



## Case Report

# Chondroid Syringoma Clinically Mimicking Pilomatricoma with a Rare Location in the Helix: Case Report and Literature Review

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Chondroid syringoma is a rare benign, skin appendageal tumor, also known as mixed tumors of the skin. The clinical presentation of chondroid syringoma is non-specific, and the diagnosis is made histopathologically. A case of a 74-year-old man presented with an asymptomatic 1.3-cm-sized, firm, skin-colored, subcutaneous nodule of 8 years' duration on the superior helical rim of the left ear. Stretching of the skin over the tumor showed the "tent sign" with many facets and angles, a pathognomonic sign for pilomatricoma. The nodule was totally excised under the impression of pilomatricoma. The histopathologic findings showed the characteristic features of chondroid syringoma. Chondroid syringoma arising in the helix is extremely rare; only four cases have been reported in the literature. This case represented the fifth documented case of chondroid syringoma occurring in the helix with clinical presentation mimicking pilomatricoma and the first case involving a male patient. A review of the relevant literature is also presented.

**KEY WORDS:** Chondroid syringoma, helix, pilomatricoma, mixed tumors of skin, pleomorphic adenoma

## INTRODUCTION

Chondroid syringoma is a rare benign, skin appendageal tumor. The reported incidence accounts for 0.010% to 0.098% of all primary skin tumors <sup>[1]</sup>. It contains epithelial and mesenchymal stromal components. Chondroid syringoma occurs most commonly on the nose, cheeks, and lips <sup>[2]</sup>.

A chondroid syringoma arising in the helix is extremely rare, and only four cases have been reported in the literature <sup>[3-6]</sup>. A case of chondroid syringoma clinically mimicking pilomatricoma with a rare location on the superior helical rim of the left ear is reported, along with a review of the literature.

## CASE PRESENTATION

A 74-year-old man presented with an asymptomatic mass of 8 years in duration on the helix of the left ear. The mass had steadily increased in size. There was no preceding history of trauma or cutaneous malignancy in this area. A physical examination revealed a 1.3-cm-sized, firm, non-tender, skin-colored, subcutaneous nodule on the superior helical rim of the left ear, which was adherent to the skin but not fixed to the underlying tissue. Stretching of the skin over the tumor showed the "tent sign" with many facets and angles, a pathognomonic sign for pilomatricoma (Figure 1). There were no overlying skin changes. The nodule was totally excised with direct closure under the impression of pilomatricoma.

Histopathologic examination of the excised lesion showed a well-circumscribed lobulated tumor located in the dermis extending into the subcutis. The tumor comprised numerous various-sized tubular lumina embedded in an abundant fibrous and myxoid stroma (Figure 2). The tubular lumina were lined by two rows of epithelial cells. There were also some solid aggregates of epithelial cells without lumina (Figure 3). The mucoid, faintly basophilic stroma stained positive with alcian blue (Figure 4). There was no calcification or ossification observed. No cellular pleomorphism or dysplasia was noted. The final pathology report showed chondroid syringoma with clear surgical margins. The patient was followed up for 3 years after resection with no sign of recurrence.

## DISCUSSION

The term chondroid syringoma was first introduced by Hirsch and Helwig <sup>[7]</sup> in 1961 and may be preferable to the former terminology, mixed tumor of the skin, because this type of tumor refers to a benign epithelial neoplasm with merely secondary changes in the stroma, while the former designation solely represented its morphological similarity to pleomorphic adenoma. A chondroid

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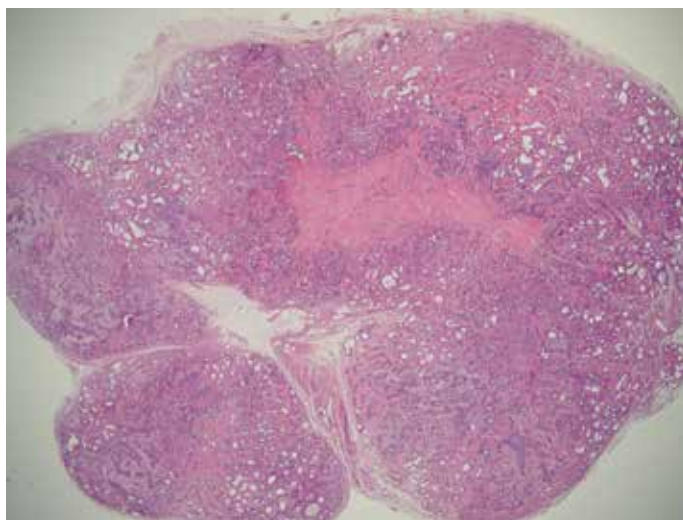
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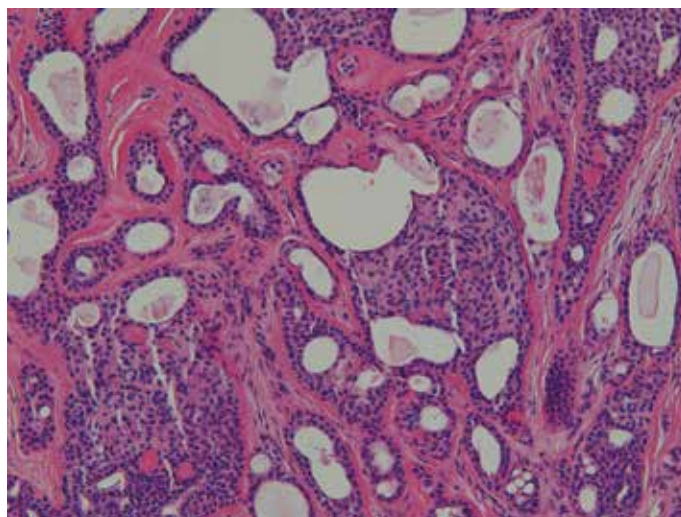


**Figure 1.** A 1.3-cm-sized, firm, skin-colored, subcutaneous nodule on the superior helical rim of the left ear, which was adherent to the skin but not fixed to the underlying tissue. On stretching the overlying skin, the mass showed several facets and irregular angles

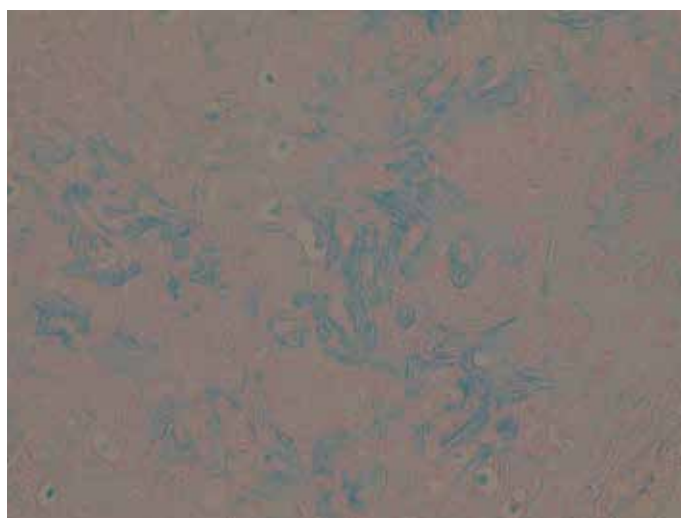


**Figure 2.** A well-circumscribed lobulated tumor comprising numerous various-sized tubular lumina embedded in an abundant fibrous and myxoid stroma (hematoxylin and eosin stain; original magnification  $\times 20$ )

syringoma is usually presented as a slowly growing, asymptomatic, firm intradermal or subcutaneous nodule of 0.5 to 3 cm in diameter [8]. The tumor is usually attached to overlying skin but is freely mov-



**Figure 3.** The tubular lumina were lined by two rows of epithelial cells. There were also some solid aggregates of epithelial cells without lumina (hematoxylin-eosin stain; original magnification  $\times 200$ )



**Figure 4.** The mucoid, faintly basophilic stroma stained positive with alcian blue (alcian blue stain, pH 2.5; original magnification  $\times 100$ )

able over the underlying tissue. The overlying skin is usually normal in texture and color. The chondroid syringoma shows a predilection for middle-aged or elder men. The male-to-female ratio of the prevalence of this condition is approximately 2:1 [8]. The most common locations are the nose, cheek, scalp, upper lip, forehead, and chin. In some instances, the tumor can develop on the trunk and extremities. The external ear is a rare location for a chondroid syringoma, and the external auditory canal was the most frequent site previously reported [9-11].

A review of the literature disclosed only 4 cases of chondroid syringoma arising in the helix that were previously reported (Table 1), and all of them were female patients. Of the four cases, two were reported as a mixed tumor of skin [3, 5] while the other two were pleomorphic adenomas [4, 6]. In 1945, Morehead reported the first case of a 53-year-old female who had a 1-cm-sized tumor with a duration longer than 10 years located on the superior surface of the auricle [3]. He suggested that mixed tumors of the skin, the salivary glands, and the breast are similar morphologically, which suggests a tumor containing tis-

**Table 1.** Cases of Chondroid Syringoma in the Helix Reported in the Literature

Author, Year	Gender	Age	Size	Duration	Designation	Special findings
Morehead [3]	F	53	1 cm	> 10 y	Mixed tumor of skin	
Ito A et al. [4]	F	50	2.5 × 2.0 × 1.7 cm	5 y	Pleomorphic adenoma	
Yim et al. [5]	F	40	0.5 cm	3 y	Pleomorphic adenoma	Gross cystic disease fluid protein stain positively
Ine K et al. [6]	F	63	0.75 cm	6 y	Mixed tumor of skin	
Wen present case	M	74	1.3 cm	8 y	Chondroid syringoma	Clinical mimicking pilomatricoma

sues representing all forms of ectodermal differentiation. The second case, described by Ito et al. [4] in 1982, was a 50-year-old female with a tumor of 5-year duration arising in the lower helix. The tumor measured 2.5×2.0×1.7 cm, which was the largest among the reported cases. The authors postulated that the tumor was derived from the sweat glands or sebaceous glands of the auricle and could be classified as a chondroid syringoma. [4] A case was reported by Yim et al. [5] involving a 40-year-old woman with a 3-year history of a 0.5-cm-sized mass on the superior helical rim. The neoplasm was proposed to originate from the apocrine glands due to a positive stain for gross cystic disease fluid protein. Ine et al. [6] described a 63-year-old woman with a nodule of 6 years' duration on the crus of the helix measuring 7.5 mm in diameter. The authors suggested that the tumor was possibly a unique ceruminous adenoma of the external auditory canal sharing features with a mixed tumor of the skin, because the crus of the auricular helix is an extension of the external meatus. In the present case, the tumor developed in a male patient.

The clinical presentation of chondroid syringoma is non-specific, and the diagnosis is typically made histopathologically. Two histopathologic subtypes have been described [12]. One is composed of variably sized and shaped tubular lumina with cystic dilatation and branching, and the other is a small, tubular lumina. The former subtype is much more common than the latter. The histopathology of this case presented with tubular branching lumina was subtype 1. The four cases in the helix reported previously were also histopathologically classified as subtype 1. The tubular lumina are lined by two layers of epithelial cells. The faintly basophilic, mucoid stroma contains mainly sulfated acid mucopolysaccharides or chondroitin sulfate, which stains positively with alcian blue and mucicarmine. The fibroblasts and epithelial cells that are scattered in the mucoid stroma resemble the cells of cartilage as a result of shrinkage of mucoid substance that leaves a hole surrounding the cells. The chondroid syringoma may originate either from eccrine or apocrine sweat glands. The double-layered tubular structures show eccrine features; the ductal cells, sometimes with decapitation secretion, reveal apocrine differentiation.

Malignant chondroid syringoma may occur de novo or rarely develop in a chondroid syringoma. Shvili and Rothem reported a case with chondroid syringoma of many years' duration that suddenly underwent malignant transformation with widespread metastases [13].

The preferred treatment for chondroid syringomas is surgical excision, which must include a margin of normal tissue [12]. Although recurrence is uncommon, it may occur due to incomplete excision or the existence of lobulated tumor islands.

In conclusion, this is the fifth documented case of a chondroid syringoma occurring in the helix with clinical presentation mimicking pilomatricoma, and the first case involving a male patient. Complete excision with an adequate margin of clearance is highly recommended for prevention of recurrence or malignant transformation.

**Informed Consent:** Informed consent was not obtained as this was a case report without recognizable photographs of patient.

**Peer-review:** Externally peer-reviewed.

**Conflict of Interest:** No conflict of interest was declared by the authors.

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