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# Schneiderian-Type Papilloma of the Middle Ear: A Review of the Literature

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**Schneiderian-type papilloma of the middle ear is a rare finding. We present a 46-year-old Aboriginal man with a large tympanic membrane perforation and a Schneiderian-type papilloma filling the middle ear. The aim of this study is to familiarize clinicians with this uncommon disease through discussion of its clinical presentation, diagnostic considerations and management. A search of English-language peer-reviewed literature was undertaken using the key words "Schneiderian-type papilloma," "inverted papilloma," and "middle ear." A total of 29 cases (including the present case) of Schneiderian-type papilloma involving the middle ear were reviewed. Common presenting symptoms include hearing loss, otalgia, and otorrhea. Middle ear disease is associated with higher rates of recurrence and malignant transformation than its sinonasal counterpart. Radical surgical resection is the only curative treatment. Schneiderian-type papilloma is a benign, but locally aggressive, epithelial neoplasm most commonly arising in the sinonasal tract. Whilst involvement of the middle ear is extremely rare, knowledge of this condition is important due to its propensity to recur and the high rate of malignant transformation.**

*Key words:* Inverted papilloma – Papilloma – Middle ear – Nasal mucosa

Schneiderian-type papilloma arising from the sinonasal tract is uncommon but well known. Middle ear disease is extremely rare and, to our knowledge, there are only 28 reported cases in the English literature. Patients typically present with hearing loss, otorrhea, otalgia, and aural fullness. The etiology is unknown and radical surgical resection is the only curative treatment. Middle ear disease is

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associated with higher rates of recurrence and malignant transformation then sinonasal disease, necessitating lifelong monitoring. The aim of this study is to familiarize clinicians with this uncommon disease through discussion of its clinical presentation, diagnostic considerations and management.

## Case Report

A 46-year-old aboriginal man in a remote community in Far North Queensland was seen by a visiting otolaryngologist with the presenting complaint of hearing loss and right-sided otorrhea. Examination revealed a large right tympanic membrane perforation, through which an irregular papillomatous mass could be seen occupying much of the middle ear space. Purulent otorrhea was noted in the right external auditory canal. A left tympanic membrane perforation was also present, but no papillomatous changes were seen. Pure tone audiometry demonstrated 25 dB conductive hearing loss bilaterally. Nasal endoscopy was normal. Preoperative computed tomography (CT) imaging of the temporal bones demonstrated significant soft tissue within the right middle ear cavity with erosion of the scutum and tegmen tympani. Erosion of the incus and stapes was also evident. Extensive soft tissue thickening and sclerosis was demonstrated throughout the mastoid air cells. The left side also demonstrated mild soft tissue thickening within the middle ear and destruction of the ossicular chain. No Eustachian tube pathology was identified.

A right modified radical mastoidectomy was performed. Intra-operatively, the middle ear cavity was found to be filled with polypoid tissue. A large dehiscence of the tegmen tympani was identified, with polypoid tissue covering the defect. The long process of incus had been eroded. A small keratin pearl was found at the orifice of the Eustachian tube in the protympanum, but the Eustachian tube was otherwise normal. The middle ear cavity was cleared of polypoid tissue except for a small amount left over the tegmen tympani dehiscence, which was cauterized. Histopathology revealed Schneiderian-type papilloma of the middle ear, with no evidence of malignancy (Fig. 1). The patient is now 18 months post-surgery with no evidence of recurrence.

## Materials and Methods

A search of English-language peer-reviewed literature was undertaken using Medline and PubMed, using the key words "Schneiderian-type papilloma," "inverted papilloma," and "middle ear." All reported cases from the literature were reviewed.

## Results and Analysis

A total of 29 cases, including the present case, were identified and reviewed from 1987 to 2012.[1–25] Age at presentation ranged from 9 to 81 years, with a mean age of 48 years. Peak incidence occurred in the sixth decade of life. There was no sex bias, with 14 male patients and 15 female patients reported. Presenting symptoms were reported in 23 cases. The most commonly reported symptoms were hearing loss (78%), otorrhea (39%), otalgia (30%), and aural fullness (17%). Ipsilateral facial nerve paralysis (4%), tinnitus (4%), and ipsilateral headache (4%) were also reported. Nine patients (31%) were reported to have right-sided disease, 14 (48%) had left-sided disease, and 2 patients (7%) had bilateral disease. Side of disease was not reported in 4 cases (14%). Fifteen patients (52%) had previous or concurrent sinonasal disease. Involvement of the Eustachian tube was clinically or radiologically identified in 7 cases (24%). Carcinoma in situ was identified in 5 patients (17%) and invasive squamous cell carcinoma was identified in 7 patients (24%). Surgical treatment and follow up was reported in 21 cases. Tympanoplasty and simple excision was the initial surgical treatment in 3 cases, with all patients experiencing recurrence. Eighteen patients were treated with more aggressive primary surgery (tympanomastoidectomy or temporal bone resection), of which 7 (39%) had at least one recurrence of disease.

## Discussion

Schneiderian-type papillomas were first described in 1854 and were named in reference to C. Victor Schneider, who described the ectodermal origin of the nasal mucosa.[22] Schneiderian-type papillomas are most commonly found in the sinonasal tract, with an incidence of 1.5 cases per 100,000 population.[26] Involvement of the middle ear is even rarer and was first described by Stone *et al*[1] in 1987. Since that time, only 28 cases have been reported in the English literature.

The etiology of Schneiderian-type papilloma remains unclear. Stone *et al*[1] first hypothesized the mechanism of direct extension through the Eustachian tube. Seven cases (24%) reported clinical or

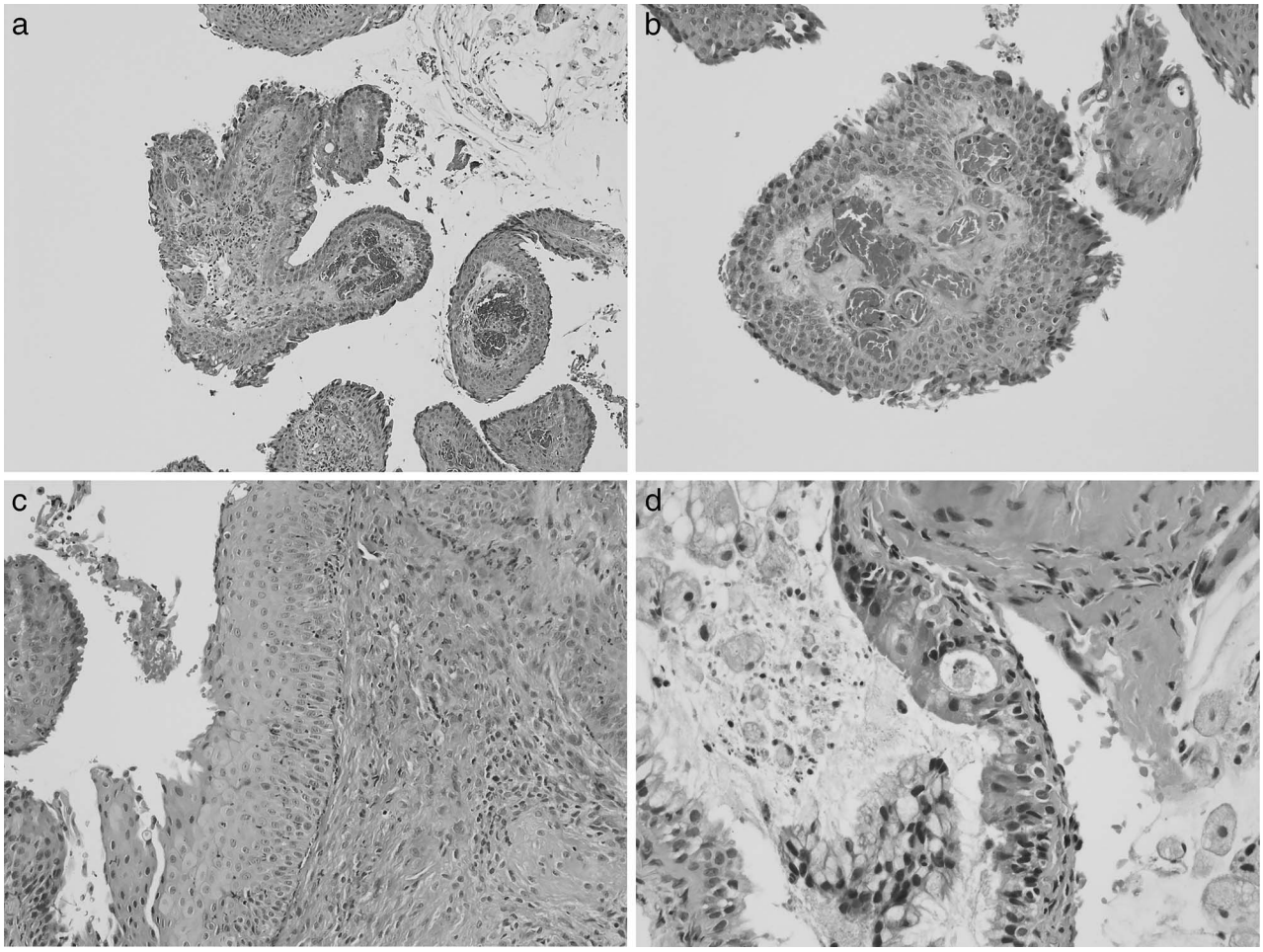


**Fig. 1** (a) Low power view demonstrating the papillary architecture, with fibrovascular cores covered with stratified epithelium. (b) Fibrovascular core, with hyalinized collagen and capillary-sized vessels, covered with transitional epithelium. (c) Mixed inflammatory cell infiltrate within the fibrovascular core and squamous/transitional epithelium. (d) Ciliated respiratory-type epithelium with many goblet cells, muciphages and a microcyst. *Staining method: hematoxylin and eosin staining.

radiological involvement of the Eustachian tube. However, several authors have reported normal Eustachian tubes in their patients.[3,4,10,16,22] This theory also cannot account for the 13 cases (45%) of middle ear Schneiderian-type papilloma without previous or concurrent sinonasal disease. On this basis, it has been hypothesized that Schneiderian-type papillomas arise from ectopic inclusion of ectodermal Schneiderian mucosa, the embryonic origin of sinonasal mucosa, into the endodermally derived mucosa of the middle ear.[7] In our case, our patient did not report any nasal symptoms and no sinonasal disease or Eustachian tube involvement was detected on pre-operative CT imaging or intra-operatively.

Histopathological features of Schneiderian-type papilloma of the middle ear are identical to those for sinonasal disease. Characteristic features of this lesion are fibrovascular cores covered by transitional, squamous, and ciliated respiratory type epithelium (Fig. 1).[27] Schneiderian-type papillomas may also exhibit microcysts, muciphages, goblets cells and an inflammatory infiltrate. Malignant transformation in sinonasal inverted papilloma is well recognized and is reported at 5 to 13%.[15] Schneiderian-type papilloma of the middle ear appears to reflect a greater association with malignancy, with a malignant transformation rate of 41% according to the analyzed cases.

Management of Schneiderian-type papilloma of the middle ear, as with sinonasal disease, is primarily surgical. Inadequate resection is associated with higher rates of recurrence. Excluding the 8 cases in which surgical management or follow-up

data is not reported, at least one recurrence occurred in 10 (48%) of the 21 eligible cases. Recurrence was reported in all 3 cases (100%) initially treated with tympanoplasty and simple excision, compared to 39% following more aggressive surgery (mastoidectomy or temporal bone resection). This compares with a recurrence rate of 40 to 78% for sinonasal inverted papilloma with limited surgery, and 0 to 14% with more aggressive surgery.[21]

Long-term follow up is essential in all patients with Schneiderian-type papilloma of the middle ear because of the propensity for recurrence and the high rate of malignant transformation. As for sinonasal inverted papilloma, magnetic resonance imaging (MRI) is recommended for follow up imaging, as it is more sensitive and specific than CT in differentiating recurrent tumor from adjacent inflammatory tissue.[21,23]

## Conclusion

Schneiderian-type papilloma of the middle ear is an extremely rare tumor, with only 28 cases reported in the English literature since its initial description in 1987. Patients typically present with hearing loss, otorrhea, otalgia, and aural fullness. Etiology is not yet known, although several hypotheses exist. Radical resection is the only curative treatment. Lifelong follow up is essential due to its propensity for recurrence and high rate of malignant transformation.

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