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Case Report

# Syringocystadenomapapilliferum Arising from the External Auditory Canal: A Rare Tumor in a Rare Site

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Syringocystadenomapapilliferum is an extremely rare benign adnexal tumor of the scalp and face region. This is a report of the case of a female patient with syringocystadenomapapilliferum originating from the bony cartilaginous junction of the external auditory canal. A provisional diagnosis of an inflammatory polyp was made based on the clinical and radiological findings. Diagnosis was established only after the histopathological examination. This article represents a report of a rare skin disease and a rare site of affection. It emphasizes the role of histopathology in the diagnosis of such a condition with debatable clinical and radiological findings.

**KEYWORDS:** Syringocystadenomapapilliferum, adnexal, external auditory canal

## INTRODUCTION

Any mass or polypoid lesion in the external auditory canal is usually challenging as a proper diagnosis cannot be made. This is due to its nonspecific clinical and radiological findings [1]. To accurately diagnose the nature of any aural polyp, imaging techniques and polypectomy should be considered to be mindful that it may uncover a serious disease process or even a surprising one [2].

Syringocystadenomapapilliferum (SCAP) is an extremely rare benign tumor originating from modified apocrine sweat (ceruminous) glands showing extensive papillary growth of epithelial elements deep into the dermis [3]. According to the World Health Organization, ceruminous gland tumors are classified into benign ceruminous adenomas, chondroidsyringomas, SCAPs and malignant adenocarcinomas, adenoid cystic carcinomas, and mucoepidermoid carcinomas [4]. Eleven cases of SCAP of the cartilaginous par0000t of the external auditory canal have been mentioned in the literature till 2006 [5]. To my knowledge, another two cases have been mentioned since that time: one associated with lipomatous apocrine adenomas [6] and the other originating from the bony part of the ear canal [7].

## CASE PRESENTATION

A 36-year-old female presented to our tertiary hospital with right ear pain for a duration of 3 days duration following self-manipulation with a cotton bud. The patient did not have a history of a previous similar condition, ear discharge, or hearing loss before this episode. She had no history of general organ system problems. She also had a history of diminished hearing in the right ear and a sense of right ear fullness for a duration of 2 months. No tragal or mastoid tenderness was found in both ears. A left ear examination showed normal findings. A right ear examination by otoscopy and ear endoscopy showed a well-circumscribed soft mass lesion filling the deep bony part of the ear canal (Figure 1). It was not tender to touch, and the probe easily passed around the swelling from all directions except the posterior part. The right ear drum could not be visualized as it was totally obscured by the swelling.

The Rinne test was positive in both ears, and Weber test was central. Pure tone audiometry showed mild conductive hearing loss of the right ear. Aspiration was attempted, with no fluid coming out from the swelling.

Computed tomography (CT) with contrast for the right petrous bone was performed to identify the mass extension and condition of the middle ear. It revealed a nonhomogenous soft tissue lesion of a density of 30–40 HU and measuring approximately 1.3×0.7 cm, which filled the deep bony part of the right ear canal starting from the bony cartilaginous junction, and the right ear drum could not be separately visualized from the lesion. The middle ear cleft was intact (Figure 2).

Because there was no destruction of the bony meatal wall as seen in the CT scan, a suggestion of a benign pathology was raised and a decision of excision biopsy of the lesion was taken.

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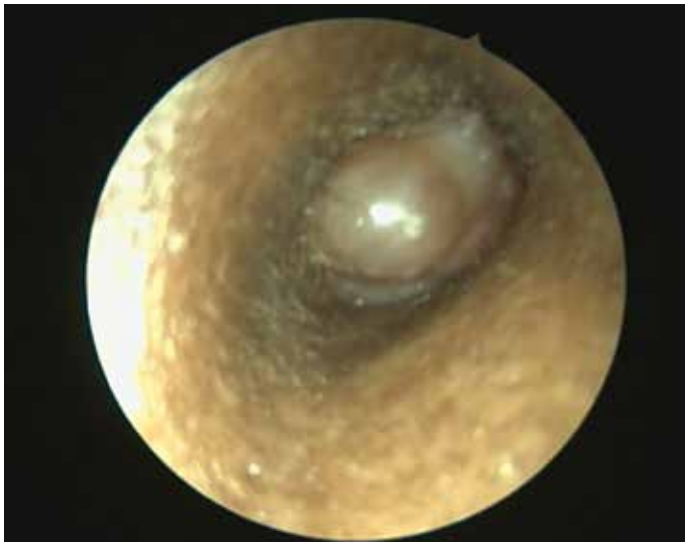

**Figure 1.** The mass lesion in the right external auditory canal as seen on ear endoscopy

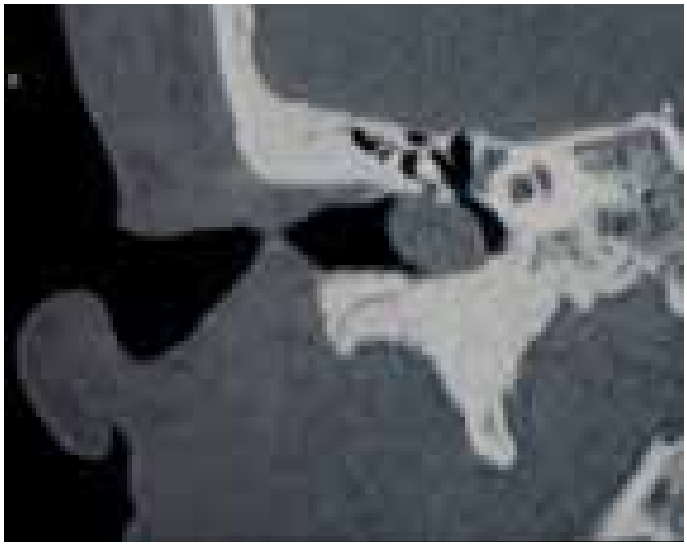

**Figure 2.** The mass in the deep part of the ear canal as seen on the enhanced CT scan pf the right petrous bone

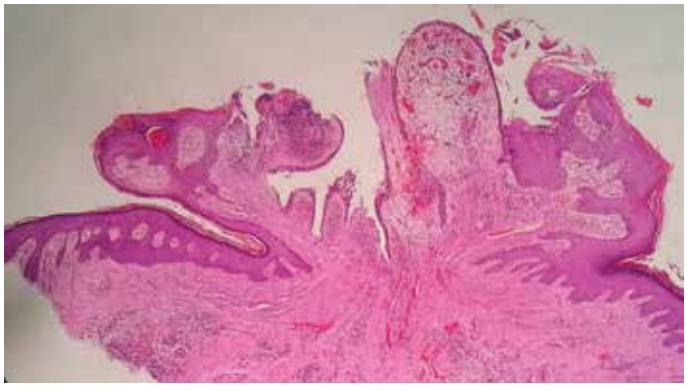

**Figure 3.** Papillomatous epidermis with multiple invaginations

Written informed consent was obtained from the patient who participated in this study. The permeatal approach was performed under general anesthesia. The mass appeared to be originating from the posterior superior part of the ear canal at the bony cartilaginous junction and filling the deep bony part of the canal. Cauterization of the base of the mass with bipolar diathermy was performed, and hemostasis was achieved. No skin grafts were added in this patient as the defect where the pedicle originated was small. The ear drum was intact.

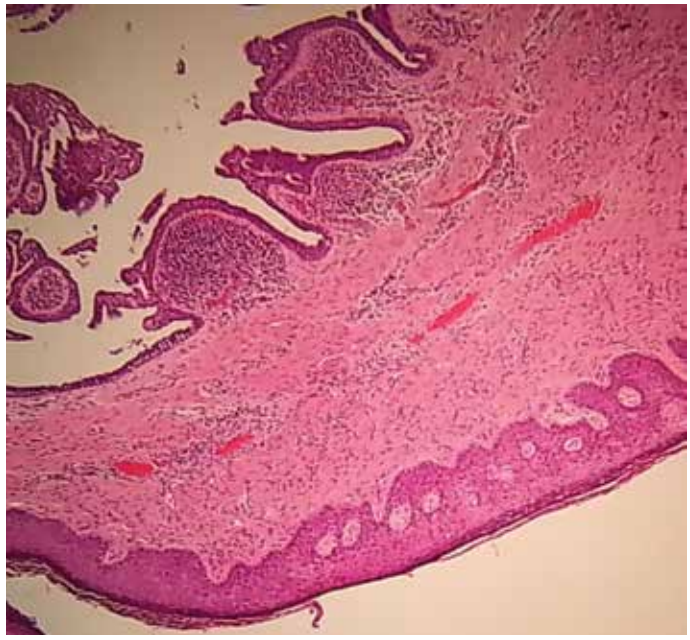

**Figure 4.** Cystic spaces with papillary projections

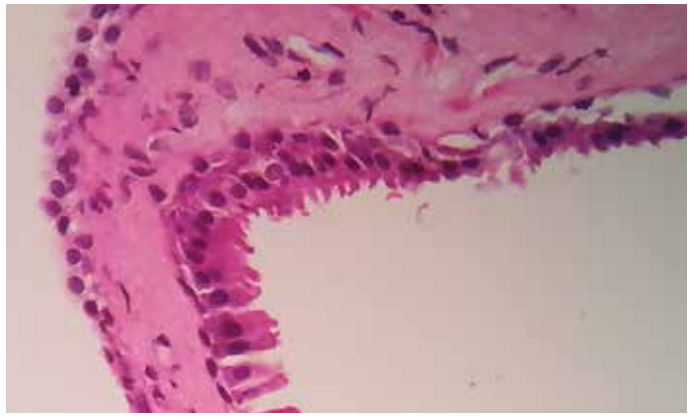

**Figure 5.** Sweat gland epithelium with apocrine differentiation and decapitation secretions

The sample was received in the histopathology department, fixed in formalin, and labeled with the patient's name. It was composed of a nodular grey white tissue covered by skin, and its gross size was 0.8×0.6×0.2 cm. Sections from the tissue (Figure 3) showed papillomatous epidermis with multiple invaginations extending from the surface epithelium. There were cystic spaces with numerous papillary projections having stroma with dense inflammatory infiltrate rich in plasma cells (Figure 4).

The invaginations and cystic spaces were lined by either squamous epithelium near the surface or double-layered sweat gland epithelium with apocrine differentiation and decapitation secretion (Figure 5), which confirmed the diagnosis of SCAP.

Postoperatively, the patient showed complete healing of the skin with no residualor recurrence of the lesion over a follow-up period of 10 months.

## DISCUSSION

Ceruminous gland tumors are rare and represent less than 5% of all tumors of the external auditory canal and auricular tumors [8].

Ceruminous glands are found in the outer cartilaginous part of the external auditory canal, and there is a general acceptance that the bony part of the canal is devoid of them.

Syringocystadenomapapilliferum is one of these rare tumors that has been mentioned in the literature in the early 20th century in dermatological case reports as nevus syringocystadenomatosuspapilliferus [9].

It has been also reported that SCAP is usually associated with a nevus in the head and neck region [10]. However, there was no nevus in the head and neck region in our patient.

In this presented case, SCAP was found to originate from the bony cartilaginous junction and fill the deep bony part of the canal. To my knowledge, only one study has demonstrated the case of a patient with SCAP arising from the bony part of the canal [7].

Radiological examinations such as computed tomography are mandatory in a case of polypoid lesions obstructing the ear canal as possibilities of a surprising pathology like foreign body [2] or any other unexpected pathology.

Magnetic resonance (MR) imaging is also a useful technique for polypoid lesions of the external auditory canal. Recently, Kamakura et al. demonstrated the MR imaging criteria of SCAP of the external auditory canal as intermediate signal intensities on T1- and T2-weighted images and slight enhancements on gadolinium-enhanced T1-weighted images [5].

Histological diagnosis is the best tool for such polypoid lesions in the ear canal, and in these cases, excision biopsy is better than incision biopsy to avoid the risk of being potentially malignant mass [11], not having sufficient biopsy tissue, or uncontrollable hemorrhage in glomus or facial palsy in schwannomas [5].

Regular follow-up should be done for such a tumor for the fear of recurrence.The patient with SCAP presented in this study had been followed for 9 months after complete excision, and no recurrence occurred during this follow-up period.

Mass lesions in the external auditory canal are difficult to diagnose based on clinical and radiological data. Histopathology usually represents the main diagnostic tool in these cases. SCAP, despite its rarity, should be kept in mind in the differential diagnosis of mass lesions in the external auditory canal. A rare site of origin of such a lesion had been described in the present study.

**Informed Consent:** Written informed consent was obtained from the patient who participated in this study.

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