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CASE REPORT

Craniofacial/Pediatric

A Unique Presentation Site of Pleomorphic Adenoma in a Young Woman: A Case Report

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Summary: The expression "pleomorphic adenoma" has been used synonymously with mixed cutaneous tumors and chondroid syringomas. It originates from eccrine or apocrine skin, salivary glands, and lacrimal glands. Histologically, it comprises an epithelial-lined glandular component embedded in the cartilaginous, myxomatous, or fibrous stroma. These lesions are usually misdiagnosed because they are extremely rare. It commonly affects middle-aged men and has a slow-growing nature. The usual manifestation is a firm nodular lesion in the periorbital region, particularly at eyelid margins. A unique presentation of this tumor was described in a young woman. The tumor presented as a small, static, nontender lump located at the junction of the superior margin of the left eyebrow and forehead. After a thorough clinical assessment, she underwent a complete surgical excision of the lesion. The most probable preoperative clinical impression at that time was that of a sebaceous cyst. However, histopathological examination revealed it to be a pleomorphic adenoma, which, to our knowledge, has never been reported in the literature at this specific anatomical site. Two years after the procedure, the patient's follow-up was uneventful, and revealed no recurrence of the lesion. Although its incidence is exceptionally low, it should always be considered in the differential diagnosis of cutaneous lesions in the head, neck, and trunk. Complete surgical excision for histopathological assessment is recommended to rule out malignancy and avoid the frequent issue of local recurrence in cases of benign tumors. *(Plast Reconstr Surg Glob Open 2024; 12:e5662; doi: 10.1097/GOX.0000000000005662; Published online 25 March 2024.)*

The tumor historically known as chondroid syringoma, or benign mixed tumor of the eyelid, has been described as originating from the skin appendages.[1] Currently, it is identified using the self-explanatory term pleomorphic adenoma, which better explains its distinctly complex configuration. Many sources, including salivary and lacrimal glands, have been documented in the literature. In addition, sweat gland (eccrine/apocrine) derivation has been reported. Eccrine sweat glands are abundant in eyelid skin, but the apocrine variety (Moll gland) is more common in relation to hair follicles at the eyelid margins.[2] This tumor consists of an epithelial-lined ductal pattern surrounded by mesenchymal stroma of a myxoid, cartilaginous, or fibrous origin. It is a rare tumor and constitutes only 0.01%–0.1% of all cutaneous tumors.[1] Middle-aged men are the most affected; the common presentation is trivial, nontender, and slow-growing swelling.[3] Periorbital and ear skin tumors of the pleomorphic variety have been described frequently in the literature, but their presentation at the superior border of the eyebrow is extremely rare.[2]

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CASE PRESENTATION

Case

A 20-year-old Saudi woman is described here, who had a unique presentation of a left eyebrow mass with insidious onset for 2 years. She presented to the plastic and reconstructive surgery clinic and complained of a mild, dull ache without any history of bleeding or discharge.

Surgical Procedure

After preoperative assessment, she underwent excision of the left eyebrow swelling under local anesthesia. A 1 × 1 cm encapsulated firm mass was removed, secured

Disclosure statements are at the end of this article, following the correspondence information.

in formalin solution, and sent for histopathological examination. The surgical wound was closed with subcuticular sutures using absorbable materials. Her postoperative course was uneventful; the histopathology report was received 1 week after surgery, which revealed well-demarcated heterogeneous elements containing pleomorphic adenoma.

Histological Findings

Microscopically, the specimen consisted of myoepithelial and myxohyaline stroma, and mitotic figures and necrosis were not observed. Epithelial ductal components, scattered spindled myoepithelial cells, and specific stroma were suggestive of a diagnosis. Based on these findings, the mass was diagnosed as a cutaneous mixed tumor, or pleomorphic adenoma.

Microscopically, the lesion showed well-demarcated borders and was composed of heterogeneous elements: epithelial, myoepithelial, and myxohyaline stroma. Focal adipose metaplasia was also observed. Necrosis or significant mitotic figures were not observed. These features confirmed the diagnosis of pleomorphic adenoma (Figs. 1–4).

DISCUSSION

Since the first description of a mixed variety of eccrine glands by Bilroth in 1859, this rare skin tumor has attracted the attention of both surgeons and pathologists.[4] Primary lacrimal and salivary glands, as their main source of origin, have been described repeatedly, but the skin appendageal origin near the eyebrow is significantly nadir.

Based on its histological origin from sweat glands and cartilage elements, Hirsch and Helwig categorized it as a chondroid syringoma in 1961.[5] The most documented presentation is a painless, gradually growing cutaneous lesion of the head and neck region with a size ranging from 0.5 to 3 cm in middle-aged men in their fifties.

Fig. 1. A photograph of the intraoperative lesion after incision depicting the excised tumor.

Fig. 2. A photograph of the left eyebrow of the same patient 2 weeks postoperative.

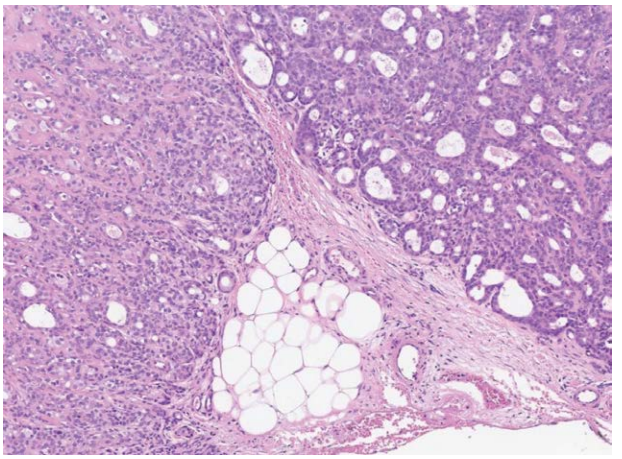

Fig. 3. Epithelial ductal component of pleomorphic adenoma (X10; hematoxylin and eosin).

Periorbital involvement was most frequently reported. Although younger patients reported this lesion, a 12-year-old girl with a lesion close to the lower eyelid punctum and a 13-year-old girl developed a subconjunctival tumor at the lateral fornix of the eye.[6] Just 26 cases had been reported by 2006 in the periorbital region, and a few more cases were published afterwards.[2] Among these patients, only a small number presented with a tumor size greater than 3 cm. However, only a few giant chondroid syringomas of the eyelids have been reported. With increasing tumor size (>3 cm), the malignancy risk also increases. However, most malignant cases arise primarily and are not caused by the malignant transformation of benign tumors.

Chondroid syringoma shares common clinical features with sebaceous/dermoid cysts, neurofibroma,

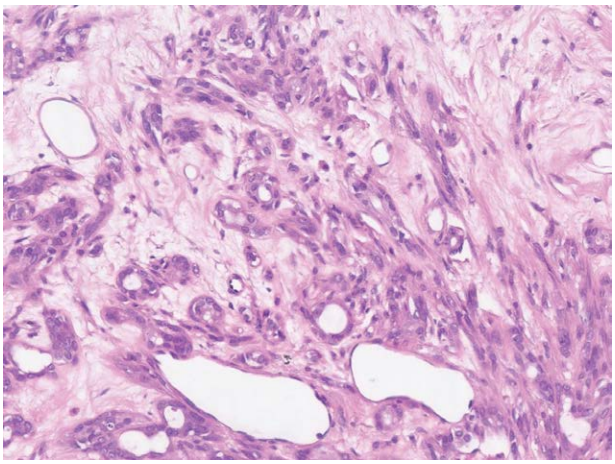

Fig. 4. Pleomorphic adenoma showing epithelial ducts, scattered spindled myoepithelial cells, and myxohyaline stroma. (X20; hematoxylin and eosin).

dermatofibroma, and a nodular variety of basal cell carcinomas, while at the periorbital site, hair follicle tumors, chalazion, and inclusion cysts are also common. An excision biopsy and histopathological analysis indicative of epithelial-lined glandular ducts surrounded by chondromyxoid substances is a confirmatory finding for its diagnosis.[7] Additionally, some histological features of the eccrine and apocrine glands are evident, which indicate their primary tissue of origin.[2] These lesions contained glandular ducts lined with single or double epithelial layers. While the four main varieties of stromal elements, including adipose, fibrous, cartilaginous, and myxoid, are well recognized, osteoid substances may also be found less commonly in stromal components.[5] Features suggestive of malignancy include many atypical or mitotic cells, cell necrosis due to rapid proliferation, pericapsular infiltration, and poor differentiation of the cartilaginous elements.

By 2013, only 30 cases were documented with malignancy, which has female preponderance, male-to-female ratio of 1:1.7, and four times more incidence in the extremities than head and neck.[7] These lesions should be managed by complete surgical excision, and close follow-up is recommended to address the high chances of recurrence and to observe for malignant changes.[2,3] Although Mohs micrographic surgery has been recommended by some authors for small size,[8] Walls et al recommended it for recurrent lesions.[9]

Lesions originating from middle-aged patients have been reported by many authors, including the lid margin, pretarsal area, and subbrow region. It is recognized by epithelial and stromal elements from either the salivary gland or any component of the lacrimal gland. The main presenting features of the subbrow lesion were painless swelling and loss of eyebrow hair.[10] Contrary to these cases, our patient was a young woman with no evidence of alopecia, and the mass was located at the superior border of the eyebrow with its junction to the forehead, originating from an eccrine gland. In addition, four out of the five histological criteria described by Priyank and Saumya were found in our patient.

CONCLUSIONS

In conclusion, pleomorphic adenoma or chondroid syringoma should be ruled out in cases of slow-growing facial skin tumors, particularly in the periorbital and forehead regions. Although more aggressive varieties with a high tendency for malignancy may be found at any site, they are more commonly found in the extremities. Both of these entities, benign and malignant, were differentiated by histopathological analysis after complete surgical excision of the tumor.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

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All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

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