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Photo vignette

A diagnostic challenge

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Abstract

A 74 year-old woman presented with a 9 month history of a slowly enlarging exophytic lesion over her left nasal alar. Examination revealed a firm, cystic mass fixed to underlying skin. It was pedunculated with occasional superficial telangiectasia and a violaceous hue. A differential diagnosis of dermoid or sebaceous cyst, dermatofibroma, and neurofibroma was considered. Histopathological examination confirmed the rare diagnosis of chondroid syringoma (CS), which has a reported incidence of 0.01- 0.098% of excised skin tumors. It is unusual for CS to present in an elderly woman, as these benign primary skin tumors normally present in adult males. Dermoscopy does not reveal any specific features and the diagnosis is difficult to make based on clinical examination alone.

Key Words: chondroid syringoma, mixed apocrine tumour, exophytic lesion, apocrine differentiation

Case synopsis

A 74-year-old woman, with an unremarkable medical history, presented with an asymptomatic exophytic lesion over her left nasal alar. The slowly enlarging mass had appeared over a 9 month period. It was 9 x 8 x 6mm, firm and cystic with occasional superficial telangiectasia and a violaceous hue (Figure 1). There was no history of trauma or insect bites and she had no systemic symptoms. The mass was non-tender and fixed to overlying skin. Cervical lymphadenopathy was absent. The patient underwent a shave excision to remove the lesion.



Figure 1. Firm, cystic, and exophytic, 9 x 8 x 6 mm nodule arising over the left nasal alar

Histological examination revealed a well circumscribed, non-encapsulated proliferation of epithelial and mesenchymal components at low power magnification (Figure 2). Within a myxoid, chondroid, and collagenous matrix a proliferation of predominantly apocrine cells was observed arranged in irregular and anastomosing islands with frequent punched out lumina (Figure 3). There were also glandular structures lined by two layers of cells; those on the luminal aspect showed apocrine differentiation with decapitation secretion (Figure 4). There was no cytological atypia or evidence of malignancy.

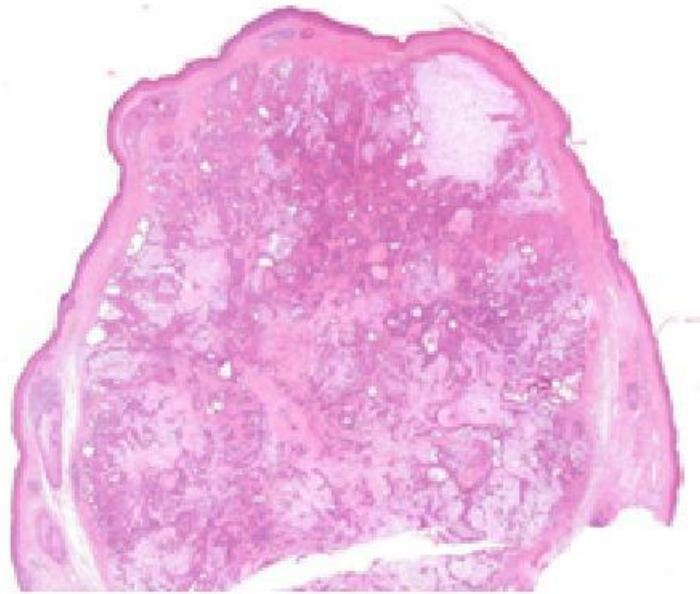


Figure 2. A well circumscribed, non encapsulated proliferation of epithelial and mesenchymal components. (Hematoxylin and eosin, 10x).

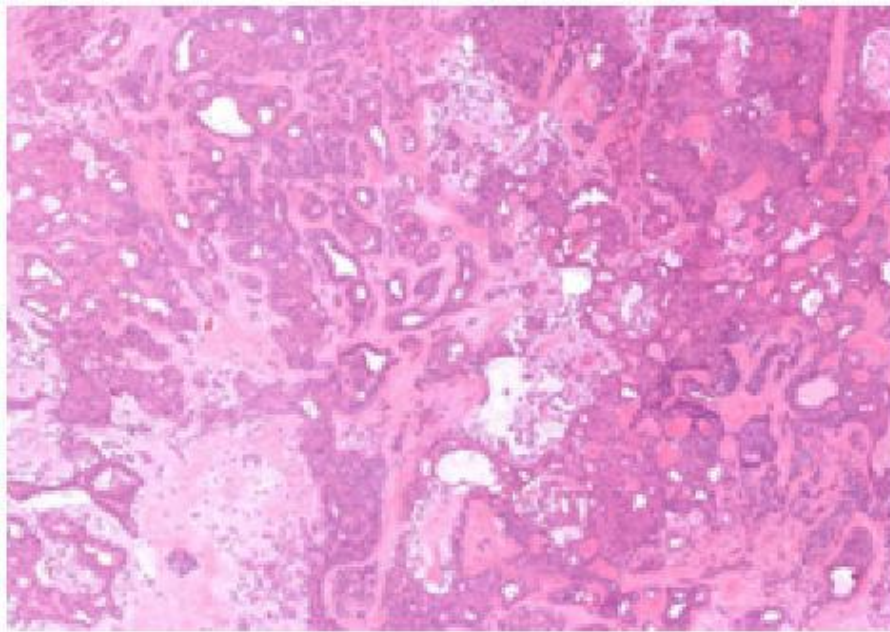


Figure 3. Epithelial, stromal (myxoid and chondroid) and glandular components. (Hematoxylin and eosin, 40x).

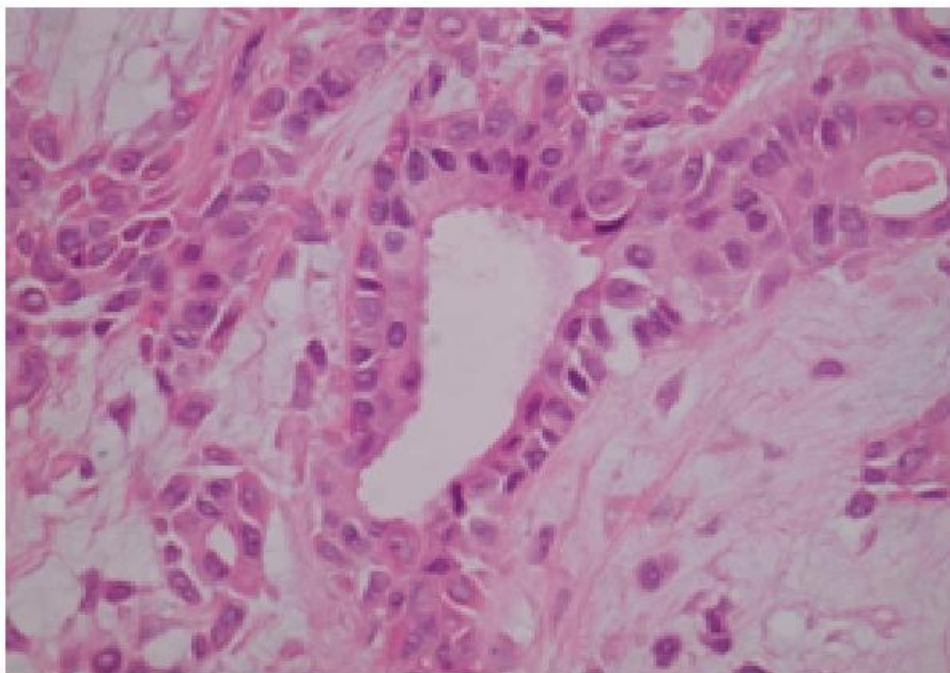


Figure 4. Apocrine cell type with decapitation secretion. (Hematoxylin and eosin, 600x).

Diagnosis: Mixed Apocrine Tumor/ Chondroid Syringoma.

Discussion

Chondroid syringomas (CS) are rare benign primary skin tumors composed of both epithelial and mesenchymal components. CS most commonly affects adult males and often arises on the head and neck. The tumors are slow growing and range in size from between 0.5cm to 3cm in diameter, although rarely they can be larger [1]. They have a reported incidence of 0.01-0.098% of excised skin tumors [2] and a predilection for sun exposed areas. However interestingly, no relationship between CS and sun exposure has been reported [1].

The first case is believed to have been reported in 1859 by Billroth. The term ‘Chondroid Syringoma’ was coined by Hirsch and Helwing to describe the presence of sweat gland elements set in a cartilaginous stroma [2]. They proposed five histological criteria for diagnosis. CS may have all five characteristics or display only some [3]. Headington later recognized that there were two types of benign mixed tumour, apocrine and eccrine [4].

Apocrine tumors are more common and not restricted to any specific site of origin. Dermoscopy does not reveal any specific features and the diagnosis is very difficult to make based on clinical examination alone [1]. The tumor may be confused clinically with various skin lesions such as dermoid or sebaceous cyst, neurofibroma, dermatofibroma, and basal cell carcinoma [5]. Microscopic histological assessment is often needed for diagnosis. Therefore, CS should be considered in the

differential diagnosis of any subcutaneous nodule in the head and neck region of a middle aged individual, especially in men. The definitive treatment of CS is surgical [3].

Very rarely CS can be malignant and case reports with this description are present. In contrast to the benign form, the malignant type commonly affects young females and predominantly occurs on the limbs. Lymphatic and visceral metastases are common, occurring in 42% and 40% of patients, respectively [6]. Malignant lesions are histologically characterized by atypia, mitoses, poorly defined borders, satellitosis, and necrosis [7].

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