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Case Report

Syringocystadenoma Papilliferum Revealed 12 Years After Surgical Treatment of Chronic Ear with Cholesteatoma: Presentation of an Unusual Case and Literature Review

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Syringocystadenoma papilliferum is a rare hamartomatous adnexal tumor and appears mainly in the head and neck region. Rarely, such tumors may appear in the external auditory canal. There are only 14 described cases of the syringocystadenoma papilliferum in the external auditory canal in the Medline database. In this study, we present the first reported case of syringocystadenoma papilliferum arising in the modified radical mastoidectomy and occurring 12 years after ear surgery. A 26-year-old man was admitted to our clinic with complaints of discharge from the left ear, ear fullness, and hearing loss. These symptoms had been worsening for 3 years. Twelve years previously, he had undergone modified radical mastoidectomy by Bondy due to a cholesteatoma. Otoscopic examinations revealed otorrhea and a reddish mass measuring 1 × 0.8 cm arising from the tegmen tympanum and fundus of the mastoid cavity. A surgical biopsy was conducted with local anesthesia in the medical outpatient clinic, and it revealed syringocystadenoma papilliferum. A tumor resection was performed with a retroauricular approach. The bone attachment of the tumor was drilled out, and the ear cavity was covered with a mashed split skin graft. The ear cavity healed, and no tumor recurrence was observed during a 9-month follow-up period. Our case and literature analysis of previously described cases led to the conclusion that cases of syringocystadenoma papilliferum of the external auditory canal may be associated with different medical histories, age distributions, and origins than other syringocystadenoma papilliferums of the head and neck region. Tubular adenoma may occur together with syringocystadenoma papilliferum in 20% of cases. Malignant transformation of syringocystadenoma papilliferum may occur but is extremely rare. Computed tomography imaging may predict malignancy of the tumor. Tumors specific to the external auditory canal may occur in the modified radical mastoidectomy after middle ear surgery.

KEYWORDS: Syringocystadenoma papilliferum, SCAP, cholesteatoma surgery, open ear cavity, modified radical mastoidectomy, tubular adenoma

INTRODUCTION

Syringocystadenoma papilliferum (SCAP) is a rare hamartomatous adnexal tumor that originates from the apocrine or eccrine sweat glands and appears mainly in the head and neck region. More than half of these tumors are present from birth or appear in early childhood.[1,2] Rarely, such tumors may appear in the external auditory canal as a reddish mass blocking the auditory canal and causing foreign body sensation, inflammation, otorrhea, and finally earache.[3] Ear obstruction leads to complaints of conductive hearing loss. The lesion does not look like an inflammatory polyp but rather a reddish or gray firm mass tender to the touch. There are only 14 described cases of SCAP in the external auditory canal in the Medline database. In this study, we present the first reported case of SCAP arising in the modified radical mastoidectomy and occurring 12 years after ear surgery.

CASE PRESENTATION

A 26-year-old man was admitted to our clinic with complaints of discharge from the left ear, ear fullness, and hearing loss. He reported that these symptoms had been worsening for 3 years. Twelve years previously, he had undergone ear surgery due to a

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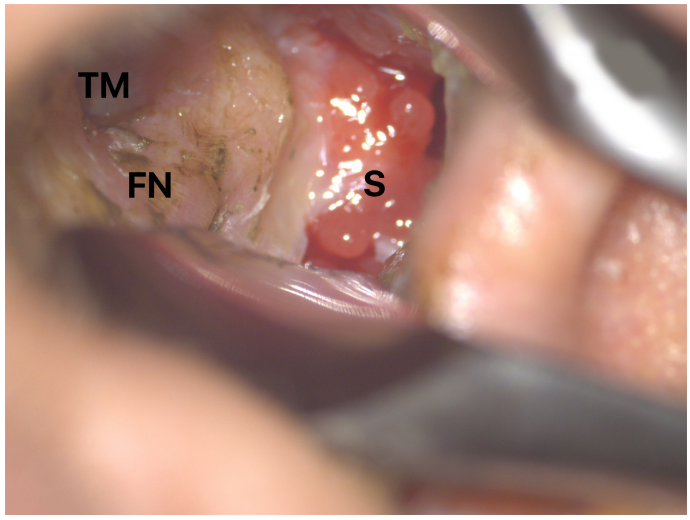

Figure 1. Left ear otomicroscopy before surgery—residual part of the tumor—syringocystadenoma (S) after surgical biopsy; TM, tympanic membrane; FN, facial nerve.

cholesteatoma. A canal wall down procedure, as described by Bondy, was performed with use of Koerner's skin flap for partially modified radical mastoidectomy coverage. The patient was regularly followed up for 6 years, after which he neglected follow-ups due to the absence of symptoms and no recurrence of cholesteatoma. Otoscopic examinations revealed otorrhea and a reddish mass measuring 1 × 0.8 cm arising from the tegmen tympani and fundus of the mastoid cavity (Figure 1). A surgical biopsy was conducted in outpatient clinic with local anesthesia, and it revealed SCAP. High-resolution computed tomography of the temporal bones revealed a mass with soft-tissue density in the superior-posterior part of the open ear cavity, without bone erosion (Figure 2). Pure tone audiometry revealed conductive hearing loss with a hearing level of 20-30 dB. A tumor resection was performed with a retroauricular approach. The tumor was broadly based against the bony surface of the tegmen, its' bone attachment was drilled out, and the ear cavity was covered with a mashed split skin graft from the thigh. Postoperative histological and immunohistological examination of the tumor confirmed SCAP. The ear cavity healed, and no tumor recurrence was observed during a 9-month follow-up period (Figure 3). Informed consent for treatment was signed by the patient before surgery.

MAIN POINTS

- This study describes the case of syringocystadenoma papiliferum of the middle ear. To the best of our knowledge, there is only 14 described cases of syringocystadenoma papiliferum in external auditory canal in Medline database.
- This is the first case occurring in a modified radical mastoidectomy after previous canal wall down middle ear surgery for cholesteatoma 12 years earlier.
- For this type of tumor, we applied tumor resection and bony margin of the tumor attachment drilled down and finally bone cavity coverage with mashing split skin graft for coverage the cavity.
- Additionally, we present a review of anatomic localization of syringocystadenoma papiliferum of the ear, performed surgical technics, and results.

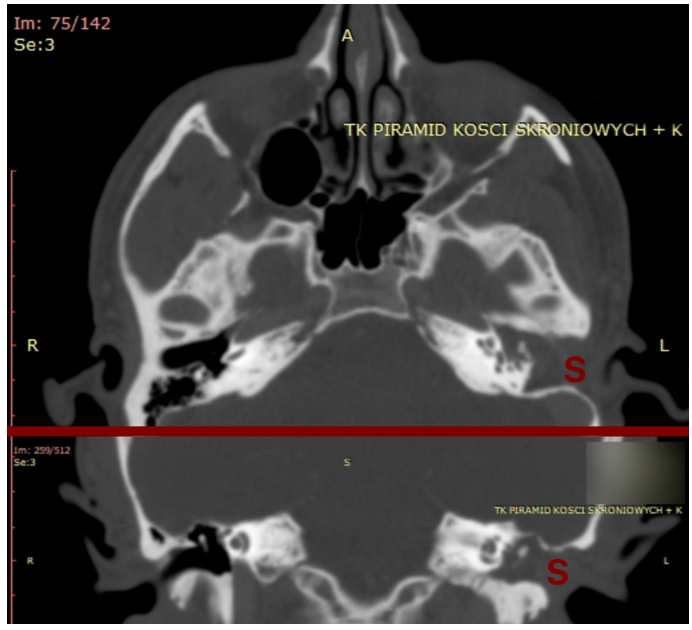

Figure 2. CT imaging of the ear—mass of the syringocystadenoma in the left ear (S). CT, computed tomography.

DISCUSSION

Syringocystadenoma papilliferum is a very rare tumor of the external auditory canal that may have a long course of development until obturation of the external ear canal occurs and the patient complains of ear fullness, pain, otorrhea, and hearing loss.[3-7] In addition, it seems to be associated with different medical history characteristics and to have a different origin than other SCAP of the head and neck region. The analyzed literature shows that it typically occurs in adults.[3-14] There is only 1 case of pediatric SCAP, described in 15-year-old male.[15] In addition, our presented case confirms the distinct age distribution in the occurrence of this tumor in the EAC, because the patient did not report an SCAP tumor in his childhood medical history when he underwent ear surgery. Certainly, in this localization, SCAP is not associated with a congenital skin lesion

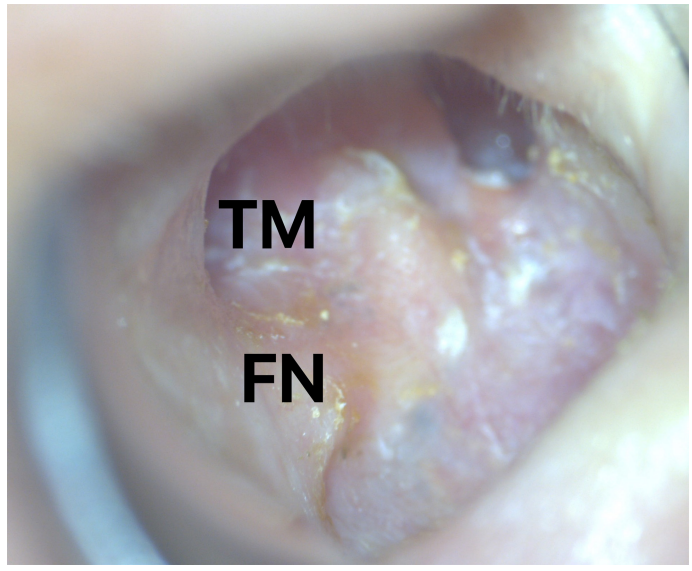

Figure 3. Left ear control otomicroscopy (9 months after surgery); TM, tympanic membrane; FN, facial nerve.

Table 1. Review of the Cases Reported in the Literature

First Author (Year)	Patients' Age	Gender	Side	Tumor Dimension	Pathology Result	Symptoms	Tumor Localization	EAC Bone Destruction	Imaging/ Approach-Surgery/ Aanesthesia	Time to Diagnosis	Free Follow-Up
Guerra-Jimenez G (2016)	62	F	R	0.3 cm	SCAP	Conductive hearing loss, tympanic membrane subtotal perforation	EAC—posterior wall	-	No/ biopsy/ local	-	-
Guerra-Jimenez G (2016)	55	M	R	Fully obturated EAC	SCAP	Conductive hearing loss, otorrhea,	Middle and external part of EAC	No	CT/ combine approach—retroarticular and endaural/ general	15 years	-
Khurayzi T (2017)	50	F	R	1 × 1 cm	SCAP	Foreign body sensation	Roof of the EAC	No	CT, MRI/ endaural/ local	-	-
Aydin. N. (2018)	40	F	R	0.8 × 0.6 × 0.3 cm	SCAP	EAR fullness	Bony part of EAC	-	-/endaural several attempts/ biopsy/ local	Several years	Several attempts
Bruschini L (2017)	72	M	L	1 × 0.6 cm	SCAP	Hypoacusis	Half of the posterior wall of the EAC	No	CT/ retroarticular/general	3 months	12 months
Arechvo A (2013)	61	F	R	Fully obturated EAC	SCAP	Ear pain, fulness, hearing loss, tinnitus, otorrhea	Bony part of the EAC	No (bony part widened)	CT/ endaural, transposition of the skin flap for defect coveradge/general	4 months	9m
Kamakura T (2005)	57	M	L	Fully obturated EAC-4 cm	SCAP *Cholesteatoma in the tympanic cavity*	Ear fullness, Conductive hearing loss	EAC+ preauricular area	-	MRI/ combine approach—retroarticular and endaural/general	10 years	-
Alzamil W (2017)	29	F	L	0.5 × 0.3 × 0.4	SCAP	Ear pain, hypoacusis	EAC+ facial recess	No incudo-stapedial joint eroded	CT/ retrograde atticotomy, incus interposition, myringoplasty/ general	3 months	Regular follow up- free from disease
Muller R (1995)	65	F	R	Fully obturated EAC	SCAP	Ear fulness, hearing loss	EAC— posterior wall	No	CT/ endaural/-	-	-
Arava S (2021)	15	M	R	0.5 cm	SCAP	History of *chronic otitis media*, ear pain, discharge	EAC—tiny mass	-	-/ excision/-	-	-
Su T (2011)	25	M	L	0.8 × 0.6 cm	SCAP+TA	Hypoacusis, otorrhea	EAC near the opening	No	CT/ endaural excision/-	Period of time	9 months
Kuczkowski J (2010)	57	F	R	0.5 × 0.6 cm	SCAP+TA	no	EAC (50% obstruction of EAC)	No	CT/ retroauricular, full thickness skin graft for coverage of the EAC/ general	-	2 years
Lee (2005)	74	F	L	1.5 × 1 cm	SCAP+TAA	Hypoacusis, otorrhea	EAC—posterior superior wall	No	CT/ excision+ split thickness skin graft/ -	10 years	8 months
Diaz R (2007)	49	M	L	Fully obturated EAC	*Ductal carcinoma* arising from SCAP	Foreign body sensation, otorrhea, ear pain	EAC—posterior wall (bony cartilaginous junction)	2 mm *bone defect*	CT/lateral temporal bone resection/ general	6 months	20 months
Zwierz A	26	M	L	1 × 0.8 cm	SCAP	Otorrhea, hypoacusis, *12 years after cholesteatoma surgery*	Tegmen and fundus of the cavity	No	CT/ retroauricular, mashed split thickness skin graft for coverage of the ear cavity/ general	3 years	9 months

EAC, external auditory canal; TA, tubular adenoma; TAA, tubular apocrine adenoma; SCAP, syringocystadenoma papilliferum; CT, computed tomography; MRI, magnetic resonance imaging; M, male; F, female; R, right; L, left.

known as nevus sebaceous. To the best of our knowledge, this is the first case of occurrence of SCAP many years after ear surgery where the patient was frequently monitored by an experienced ear, nose, and throat doctor. It is also the second case of SCAP linked with cholesteatoma and the third case related to chronic ear inflammation.[6,15] There was no chronic ear history in the other 12 analyzed cases.[3-5,7-14]

In many cases, the tumor appears to originate in the skin of the posterior-upper wall of the EAC at the border of the cartilage–bone junction.[3-5,7-10,14] This is the cartilaginous two-thirds part of the EAC where the ceruminous glands are located.[10] In the described case, the appearance of the tumor in an unusual location—the tegmen and fundus of the modified radical mastoidectomy—could also have been caused by transposition of skin rich in ceruminous glands from the posterior wall of the external auditory canal during the primary ear surgery.

Frequently, SCAP and tubular adenoma (TA) may occur together. In the analyzed literature, 3 out of 15 cases showed the coexistence of both pathological types of tumors (Table 1).[12-14] Further, Diaz[3] described a case of ductal carcinoma arising from SCAP. In this case, a bony defect was observed via computed tomography (CT) imaging. A preserved tympanic membrane and a lack of bony erosion in the external auditory canal revealed via CT scan may therefore indicate the absence of tumor malignancy.[4,5,7,8,10,12,13] Biopsy and CT scanning seem to play important roles in surgical decision-making concerning the range of the procedure.[11] A retroauricular or endaural surgical approach may be used to reduce the risk of tumor recurrence, but surgical biopsy does not guarantee radicality treatment (Table 1).[9] In some cases of large skin resection, bone coverage with a skin graft may accelerate healing.[5,13,14] Both full-thickness and split-thickness skin grafts are used with good results.[13] Due to the risk of massive skin loss in the ear cavity, we used a mashed split skin graft and achieved a well-epithelialized cavity.

CONCLUSION

Our case and literature analysis of previously described cases led to the conclusion that cases of SCAP of the EAC may be associated with different medical histories, age distributions, and origins than other SCAPs of the head and neck region. In 20% of cases, SCAP and TA may occur together. Malignant transformation of SCAP may occur but is extremely rare. Computed tomography imaging may predict malignancy of the tumor. Tumors specific to the external auditory canal may occur in case of modified radical mastoidectomy after middle ear surgery.

Informed Consent: Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient/parent/guardian/ relative of the patient.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – A.Z.; Design – A.Z.; Supervision – A.Z., P.B.; Materials – A.Z.; Data Collection and/or Processing – A.Z., K.M.; Analysis and/or Interpretation – A.Z.; Literature Review – A.Z.; Writing Manuscript – A.Z.; Critical Review – P.B.

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