CASE REPORT

Benign cylindroma: a rare differential diagnosis of external ear canal tumour

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SUMMARY

Cylindromas are rare adnexal skin tumours that usually occur in the head and neck area. This article reports a case of benign cylindroma arising in the external auditory canal. A 75-year-old man presented with a 3-month history of right aural fullness; no other symptoms were reported. Ear microscopy examination revealed a skin mass on the floor of the external ear canal. A CT scan of the skull showed a well-circumscribed soft tissue mass, with no signs of underlying cortical bone erosion. Tumour resection through an endaural approach was performed. Histological examination revealed a benign cylindroma with margins free of tumour, so that no further treatment was necessary. 8 months after surgery the patient is asymptomatic with normal findings in the ear microscopy examination. Benign cylindroma has a high recurrence rate and can undergo malignant transformation. Therefore, complete surgical removal and close follow-up are of great importance.

BACKGROUND

The skin of the cartilaginous part of the external auditory canal (EAC) is characterised by the presence of hair follicles with associated sebaceous and ceruminous glands. Neoplasms of the EAC arising from these adnexal skin structures are rare. It is reported that tumours arising from glandular elements represent fewer than 20% of all EAC neoplasms.^{1–3}

Adnexal skin tumours represent a wide histological spectrum with a complex classification.⁴ Cylindromas are rare adnexal skin tumours that usually occur in the cutaneous areas of the head and neck, particularly on the scalp or face. Cylindromas are considered to be benign tumours; however, malignant transformation has been described.^{3 5 6}

This article reports a case of benign cylindroma arising in the EAC and discusses the diagnostic and therapeutic strategies for this rare neoplasm.

CASE PRESENTATION

A 75-year-old otherwise healthy man presented at the outpatient department with a 3-month history of right aural fullness. Other accompanying otological symptoms were denied.

Ear microscopy examination of the right EAC revealed a compressible, sessile skin mass on the floor of the canal at the border between the cartilaginous and bone areas. The tumour obstructed about three-quarters of the lumen, had a normal skin colour and was painless to palpation. After

Figure 1 Preoperative CT scan in axial plane showing a well-circumscribed soft tissue tumour in the right external auditory canal (arrow). The tumour appeared to be limited exclusively to the soft tissues without signs of extension into the adjacent structures or bone erosion.

removing the accompanying cerumen plug, a normal tympanic membrane could be partially recognised. The rest of the head and neck examination, including the left ear, was unremarkable.

The pure tone audiogram showed symmetric, normal hearing bilaterally, and the caloric vestibular test was normal for both sides as well.

INVESTIGATIONS

A CT of the skull revealed a well-circumscribed soft tissue mass with a maximal diameter of 10 mm in the right EAC. The tumour appeared to be limited exclusively to the soft tissues of the EAC. On imaging, there was no evidence of extension into the adjacent structures and no signs of underlying cortical bone erosion or remodelling (figure 1).

Figure 2 Endaural approach after Heermann's incision (arrow). The nodular tumour in the inferior part of the canal was exposed (star). The patient was in a reclined position on the operating table, so that the incision on the ear canal roof (Heermann's) appears on the left in the picture.

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Figure 3 Histological picture of the tumour (hematoxylin and eosin) showing the lobulated tumour with oval to polygonal mosaic arrangement and sharp margins with no effect on the basal cell layer of the epidermis.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis should include all benign and malignant tumours of the EAC. In order to obtain the exact diagnosis, surgical excision and subsequent histopathological examination is needed. It is important to consider that squamous cell carcinoma represents the most common diagnosis of EAC tumours.³

TREATMENT

The patient underwent a tumour resection through an endaural approach under general anaesthesia. Using a surgical microscope, the nodular tumour was dissected at its base up to the EAC bone and removed in toto (figure 2). No signs of extension into the adjacent structures, or bone erosion, could be detected and the eardrum was normal. After haemostasis, ear packing was inserted.

A definitive histological examination revealed the presence of benign cylindroma. The tumour was covered by squamous cell epithelium and the resection margins appeared free of tumour. The lobulated tumour tissue showed an oval to polygonal mosaic arrangement of cell proliferation (figure 3). Using immunohistochemical staining methods, as antibodies against ki67, malignant proliferation could be excluded.

OUTCOME AND FOLLOW-UP

The postoperative course was uneventful. Ear packing was removed 3 weeks after surgery and normal wound healing was noted. Follow-up was carried out and 8 months after surgery the patient is asymptomatic with a normal EAC in the ear microscopic examination.

DISCUSSION

Cylindromas are rare adnexal skin tumours that belong to the sweat gland neoplasms.³ They usually affect adult females with a female-to-male ratio of 6–9:1. They mostly occur in the cutaneous areas of the head and neck, particularly on the scalp or face. Other locations such as torso and extremities may also be affected. In cases where multiple lesions cover the scalp, the neoplasms are referred to as ‘turban tumours’. The presence of cylindroma in the EAC can be explained by considering the histological structure of the EAC skin. In contrast with the bony segment of the EAC, the skin of the lateral or cartilaginous part possesses hair follicles and associated sebaceous and ceruminous glands. From an embryological point of view, these ceruminous glands are considered to be modified apocrine sweat glands.^{3 4 7 8} The first cases of cylindroma in the EAC were described in the early 1960s and since then several cases have been reported, particularly due to the possibility of malignant transformation.^{9 10}

The histogenesis of cylindromas is controversial, since an eccrine as well as an apocrine origin is discussed. Most reported cylindromas derive from eccrine sweat glands of the skin, however, development from apocrine glands—for instance ceruminous glands of the EAC—can develop this tumour as well.^{4 11} It is believed that cylindromas may arise from a common progenitor capable of dual (apocrine and eccrine) differentiation, such as a peripheral basal cell of the glandular acinus.^{3 12}

Cylindromas mostly appear sporadically as solitary, smooth, nodular lesions in middle-aged and elderly persons and can be up to several centimetres in size. Multiple cylindromas are also possible. These are inherited and mostly arise in the familial cylindromatosis and associated Brooke-Spiegler syndrome.⁸ Although cylindromas are usually benign tumours, malignant degeneration to malignant cylindroma or adenoid cystic carcinoma is possible. Signs such as pain, sudden growth, bluish to pink discoloration or ulceration and bleeding, may suggest malignant transformation.^{13 14}

Histologically, benign cylindromas are characterised by multiple, well-defined islands and nodules of epithelial cells. These structures are covered by a thick periodic acid-Schiff-positive basement membrane-like, hyalinised material, and are arranged in a ‘jigsaw puzzle’ pattern. In the case of malignant cylindroma, an infiltrative growth pattern with loss of the typical jigsaw pattern and infiltration of a hyaline sheath and peripheral structures is histologically noticeable.^{4 8}

The presented case of cylindroma of the EAC showed no clinical or radiographic signs of malignancy. However, definite diagnosis requires histological examination of the tumour. As aforementioned, squamous cell carcinoma represents the most common diagnosis of EAC tumours.³ In the reported case, the nodular tumour was dissected at its base up to the EAC bone and removed in toto. Histological examination revealed a benign cylindroma with margins free of tumour, so that no further surgical treatment was necessary. In the absence of any signs of malignant transformation, no adjuvant treatment was necessary. The patient is in regular follow-up and to date no suspicion of recurrence has been raised.

The modes of treatment of EAC tumours are well discussed in the literature. It is generally supported that benign lesions require wide local excision, whereas in case of malignant neoplasms, an en bloc resection of the external canal and the surrounding structures with subsequent radiotherapy is needed.^{15 16} In case of benign cylindroma, surgical excision is the treatment of choice. Despite its benign nature, cylindroma has a recurrence rate of over 40%, and it can also undergo malignant transformation. Therefore, the complete surgical removal with free resection margins, as well as a close follow-up, is of great importance. In addition to the classic surgical resection, treatment with CO₂ laser has been reported, especially in cases with multiple cylindromas.^{8 17}

Learning points

- ▶ Cylindromas are rare benign tumours of the skin that mostly occur in the head and neck area.
- ▶ Cylindroma of the external ear canal is a rare differential diagnosis. Squamous cell carcinoma represents the most common diagnosis of external ear canal tumours.
- ▶ In the case of cylindroma, because of the high recurrence rate and the possible malignant transformation, a complete surgical removal is necessary.

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