Pilomatricoma

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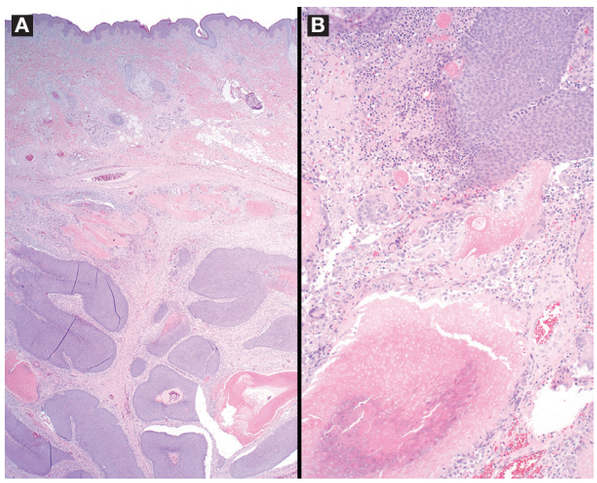

Figure 1. ***A:*** *Low-power photograph shows a basaloid tumor deep in the dermis and subcutaneous tissues.* ***B:*** *A basaloid proliferation juxtaposed toa ghost cell fragment exhibits early signs of calcification. Numerous giant cells are present between the fragments.*

Pilomatricoma, also referred to as *pilomatrixoma* and *calcifying epithelioma of Malherbe,* is a benign dermal-subcutaneous tumor derived from the matrix of the hair follicle. Its development is associated with a known mutation in the *CTNNB1* gene, the gene that encodes for beta-catenin. Pilomatricomas are relatively common tumors. They usually arise during the first 2 decades of life, and they have no predilection for either sex.

The most commonly affected sites are the head and neck and the upper limbs. The lesion presents as a solitary, rubbery to hard mass that often leads to a "tent sign" appearance to the skin. The occurrence of multiple tumors is rare; when they do occur, they are usually syndrome-associated lesions. Simple excision is curative, although some rare cases undergo malignant transformation to pilomatrical carcinoma.

These tumors can be as large as 3 cm in diameter. Their cut surface is gritty to chalky as a result of tumor calcification. Tumors are well-circumscribed, dermal to subcutaneous nodules composed of several components, including basaloid proliferation, shadow cells, dystrophic calcifications, and foreign-body giant-cell reactions (figure 1).

The basaloid cells are tightly cohesive, usually more prominent at the periphery of the tumor, and composed of many layers of small, monotonous cells. The cells are arranged in a syncytium with indistinct cell borders and a very high nucleus-to-cytoplasm ratio (figure 2). They have a high mitotic index, and they merge imperceptibly to abruptly with the keratinizing shadow cells.

The shadow cells have abundant eosinophilic cytoplasm and a negative space where the nucleus was once located (figure 2). As lesions age, the proportion of shadow cells increases. The dystrophic calcifications begin within the shadow cells as fine granules (figure 1). They may ultimately become the dominant finding,

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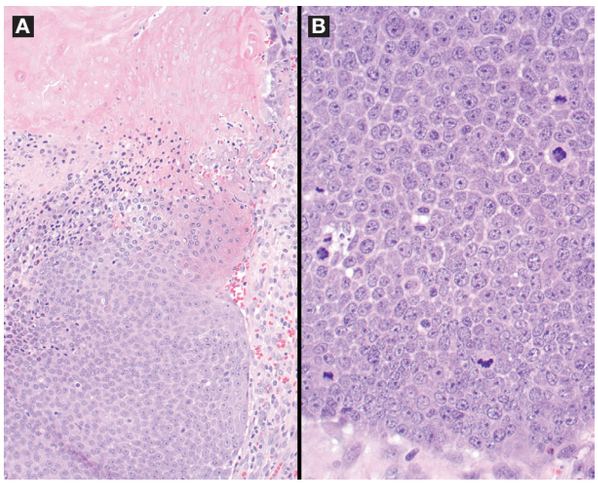

Figure 2. ***A:*** *High-power view shows the basaloid proliferation gradually blending into the keratinizing eosinophilic shadow-cell area. The nuclei are pyknotic just as they transition to the shadow cells (upper).* ***B:*** *A syncytium of basaloid cells exhibits a very high nucleus-to-cytoplasm ratio and numerous mitoses.*

resulting in ossification in up to 20% of cases.

Tumors are frequently cystic and might have ruptured, demonstrating a well-developed, foreign-body, giant-cell reaction. The shadow cells are usually part of the material that is destroyed by the giant-cell reaction.

In rare cases, melanin and extramedullary hematopoiesis may be seen. There is a prominent nuclear reaction with beta-catenin, but this is usually unnecessary for the diagnosis. The diagnosis is usually straightforward, although the basaloid population on fine-needle aspiration samples may be misinterpreted as a carcinoma.

Histologically, the differential diagnosis includes basal cell carcinoma (lacks shadow cells; shows clefting around basaloid groups), neuroendocrine carcinoma such as Merkel cell carcinoma (lacks shadow cells; shows CK20 immunoreactivity), and a proliferating trichilemmal cyst (usually large squamous cells and no shadow cells).

Suggested reading

Duflo S, Nicollas R, Roman S, et al. Pilomatrixoma of the head and neck in children: A study of 38 cases and a review of the literature. Arch Otolaryngol Head Neck Surg 1998;124(11):1239-42.

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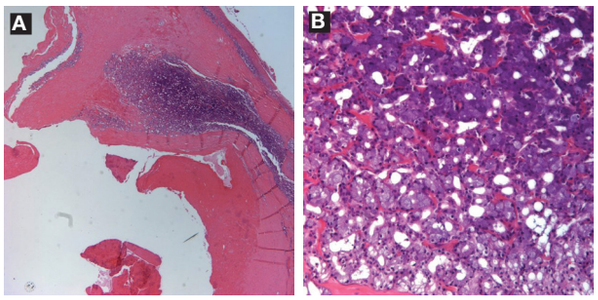

*Figure 2. Microscopically, the surgical specimen exhibits the infiltrative mass of cells arranged in nests and microcysts (**A**) and cells with basophilic cytoplasm, eccentric nuclei, and nuclear pleomorphism (**B**).*

Acinic cell neoplasms are rare in the major salivary glands, and they are even more rare in the minor salivary glands and other sites.[1-3] We describe an unusual presentation of a parapharyngeal acinic cell carcinoma (ACC) that arose from a minor salivary gland.

The patient was a 51-year-old man who presented with a progressively enlarging, left-sided parapharyngeal mass. Computed tomography (CT) demonstrated a circumscribed, low-density, peripherally enhancing mass on the left (figure 1). CT also showed adjacent multilevel, nonspecific, subcentimeter lymph nodes. The mass was completely excised. Grossly, the specimen was a pink-maroon rubbery mass measuring 4.5 × 4.0 × 2.0 cm. Sectioning revealed a cystic cut surface with a tan-pink hemorrhagic and fibrous wall.

Microscopically, the lesion consisted of a fibrous walled pseudocyst with an infiltrative mass of cells arranged in nests and microcysts (figure 2). The cells featured granular basophilic cytoplasm and an eccentric nucleus. Mild nuclear pleomorphism was seen, and the mitotic activity was low. No evidence of metastatic lymphadenopathy was present.

On clinical and imaging follow-up at 6 months, the patient remained stable.

Salivary gland carcinomas represent 0.3 to 0.9% of all cancers in the United States; of these, 6 to 10% are ACCs.[1] ACCs are most common in whites and in women, and the average age at onset is 52 years.[4] In 80% of cases, these slowly growing malignant tumors occur in the parotid gland and are accompanied by by4. Kim SA, Mathog RH. Acinic cell carcinoma of the parotid gland: A 15-year review limited to a single surgeon at a single institution. Ear Nose Throat J 2005;84(9):597-602.

5. Ota Y, Arai T, Yamazaki H, et al. Acinic cell carcinoma of the sublingual gland accompanied by bone formation. Tokai J Exp Clin Med 2001;26(4-6):127-30.