA and a 2-fold increase in elastolysis compared with normal skin (155). An idiosyncratic reaction to ultraviolet light has also been proposed as an etiologic or exacerbating factor (185,186). Recent studies suggest that UV exposure may lead to increased expression, by fibroblast-like cells, of elastin-degrading matrix metalloproteinase MMP-9 (187).

Principles of Management. There are no proven therapeutic regimens. Topical retinoids may improve wrinkling but do not alter the course of the process. A long list of treatment options such as colchicine, topical and systemic steroids, and chloroquine have not produced any benefit (166). Because sun exposure may play a role in the pathogenesis of mid-dermal elastolysis, photoprotection is strongly recommended.

MACULAR ATROPHY (ANETODERMA)

Clinical Summary. Atrophic patches located mainly on the upper trunk characterize macular atrophy, or anetoderma. The skin of the patches is thin and blue-white and bulges slightly. The lesions may give the palpating finger the same sensation as a hernial orifice.

Two types of macular atrophy are generally distinguished: the Jadassohn–Pellizzi type, in which the atrophic lesions initially appear red and, on histologic examination, show an inflammatory infiltrate; and the Schweninger–Buzzi type, which clinically is noninflammatory from the beginning. However, not every case can be clearly assigned to one or the other of these two types; many clinically noninflammatory cases show an inflammatory infiltrate when examined histologically (188). The justification for distinguishing between the inflammatory and noninflammatory types has therefore been questioned (189). In many patients, new lesions continue to appear over a period of several years. Rare congenital and familial cases have been reported (190–194).

The so-called secondary form of macular atrophy occurs in the courses of various diseases, such as syphilis, lupus erythematosus, tuberculosis, sarcoidosis, and leprosy (195–198). The macular atrophy then represents the atrophic stage of the preceding disease (199). Other cases of secondary anetoderma have occurred in association with porphyria (200), urticaria pigmentosa (201,202), pilomatrixomas (203,204), Down syndrome (205,206), acrodermatitis chronica atrophicans (189), Takayasu arteritis (207), Graves disease (208), Addison disease (208), autoimmune hemolysis (208), systemic sclerosis (208), varicella (209), anticoagulopathy, and antiphospholipid antibodies (210–216). HIV (217), Sjogren syndrome (218,219), anti-glutamic acid decarboxylase antibodies (210,211), anti-nuclear factor (166), anti-beta-2-glycoprotein antibodies (212,213), and neoplasms (214,215), and generalized granuloma annulare (226). Macular atrophy has also been observed after iatrogenic procedures such as leech application (227), monitoring electrode placement in premature infants (228,229), hepatitis B vaccination (230), and after prolonged treatment with penicillamine (231).

Histopathology. Early, erythematous lesions usually show a moderately pronounced perivascular infiltrate of mononuclear cells (232). In a few instances, however, the early inflammatory lesions show a perivascular infiltrate in which neutrophils and eosinophils predominate and nuclear dust is present, resulting in a histologic picture of leukocytoclastic vasculitis (233,234). Microthrombosis has also been noted in patients with anetoderma and associated antiphospholipid antibodies (221–224).

The elastic tissue may still appear normal in the early stage of an erythematous lesion (225). Usually, however, it is already decreased or even absent within the lesion (235). In cases in which there is a decrease in the amount of elastic tissue, mononuclear cells may be seen adhering to elastic fibers (232). Elastaphagocytosis within macrophages and giant cells may be seen (236).

Longstanding, noninflammatory lesions generally show a more or less complete loss of elastic tissue, either in the papillary and upper reticular dermis or in the upper reticular dermis only (Fig. 15-9). A perivascular and periadnexal lymphocytic infiltrate of varying intensity is invariably present, so that a distinction of an inflammatory and a noninflammatory type is not justified. In some instances, the involved areas show small, normal elastic fibers, which are probably the result of resynthesis, or abnormal, irregular, granular, twisted, fine fibers (188).
Antibodies may induce lesions remains to be elucidated. Some authors have postulated microthrombosis produced by antiphospholipid antibodies with resultant ischemia of dermal tissues may induce elastic fiber degeneration (212,215). Others have hypothesized that the antibodies may alter the inhibitors of elastolytic enzymes, thus allowing the destruction of elastic fibers (241).

**Principles of Management.** Various modalities such as aspirin, phenytoin, dapsone, vitamin E, colchicine, and hydroxychloroquine have been tried, and may have some short-term benefit, but the long-term outcomes have been disappointing (155).

**PERIFOLLICULAR ELASTOLYSIS**

**Clinical Summary.** Perifollicular elastolysis is a relatively common condition consisting of small, hypopigmented, follicular papules on the face and upper trunk. The papules may protrude or herniate (242–245). There is a strong association with acne vulgaris and some have suggested the term *papular acne scars* for the disorder (245).

**Histopathology.** There is an absence of elastic fibers localized to the regions around pilosebaceous units (242).

**Pathogenesis.** Perifollicular elastolysis most likely represents a form of anetoderma related to acne scarring (245).
Elastase-producing strains of *Staphylococcus epidermidis* have been found in lesional hair follicles and have been proposed as the causative factor (242,244).

*Principles of Management.* There is no known treatment.

**ACRO-OSTEOLYSIS**

**Clinical Summary.** The term *acro-osteolysis* refers to destructive lytic changes on the distal phalanges. Three types are recognized: familial; idiopathic, nonfamilial; and occupational, which is associated with exposure to vinyl chloride gas. In addition, acro-osteolysis may be a feature of genetic syndromes such as Haim–Munk syndrome, pykodysostosis, Hutchinson–Guillford syndrome, and Hajdu–Cheney syndrome (246–249).

The *familial* type affects mainly the phalanges of the feet and is associated with recurrent ulcers on the soles (250).

The *idiopathic* type affects the hands more severely than the feet. Involvement of the distal phalanges of the fingers causes shortening of the fingers. This variant may be associated with Raynaud phenomenon. Only one case has been described, with cutaneous lesions consisting of numerous yellow papules 2 to 4 mm in diameter and showing a linear distribution and coalescence into plaques, mainly on the arms (250).

*Occupational* acro-osteolysis, like the idiopathic type, causes shortening of the fingers due to osteolysis. This variant is often associated with Raynaud phenomenon and progressive thickening of the skin of the hands and forearms simulating scleroderma. There may be erythema of the hands, and the thickening may consist of papules and plaques (251). The skin of the face may show diffuse induration (252). In addition, there may be thrombocytopenia, portal fibrosis, and impaired hepatic and pulmonary function (253). A variant of occupational acro-osteolysis has been described in guitar players and is believed to be secondary to mechanical stress (254, 255).

**Histopathology.** The histologic changes in the papules and plaques of idiopathic and occupational acro-osteolysis consist of thickening of the dermis, with swelling and homogenization of the collagen bundles, indistinguishable from scleroderma. Staining for elastic tissue shows disorganization of the elastic fibers, which appear thin and fragmented (250–252).

**Pathogenesis.** Vinyl chloride disease is an immune-complex disorder associated with hyperimmunoglobulinemia, cryoglobulinemia, and evidence of in vivo activation of complement (253). The immunologic nature of the disease explains why it is developed by fewer than 3% of the workers exposed to vinyl chloride gas (251).

*Principles of Management.* Other than treatment and supportive care of the underlying cause, when one is identified, there are no specific therapeutic options.

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