

Slowly growing nodule in supralabial region

A 63-year-old man with a history of chronic obstructive pulmonary disease presented with a 16 mm × 15 mm solitary lobulated nodule in the right supralabial region. The tumor that appeared 15 years ago was slowly growing and asymptomatic. On physical examination, the tumor was firm, well-circumscribed, without pain on palpation and the overlying skin showed no inflammatory signs or abnormal pigmentation [Figure 1]. An excisional biopsy was performed. The tumor was well delimited with no adhesions, which allowed complete removal. The histopathologic examination demonstrated a well-circumscribed dermal tumor [Figure 2a] with clusters and solid cords of cells as well as ductal structures

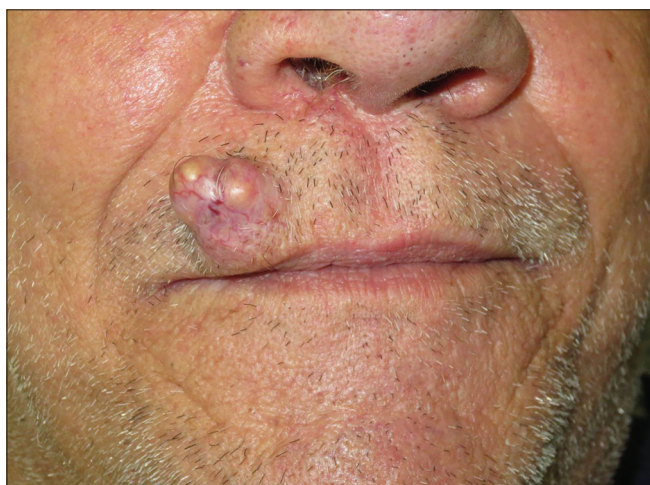


Figure 1: Firm and well-circumscribed solitary lobulated nodule in the right supralabial region

through a myxoid, chondroid and fibrous stroma. Keratinous cysts and calcifications were present [Figure 2b and c]. Tubuloalveolar structures were lined internally by epithelial cells and externally by myoepithelial cells. Decapitation secretion was observed [Figure 2d]. Surgical margins were not affected. No tumor recurrence occurred after 15 months of follow-up.

Question

What is your diagnosis?

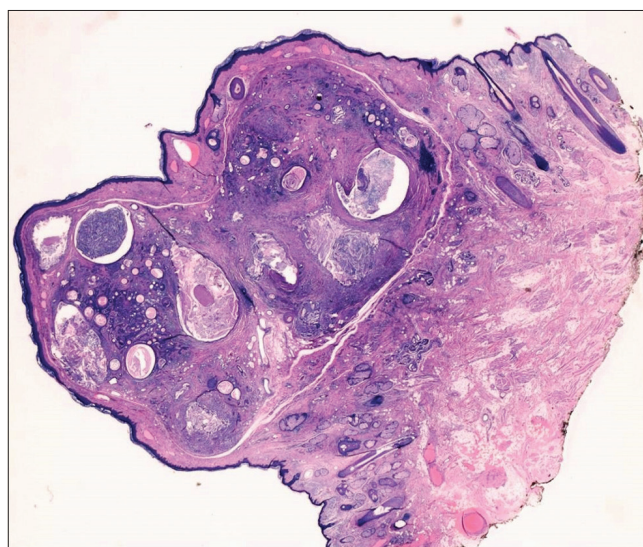


Figure 2a: Well-circumscribed dermal tumor [hematoxylin and eosin (H and E), ×25]

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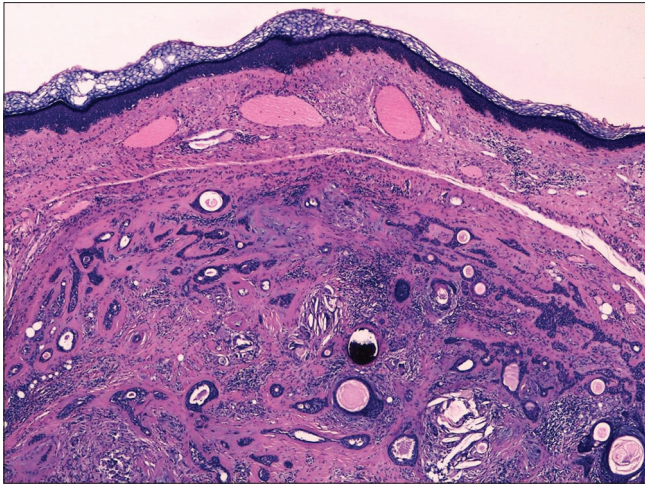


Figure 2b: Cluster and solid cords of cells as well as ductal structures. Keratinous cysts and calcifications can be observed (H and E, $\times 40$)

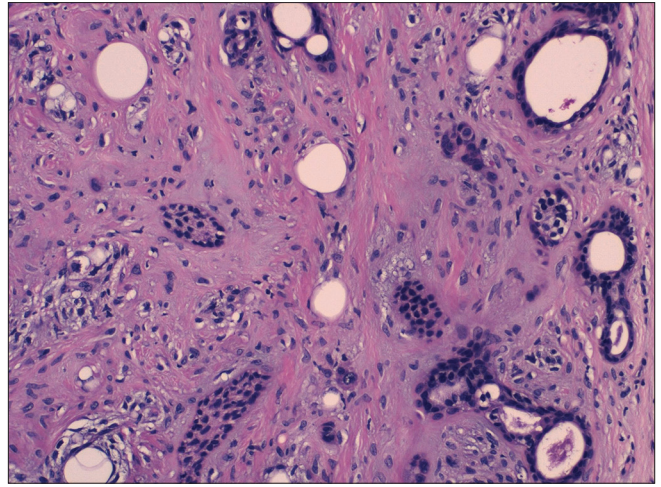


Figure 2c: Clusters, solid cords and ductal structures of epithelial cells included in a fibrous and chondroid matrix (H and E, $\times 200$)

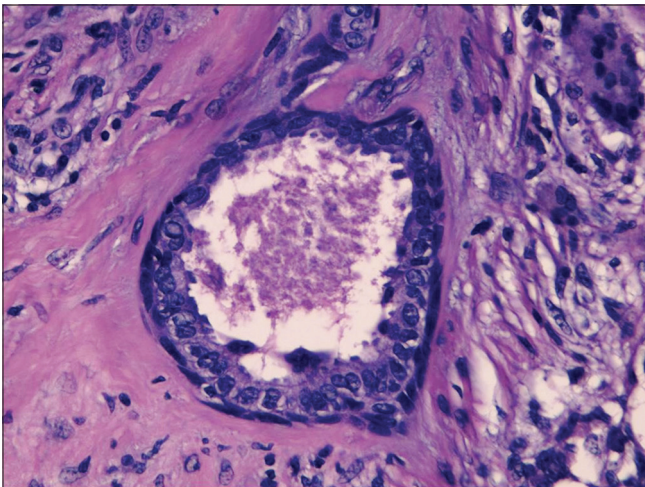


Figure 2d: Tubuloalveolar structures lined internally by epithelial cells and externally by myoepithelial cells. Decapitation secretion can be observed (H and E, $\times 400$)

Answer

Apocrine mixed tumor of the skin.

Discussion

Mixed tumor of the skin (chondroid syringoma) is a rare tumor that was first described in 1859 by Billroth and is considered analogous to pleomorphic adenomas of salivary glands.¹ This tumor typically presents in the middle-aged individuals and males are more commonly affected than females. It usually presents as a solitary, asymptomatic, flesh-colored or reddish, well-circumscribed and slow-growing benign tumor of the head and neck involving especially the nose, upper lip and cheek.^{2,3} Malignant chondroid syringoma is rarer and occurs typically in women and appears on the extremities or trunk.²

Chondroid syringoma contains both epithelial and mesenchymal elements. Most of them are classified as apocrine chondroid syringoma.³ It consists of a multilobulated and well-delimited tumor in the deep dermis or subcutaneous fat with an epithelial component distributed through a myxoid, chondroid and fibrous stroma.^{3,4} The epithelial component is composed of clusters and solid cords of cells as well as tubuloalveolar and ductal structures. The tubuloalveolar structures are lined internally by two or more rows of pale-staining eosinophilic cytoplasm and oval basophilic nuclei cells (epithelial cells) and externally by pale-staining cytoplasm and hyperchromatic-spindled nuclei cells (myoepithelial cells). Tubuloalveolar structures intersect in a retiform fashion. Decapitation secretion is found in most cases. The ducts are lined by cuboidal epithelium. In some of these ducts, keratinous cysts may form. Apocrine chondroid syringoma can present a wide range of metaplastic changes and differentiation in the epithelial, myoepithelial and the stromal component.^{3,4} In the epithelial component, apocrine, follicular and sebaceous differentiations are the most frequent.³ Lipomatous metaplasia, chondroid metaplasia and osseous metaplasia are occasionally present.³ Some of these tumors are classified as atypical mixed tumor due to their borderline features of malignancy with an infiltrative pattern but without dissemination in their follow-up.^{1,3} Immunohistochemically, the epithelial component expresses low molecular weight keratin, epithelial membrane antigen and carcinoembryonic antigen.⁵ The myoepithelial cells express vimentin, S-100 and sometimes muscle-specific actin. Finally, stromal cells are vimentin and S-100 positive.^{1,2,4,5}

The main differential diagnosis should include mixed tumor of salivary gland (for tumors located in the facial region), pilomatricoma, chondroma, osteoma, mesenchymal hamartomas of the skin, dermoid or epidermoid cysts, neurofibromas, dermatofibromas and basal cell carcinomas.^{2,5} Definitive diagnosis can be made by total excision and histopathologic examination.² The combination of epithelial and mesenchymal components is the key finding that allows the correct differentiation from other skin tumors.⁴

The treatment of choice is complete excision, although electrodesiccation, dermabrasion and vaporization with argon or CO₂ laser could be other options.^{1,2,5} Mohs surgery has been used in one patient with recurrent benign chondroid syringoma of the eyebrow with no recurrences.⁵

We highlight the importance of clinical suspicion of this entity when we face a slowly growing nodule. Histological examination will be essential for the correct diagnosis, and complete excision will be necessary for avoiding recurrences.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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