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Review Article

Chondroid Syringoma of the External Ear: A Case Report and Literature Review

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ABSTRACT

We describe an unusual case of Chondroid Syringoma with histological analysis and literature review. Chondroid syringoma, also known as "mixed tumor of the skin" or "pleomorphic adenoma of the skin" is a rare sweat gland tumor with epithelial and mesenchymal features. The incidence is 0.01% of all is 0.01% of all primary cutaneous tumors.

Keywords: Chondroid syringoma; Helix; Sweat gland tumor

INTRODUCTION

The tumors are benign; however, malignant variants were also described [1,2]. Chondroid Syringoma is usually localized in head and neck (79%), followed by extremities (10-15%), moreover scrotal, vulvar, axillar, and cerebral cases are reported but are very unusual and rare anatomic localization [1,3].

Studies in literature found that malignant variant is more common in females and the extremities region than in other sites with a rate of 82%, frequently infiltrating regional lymph node (48%) and with metastasis localized in bones and visceral organs (45%) [2,4]. The most common clinical presentation of a chondroid syringoma is a slow-growing, painless, elevated, firm, and round-shaped mass. Sungur et al. described a case of a benign variant in a patient with an ulcerated, rapid growth mass with necrosis aspects. We describe a case of chondroid syringoma of the helix with histological analysis and literature review.

CASE PRESENTATION

An 85-years-old woman presented to our Ear-Nose-Throat (ENT) Department with a single, firm, and painless nodular lesion of 1.5 cm. The lesion was hidden under whole skin and was localized on the right helix over ten months. The patient underwent a dermatological investigation with the hypothesis of a nodular basal cell carcinoma of the right helix; therefore, a complete exeresis was indicated in order to improve diagnosis definition and outcome. A surgical exeresis of the suspected lesion was executed under local anesthesia, at our head and neck dermatosurgery unit, as following: a wedge resection of the helix, including cartilage and skin was performed with macroscopical free limits, and then the surgical specimen was sent in formalin to the Pathology Department.

The specimen was examined with the classical Hematoxylin-Eosin method. Microscopically the tumor showed findings consistent with chondroid syringoma: dermic multinodular neoplasia with epithelial-myoepithelial pattern within a chondromyxoid stroma. The lesion was wholly excided with a free margin > 1 mm (Figure 1). Based on the histologically benign features, and according to literature, we decided to proceed with only clinical follow-up. At one month follow up visit, we did not find any signs of recurrence.

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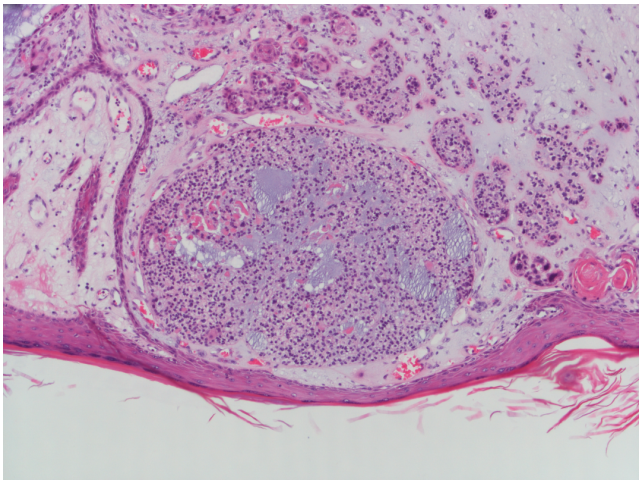

Figure 1: Histologic slice of the tumor in witch can be observed mixed epithelial and mesenchimal elements within a chondromixoid stroma.

DISCUSSION

We propose this case because, to the best of our knowledge, it is unique: helix localization is extremely rare, as there is some literature discussing chondroid syringoma of the external ear canal, or described in ear lobule [5-7], however none of that mentioned helix localization.

In 1961 Hirsc et al. coined the term of chondroid syringoma referring to a kind of a skin tumor type presenting with a mixed epithelial and mesenchymal differentiation. In addition, they established histopathological criteria for its diagnosis (Table 1). The literature review demonstrated histological characteristics that suggest a possible malignant variant: cytologic atypia, infiltrative margins, presence of necrosis, satellite tumor nodules, the involvement of deep structures, a high number of mitosis, excessive mucoid matrix and a poorly differentiated chondroid matrix [1,8-11].

Histopathological Diagnostic Criteria
1. Nests of cuboidal or polygonal cells
2. Tubuloalveolar structure with glandular elements
3. Ductal structure
4. Occasional keratine cysts
5. Chondroid matrix (basophil) or hyaline (eosinophil)

Table 1: Chondroid syringoma, Histopathological Diagnostic Criteria.

We describe, in our case, a specimen of a typical benign chondroid syringoma without histological finding suggestive for malignancy. The clinical differential diagnosis of such a tumor is made with basal cell carcinoma, dermous or sebaceous cyst, neurofibroma, dermatofibroma, histiocytoma, pilomatricoma, seborrheic keratosis. In most cases, the final diagnosis is performed after a histopathological examination. According to the literature, we performed a complete surgical excision with free margins, which is the gold standard procedure for diagnosis and treatment for benign chondroid syringomas.

CONCLUSION

The purpose of our case is to increase the awareness of chondroid syringoma during the differential diagnosis of suspected skin masses.

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