CASE STUDY

Superficial Angiomyxoma of the Parotid Region and Review of the Literature

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Abstract Superficial angiomyxoma (SA) is a rare benign cutaneous neoplasm first described by Allen et al. in 1988. To the best of our knowledge, we report the first case of superficial angiomyxoma located in the parotid region. We also stress the importance of distinguishing this entity from other lesions that may be involved in this location such as cutaneous neoplasms, parotid tumours or cysts. We emphasise the need to rule out the Carney complex, which has been associated with these tumours.

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Angiomixoma superficial de la región parotídea y revisión de la literatura

Resumen El angiomixoma superficial (AS) es una neoplasia benigna cutánea infrecuente que fue descrita por primera vez por Allen y colaboradores en 1988. Presentamos el primer caso publicado en la literatura médica de angiomixoma superficial localizado en la región parotídea, destacando la importancia de distinguir esta entidad de otras lesiones que pueden aparecer en esta área, como neoplasias cutáneas, tumores o quistes. Así mismo hacemos hincapié en la necesidad de descartar la asociación con el complejo Carney ante el diagnóstico de este tipo de lesiones.

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Introduction

Superficial angiomyxoma (SA) is a rare cutaneous neoplasm, which was first described in 1988 by Allen et al., who reported 30 cases in 28 patients. SA is included in the group of soft tissue, myxomatous tumours characterised by their unique ability to produce a large amount of myxoid material, along with the proliferation of tumour constituent cells.1 We report the case of a patient who presented SA in the parotid region, highlighting the importance of distinguishing this entity from other lesions which may appear in this region.

Clinical Case

The patient was a 61-year old male who attended consultation due to a painless, right parotid tumour, with progressive
growth for 1 year. The patient had no relevant medical history. Physical examination revealed a mobile and elastic mass, approximately 2 cm in size, located in the left parotid region at the level of the mandibular angle. Facial nerve function was normal.

We performed a contrast-enhanced CT scan (Fig. 1), which revealed a sharply defined, hypodense lesion, measuring 27 mm × 16 mm and located immediately in front and below the superficial lobe of the right parotid gland. Fine needle aspiration revealed a mesenchymal lesion accompanied by scattered adipocytes and fibroblasts within a myxoid stroma.

The patient underwent surgical excision of the lesion via a retromandibular approach. Macroscopically, it measured 30 mm × 2.7 mm × 20 mm, presented a whitish colour, had a myxoid appearance and was circumscribed by a thin capsule. Microscopically, it consisted of a sparse proliferation of spindle, stellate, and oval cells within a fibromyxoid stroma, accompanied by lymphocytes and delicate capillaries which did not form a constant network (Fig. 2). We did not find any signs of malignancy or cytological aggressiveness. Immunohistochemical analysis showed positive staining for CD34 and negative for cytokeratins, S-100, desmin, actin, CD99 and CD112. These findings were consistent with the diagnosis of superficial angiomyxoma.

Given the diagnosis, we performed an echocardiogram which revealed the absence of atrial myxomas. Moreover, physical examination found no other tumours or pigmented lesions which suggested an associated syndrome. A complementary, biochemical analysis showed no signs of endocrine hyperactivity.

After 10 months of follow-up, there were no signs of recurrence.

**Discussion**

The term “superficial angiomyxoma” was first used in 1989 to designate a benign, myxoid-type neoplasm characterised by the presence of scattered cell nests and small vessels. This entity was named “superficial angiomyxoma” in order to distinguish it from “aggressive angiomyxoma”, which primarily affects the female genital region.

Histological examination of SA reveals a noninvasive lesion with well-defined margins. Microscopically, it shares some of the characteristics of other myxomatous lesions (cutaneous mucinosis and aggressive angiomyxoma), such as the presence of spindle, stellate, and oval cells within a myxoid stroma. However, SA typically shows a scattered distribution of blood vessels of small and medium calibre and thin walls. An important diagnostic clue is the presence of inflammatory cells, which are absent in other myxomatous lesions. Occasionally, epithelial inclusions may be present, but this was not so in our case. Immunohistochemical analysis is often nonspecific and may show variable positivity for CD34 and protein S-100, smooth muscle actin and pankeratin.

Clinically, SA often appears as a skin nodule, papule or polypoid lesion, mainly affecting the trunk and lower limbs, followed by the head and neck and, finally, the upper limbs.

We conducted a retrospective review of medical literature in English in Medline and found only 28 cases in the head and neck region. We observed a slightly higher prevalence in men (57%) than in women. The age of patients ranged from 12 to 82 years with a mean age of 36.45 years. These data are consistent with those reported in other locations.

The present case represents the first one with parotid involvement described in the literature. In this location, the differential diagnosis must include parotid gland tumours and cysts of the first branchial arch. Fine needle aspiration and MRI studies may be useful in this regard.

The differential diagnosis must include focal cutaneous mucinosis, trichodiscoma, fibrofolliculoma, perifollicular fibroma, trichofolliculoma, trichogenic myxoma and other cutaneous lesions, such as epidermoid cysts, lipomas, neurofibromas, abscesses, lymphangiomata, fibroma, and dermatofibrosarcoma. It is important to rule out an association with Carney complex in all patients, especially in those
forms located in the external ear. This autosomal dominant syndrome includes the presence of cardiac and cutaneous myxomas, skin hyperpigmentation, and endocrine hyperactivity (Cushing syndrome, sexual precocity, and acromegaly). All these characteristics were absent in our patient.

Although they do not generate distant metastases, a general tendency to recur has been described in up to 30%–40% of cases, indicating the need for postoperative monitoring. However, the recurrence rate was lower (16%) in those cases described in the head and neck region.

Conflict of Interests

The authors have no conflicts of interest to declare.

References