

Syringocystadenoma Papilliferum of the External Auditory Canal in an Adult Female: A Case Report

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ABSTRACT

We present a 50-year old lady with rare benign external auditory canal neoplasm which is syringocystadenoma papilliferum. All of clinical presentation, radiological findings and histopathological examination reveal syringocystadenoma papilliferum. This study presents a rare localization of such adnexal benign tumor in the head and neck in adulthood.

Key Words: Syringocystadenoma Papilliferum, Ceruminous Gland Tumor, External Auditory Canal Mass.

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INTRODUCTION

Syringocystadenoma papilliferum (SCAP) was first described in the dermatological literature in the beginning of the 20th century as a "naevus syringadenomatosus papilliferus".¹ It is a rare benign hamartomatous adnexal tumour, which originates from the apocrine or the eccrine sweat glands.² Most patients present with a solitary lesion in the head and neck region at birth or in early childhood.^{3,4} Presentation with multiple lesions is rare; those arising outside the head and neck region are even more uncommon.⁵ It is relatively a rare neoplasm, predominantly a childhood tumour. In about 50% of those who are affected, it is present at birth, and in a further 15%-30%, the tumour develops before puberty.⁶ Only two cases of malignant transformation yet reported.^{7,8}

We present here a case of right external auditory canal Syringocystadenoma papilliferum mass in adult woman patient. After we reviewed the literatures, we found it a very rare cause of benign mass in head and neck. Few reported case of Syringocystadenoma papilliferum (SCAP) in scalp and ear auricle. In our case, it was the underlying pathology of an external auditory canal mass.

CASE REPORT

A 50-year-old woman, presented to otology clinic with right external auditory canal foreign body sensation for one year. She also complained of decrease hearing. However, she does not have any other significant symptoms notably no ear discharge. Her past surgical and medical history was unremarkable. On physical Examination, there was a soft mass at the external

auditory canal of her right ear. Measured 1 x 1 cm. with smooth pale surface. Originating from the roof of the canal at the level of the bony-cartilaginous junction and obstructing the whole canal. There for the tympanic membrane was not visualized. The rest of E.N.T. examination was within normal.

Audiological assessment was done, including pure tone audiometry, speech audiometry and impedance. It showed bilateral symmetrical mild high frequency sensorineural hearing loss. Speech reception threshold was 20 dB for both sides. Moreover, speech discrimination score for right ear was 92% and 96% for left side. Tympanogram was type A bilateral and normally present acoustic reflex.

We requested temporal bone high-resolution computer tomography (HRCT) scan and Magnetic Resonance imaging (MRI).

HRCT showed well-defined lesion related to superior wall of external canal with no bony erosions (Fig 1). On MRI this well-defined lesion showed high intensity signal on T2WI and iso-intensity signal on T1WI measuring 8 by 8 mm (Fig 2).

After discussion with the patient, we decided to excise the lesion under local anesthesia. Surgery was performed with no difficulties, a cystic mass completely dissected with maximum preservation of external auditory canal skin, and the specimen sent for histopathological study. The tympanic membrane was normal and intact behind the mass. Dressing of the canal done using a small otowick.

Histopathological study revealed a cystic invagination extending downward from the epidermis. Epidermis is acanthotic and shows

papillomatosis. Villous projections were seen within these invaginations (fig 3 & 4). This picture suggests the diagnosis of syringocystadenoma papilliferum.

Postoperatively the patient showed complete healing of external auditory canal skin with no residual mass. Our patient now been followed for one year; showed no new symptoms.

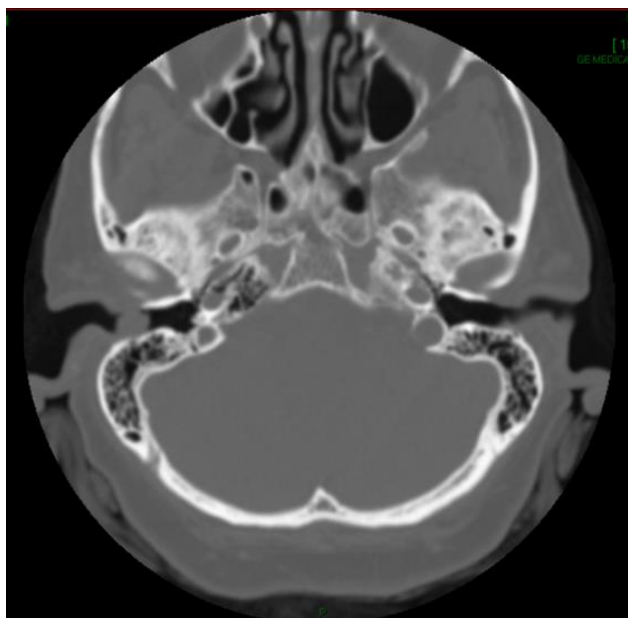


Fig 1: (HRCT scan) shows well defined mass localized at bony-cartilaginous junction of the right external auditory canal without bony erosion. No extension to tympanic membrane and the middle ear cavity.

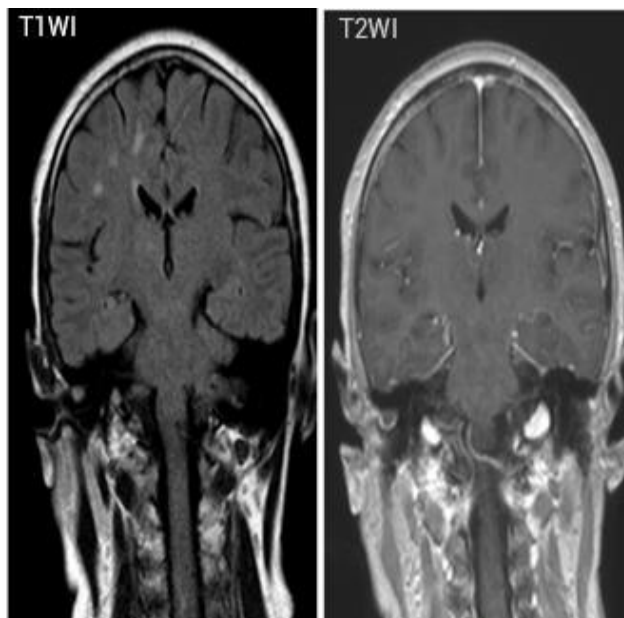


Fig 2: (MRI) showing clear originating from bony-cartilaginous junction of the right external auditory and no extension intracranial.

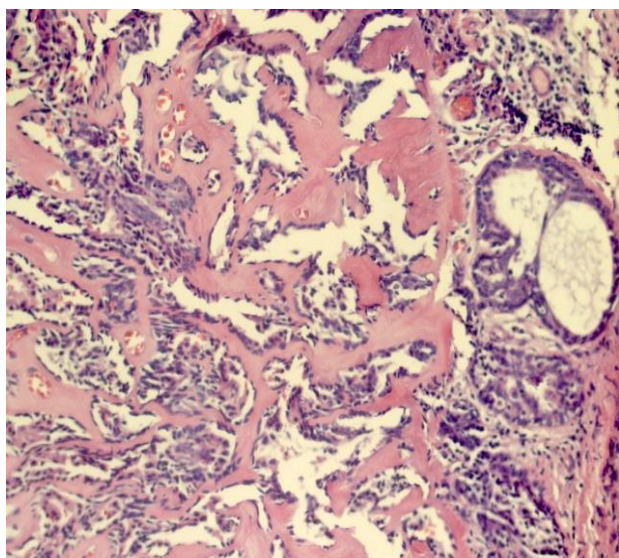


Fig 3: Cystic invagination extends downward from the epidermis.

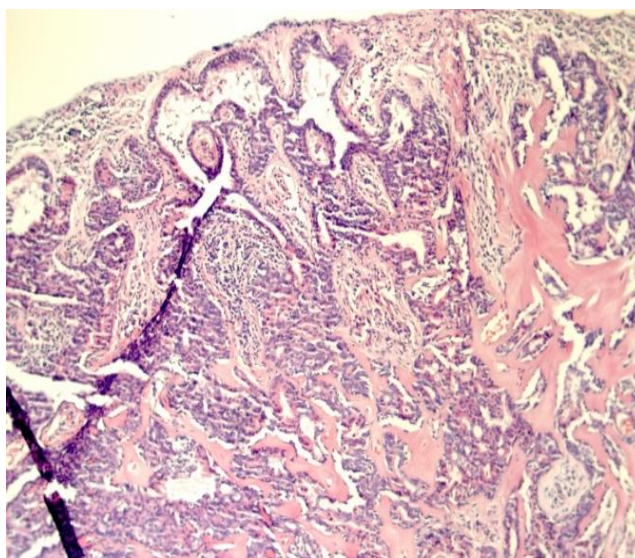


Fig 4: Papillomatous projections with invaginations.

DISCUSSION

Ceruminous glands are normally located in the outer one-third to one-half of the external auditory canal skin overlying the cartilaginous region.⁹ Ceruminous glands tumors have previously been characterized as well-differentiated localized benign neoplasms that are occasionally cystic and demonstrate the papillary proliferation of glands that are histologically similar to normal ceruminous glands.¹⁰ SCAP was previously reported to be often associated with pre-existing nevus sebaceous.¹¹ Three clinical types have been described syringocystadenoma papilliferum:

a. Plaque type: Presenting as an alopecic patch on the scalp and may enlarge during puberty to become nodular, verrucous or

crusted. Plaques commonly tend to be associated with a naevus sebaceous of Jadassohn in one-third of the cases.

b. Linear type: Consists of multiple reddish pink firm papules or umbilicated nodules 1-10 mm in size commonly occurring over face and neck.

c. Solitary nodular type: They are domed pedunculated nodules 5-10mm in size with a predilection for the trunk shoulder and axillae.¹

Syringocystadenoma papilliferum is exceedingly rare neoplasm. Almost half are present at birth or appear during infancy (6). In our case, it is a very rare exception of presentation during adulthood and a similar case was reported which was presented clinically exactly the same as our patient.¹¹

Because of malignancy transformation risk, surgical excision is the standard treatment with the regular follow up to insure the absence of recurrence.

CONCLUSION

Syringocystadenoma papilliferum is exceedingly rare neoplasm. Reported to be more at birth or during infancy. Should be considered if slowly cystic, soft and well defined growing mass at lateral third of ear canal.

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