CHAPTER 13

Cutaneous Myxoma

**CLINICAL FEATURES**

Cutaneous myxoma has been classified as a specific neoplasm ever since it was found in patients with Carney complex. This syndrome is inherited as an autosomal dominant trait combining endocrine hyperactivity (Cushing syndrome, testicular tumors, and acromegaly), spotty skin pigmentation, psammomatous melanotic schwannoma, and multiple myxomas (cutaneous, cardiac, and mammary). Most patients with Carney complex have a mutation in the \textit{PRKAR1A} gene, which encodes the R1 alpha subunit of protein kinase A. Cutaneous myxomas in patients with Carney complex may occur on any part of the body surface but are particularly frequent in the external auditory canal (Fig. 13-1) and on the eyelids. Among the described cases, the cutaneous myxoma in a patient with other elements of Carney complex progressed to scleromyxedema. Multiple cutaneous myxomas with no other elements of Carney complex have also been described. Solitary cutaneous myxomas are not normally associated with Carney complex and present as cutaneous or subcutaneous nodules primarily located in distal skin areas of the limbs or the head (Fig. 13-2). When a prominent vascular component is present, the lesion is called angiomyxoma. Clinically, cutaneous myxomas present as slow-growing nodules that have usually been present for many years before they are surgically removed. Some myxomas can be present at birth; some large lesions have also been described.

**HISTOPATHOLOGIC CHARACTERISTICS**

Histopathologically, cutaneous myxomas present as well-circumscribed multilobular tumors with abundant vascular components, particularly in the periphery of the lesion, with collagenous septa compartmentalizing the lesions. Each individual lobule is made up of abundant myxoid material with low cellularity. Stromal cells display a fusiform or stellate morphology and are embedded in the myxoid ground substance (Fig. 13-3). Cytologic atypia is minimal, and normally no mitotic figures are seen. Occasionally, thin-walled dilated blood vessels with abundant red blood cells within their lumina are seen scattered throughout the myxoid material, creating an angiomatous appearance. Variable degrees of hyperplasia are frequently seen in the epidermis covering the lesion, as well as in the epithelia of the adnexa trapped within the tumor, with formation of infundibular cysts, thin basoloid epithelial strands, or basoloid buds that correspond to primitive follicular germs similar to those observed in the fibroepithelioma of Pinkus. These findings of follicular induction in the epidermis covering a myxoma are quite frequent in myxomas of patients with Carney complex (Fig. 13-4). The presence of an inflammatory infiltrate scattered within the lesion is also frequent; in this infiltrate, the presence of neutrophils is a characteristic finding that allows differentiating the myxoma from other cutaneous myxoid lesions.

Immunohistochemical studies have shown that vimentin and alpha-smooth muscle actin may be expressed in the cellular components of the myxoma, but not CD34, S-100 protein,
FIGURE 13-3. Histopathology of a cutaneous myxoma in a patient with no other elements of Carney complex. A: Panoramic view showing a well-circumscribed, exophytic nodular lesion with no attachment to the epidermis. B: The lesion is made up of abundant myxoid material and low cellularity and is separated from the epidermis by a thin collagenous grenz zone. C: Myxoma cells display a fusiform or stellate morphology and are immersed in a myxoid matrix with little fibrillar collagen. D: Detailed image at high magnification of fusiform or stellate cells within the cutaneous myxoma.

factor XIIIa, Leu-7, Kp1, MAC387, or desmin,12,16 which suggests a myofibroblastic nature of these cells. However, other studies found focal expression of CD34 with associated negativity for alpha-smooth muscle actin, desmin, and S-100 protein in the myxoma spindle cells. Electron microscopy studies have shown that these cells exhibit ultrastructural features of fibroblasts.11

Cutaneous myxomas can arise in the genital region,17 and thus the histopathologic differential diagnosis must be established with aggressive angiomyxoma. The deep-seated aggressive angiomyxoma is a genital lesion with a larger size that extends to subcutaneous structures and displays a vascular component different from that in angiomyxomas. The width and wall thickness of neoformed vessels in aggressive angiomyxoma are variable, from small thin-walled capillary vessels to large thick-walled vessels with perivascular hyalinization.

TREATMENT

Cutaneous myxomas are benign neoplasms, although persistence has been described in up to 38% of the cases after incomplete surgical resection,1,16 probably due to the ill delimitation of the subcutaneous lesions.
REFERENCES


FIGURE 13-4. *Histopathology of a cutaneous myxoma in the external auditory canal in a patient with Carney complex.* A: Panoramic view showing a hemispheric papule. The lesion involves the whole thickness of the dermis. B: Follicular induction is seen in the epidermis covering the lesion. C: Follicular induction consists in the development of structures that reproduce a hair germ and a follicular papilla from a follicular follicle in anagen. D: Detailed image of induced follicular germ and papilla.