PATHOLOGY CLINIC

Pilomatricoma

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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. They usually arise during the first 2 decades of life, and they have no predilection for either sex.

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. 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. As lesions age, the proportion of shadow cells increases. The dystrophic calcifications begin within the shadow cells as fine granules. They may ultimately become the dominant finding.
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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.


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.

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