

รายงานผู้ป่วย

Neurofibroma of the chorda tympani nerve.

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Neurogenic tumors of the facial nerve and its branches in the middle ear and the temporal bone are rare. Almost all of them presents as neurilemmoma. The author reports the first solitary neurofibroma of the chorda tympani nerve coincidentally found during tympanoplastic procedure. The treatment was simple resection and the tumor has not recurred.

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รายงานผู้ป่วย 1 ราย ที่พบเนื้องอกนิวโรไฟโบรมาของเส้นประสาทคอรัดา ทิมปานีโดยบังเอิญขณะ ทำการผ่าตัดซ่อมแซมหูชั้นกลางและเยื่อแก้วหู ได้ตัดเนื้องอกออกทั้งหมดทั้งก้อนและไม่กลับเป็นใหม่อีก นับว่าเป็นเนื้องอกชนิดนี้รายแรกของโลกที่พบเกิดจากเส้นประสาทคอรัดา ทิมปานี

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Middle ear neoplasms are rare and are commonly divided into two categories—the malignant and the benign tumors. Among the malignant tumors, the squamous cell carcinoma is the most common but adenocarcinoma and adenoid cystic carcinoma are also frequently reported.⁽¹⁾ Glomus tumor is the most important of the benign type. Nerve sheath tumors, congenital cholesteatoma, meningioma, adenoma and hemangioma also present as middle ear masses.⁽¹⁻²⁾

Intratympanic nerve sheath tumors, the neurilemmoma and neurofibroma, usually arise in the facial nerve especially in the geniculate ganglion.⁽²⁻³⁾ Branches of the facial nerve have been reportedly involved by these tumors⁽⁴⁻¹¹⁾. The less common neurofibroma presents as either a facial nerve involvement in von Recklinghausens disease or a solitary mass^(7,12). Goethem in 1985 reported a case of tactile neurofibroma issuing from the stapedial nerve.⁽⁹⁾ We present a patient with a solitary neurofibroma of the chorda tympani nerve which was coincidentally discovered during tympanolasty surgery. We believe, to the best of our knowledge, that this is the first case of chorda tympani neurofibroma to be reported.

Case report

The patient was a 40-year-old Thai female who came to the E.N.T. department of Chulalongkorn Hospital with a long history of left ear infection and hearing loss, of more than 30 years. She had aural discharge on and off, ranging from serous to purulent. Otoscopic examination revealed a normal appearance on the right side and a subtotal tympanic membrane perforation on the left. The mucosa of the middle ear was somewhat thickened without cholesteatoma or granulation tissue. Audiologic tests showed a mild sensorineural hearing loss of 5 to 10 dB in the normal ear and a mixed hearing loss with an air conduction threshold of 70 dB and air-bone gap of 30 dB in the diseased ear. The remaining examination and laboratory work-ups were unremarkable.

She was scheduled for a tympanoplastic surgery to her left ear. The operation was undertaken through a 6-mm. Shea's aural speculum, by the senior author (V.S.). The findings were consistent with chronic middle ear diseases, the malleus handle and the long process of incus were missing. The chorda tympani nerve was found with a 0.5 cm bulbous enlargement of the proximal half. (Fig. 1) The stapes was intact and mobile. Removing all the pathologic tissues, the ossicular chain was reconstructed by an autograft incus bridging between the malleus and head of the stapes. The chorda tympani mass was resected and sent for histopathology. Lateral placing temporalis fascia graft and canal skin flap were

employed to cover the tympanic membrane defects and gelfoam packing was used to stabilize the graft. Postoperative recovery was uneventful and she was discharged on the third postoperative day. The packing was removed on the tenth postoperative day when the graft appeared healthy and remained so for at least 1 year, with acceptable serviceable hearing. Microscopic report of the specimen was compatible with neurofibroma. (Fig. 2)

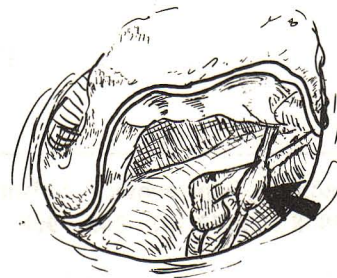


Figure 1 The bulbous enlargement of the proximal half of chorda tympani nerve (arrow) was found during surgery.

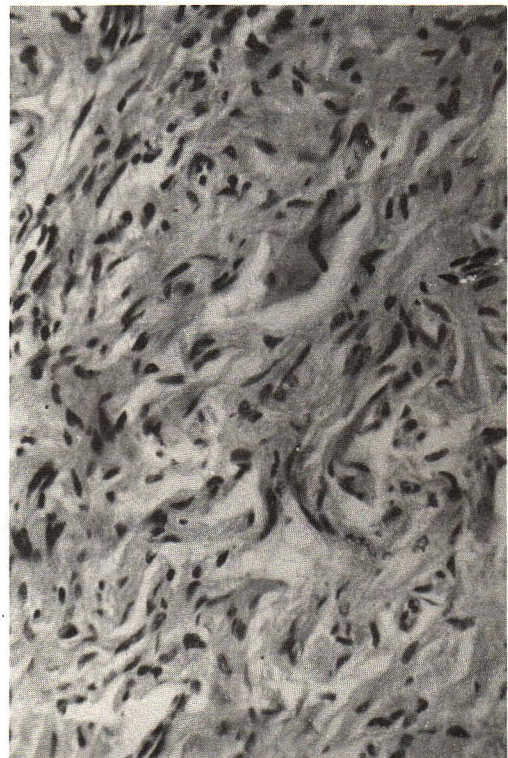


Figure 2 The non-encapsulate tumor consists of elongated cells with thin nuclei arranged in between the wavy collagen fibers. (H&E.-High power).

Discussion

Neurilemmomas of the middle ear are commonly found originating from the facial nerve, autonomic nerves and the hypoglossal nerve.^(1,13) Facial nerve neurilemmoma is believed to arise from the sensory component of the facial nerve.⁽¹⁴⁾ The presenting symptoms can be divided into three groups.⁽¹⁵⁾ 1) Facial nerve dysfunctions, paralysis and hyperkinesia especially with hemifacial spasm. The facial nerve paralysis is usually slowly progressive in nature and remains for more than three weeks duration. 2) A hearing loss which may be of sensorineural or conductive type according to the location and size of the tumor. 3) Otalgia.

The neurofibroma arise from the Schwann's cells or perineural cells which surround the nerve bundle.⁽¹⁶⁾ The more common form is the multiple neurofibromas or von Recklinghausen disease. Solitary or simple neurofibroma is rarely seen but occurs mostly in the middle ages (30-40 years)^(12, 17) Histologic features of the neurofibroma consist of elongated cells arranged in between the collagen and reticulin fibers. Their nuclei are thin and elongated. The fibers are arranged in wavy pattern along the course of the nerve. Between these fibers, mucinous or myxoid ground substance is present with lymphocytic and macrophage infiltration. Ten to fifteen per cent of these tumors especially from von Recklinghausen's cases undergo malignant transformation.⁽¹⁶⁾ In our patient, all systems were reexamined

especially the skin, ear, nose, throat, eye and neurological system, searching for evidences of von Recklinghausen's disease but none was found including the family history of this disease.

Solitary or simple neurofibroma of the chorda tympani nerve was then concluded. Searching through the literature, no report in English and Thai language has mentioned of this entity. Because this tumor was too small and found coincidentally during tympanoplasty, the functions of the nerve was not previously determined. We believe that this should be the first reported case of chorda tympani neurofibroma in the world.

Summary

The authors present a case of chorda tympani neurofibroma in a 40-year-old woman, being found co-incidentally during a tympanoplastic surgery. The tumor was too small to have presented with symptoms and the treatment in this case was a simple excision. The author believe that it was the first case of solitary neurofibroma arising in the chorda tympani nerve ever reported.

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