

Primary Pleomorphic Adenoma (Chondroid Siringoma) of the External Auditory Canal. Case Report and Literature Review

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Chondroid siringoma of the external auditory canal is an extremely rare neoplasm, representing the cutaneous counterpart of pleomorphic adenoma of salivary glands. This tumour is thought to derive from the apocrine duct of the follicular-sebaceous-apocrine unit. We report the case of a 68-year-old male in whom the clinical and radiological examinations showed a well-circumscribed tumour limited to the external auditory canal. The diagnosis was confirmed by histologic examination. We also reviewed the literature.

Key words: Chondroid siringoma. Tumour of the ear canal. Pleomorphic adenoma.

Adenoma pleomórfico primario (siringoma condroide) del conducto auditivo externo. A propósito de un caso y revisión de la literatura

El tumor mixto cutáneo de conducto auditivo externo (siringoma condroide) es extremadamente raro. Estas tumores representan los homónimos cutáneos de los adenomas pleomórficos salivales. Se cree que derivan de la llamada unidad foliculosebácea apocrina. Presentamos un caso en un varón de 68 años. El examen clínico y radiológico mostró una lesión bien limitada y circunscrita al conducto auditivo externo. El diagnóstico se confirmó mediante estudio histopatológico. Hacemos revisión de la literatura.

Palabras clave: Siringoma condroide. Tumor de conducto auditivo. Adenoma pleomórfico.

INTRODUCTION

Chondroid siringomas of the external auditory canal are extremely difficult tumours to encounter. Around 25 cases have been reported with this kind of tumour¹ which might be considered as the cutaneous counterpart of pleomorphic adenoma of salivary glands.¹⁻⁶

All authors coincide that tumours in this location originate in the ceruminous glands in the skin of the external auditory canal.

These lesions are encapsulated and histopathology tests reveal they are benign; they are generally diagnosed by chance.

In the case reported here, the patient came to the clinic complaining of left hearing loss due, according to our diagnosis, to the obstruction caused by the tumour.

CASE REPORT

Male, 68 years old, a carpenter by profession, with a personal history of cardiopathy, came to the out-patients' clinic complaining of slowly progressive hearing loss in his left ear.

The otoscopy revealed a papulonodular lesion, covered by normal-looking skin, that occupied the external auditory

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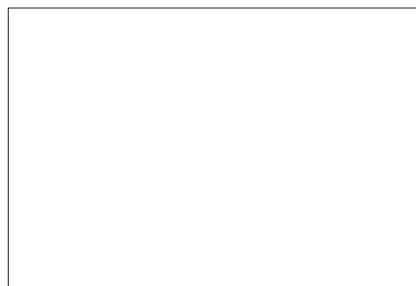
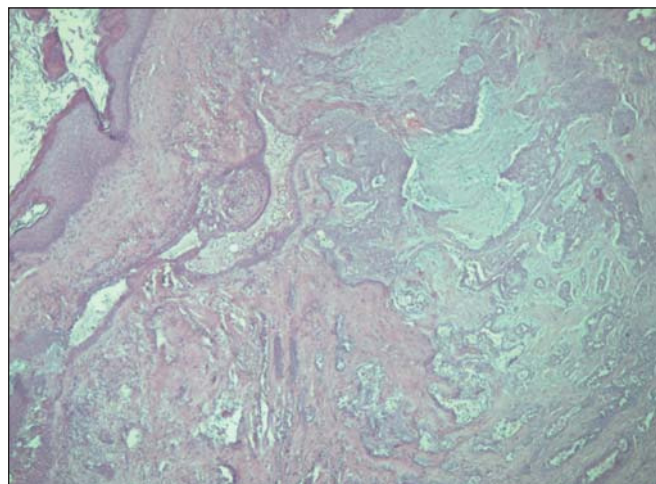


Figure 1.
Computerized tomography in which the external auditory canal is seen to be occupied by a hypodense mass that does not erode its margins and partially occludes its lumen.

Table. Chondroid Syringoma: Histopathological Diagnostic Criteria

1. Niches of polygonal cells
2. Tubuloalveolar structure with glandular elements
3. Ductal structure
4. Occasional keratin cysts
5. Chondroid matrix (basophil) or hyaline (eosinophil)

**Figure 2.** Histologic slice of the tumour in the external auditory canal in which a mixed stroma can be observed to attract basophiles and, on top of this, there are epithelial cells with ductal differentiation (haematoxylin-eosin).

canal. The threshold tonal audiometry confirmed a transmission hearing loss.

Computerized tomography (Figure 1) confirmed the presence of a dense lesion in the soft tissue, limited to the middle third of the external auditory canal, without signs of bone erosion. Surgical exeresis was performed. The histopathology diagnosis was chondroid syringoma.

Following regular check-ups at the outpatients' clinic for 3 years, no relapse has been observed.

DISCUSSION

In 1961, Hirsh et al⁶ coined the term chondroid syringoma to refer to a kind of skin tumour presenting a stroma similar to cartilage and with glandular elements, originating in the apocrine sweat glands (it is also known as a mixed cutaneous tumour). In addition, they established the histopathological criteria for its diagnosis^{6,8} (Table).

Headington⁷ described 2 histological variants, one apocrine and the other eccrine, and therefore argues that the origin of these tumours lies in the follicle-sebaceous-apocrine unit.⁴

This sub-class of tumours includes skin tumours with epithelial strain cells arranged tubularly on a myxoid or chondroid stroma or a mixture of both and there are also signs of glandular or ductal differentiation (Figure 2). In both the histology and the immunohistochemistry, they look similar to mixed tumours (pleomorphic adenoma) of salivary and/or lachrymal glands.¹⁻⁸

They are usually asymptomatic, slow-growing, benign single tumours with an eruptive/papulonodular appearance in subcutaneous or intradermal location; they tend to appear preferably in the cervicofacial region.⁸ In general, magnetic resonance imaging confirms it is a localized lesion that does not erode its settlement margins with hypodensity in T1 and hyperdensity in T2 (similar to mixed parotid tumours) that are grounds for suspicion^{9,10} corroborated by a pathology study. They are exceptionally associated with malignant lesions or degenerate,^{11,12} a behaviour observed with greater frequency when the tumour is located in the lower limbs.⁸

The demonstration of a lesion of these characteristics in the outer ear is exceptional and, although they generally present as a firm or polypoid tumour, some cystic, and/or lipomatous tumours have been described.^{4,8}

Their treatment is surgical by the endaural route and recurrence is exceptional when exeresis is complete.

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