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Alexander T. Laoag, M.D.[1]
Antonio H. Chua, M.D.[1, 2]
Thanh Vu T. De Guzman, M.D.[1]
Samantha S. Castañeda, M.D.[1, 3, 4]
Jose A. Malanyaon, Jr., M.D.[1,5]

[1]*Department of Otorhinolaryngology Head and Neck Surgery Jose R. Reyes Memorial Medical Center*

[2]*Department of Otorhinolaryngology Head and Neck Surgery University of the East – Ramon Magsaysay Memorial Medical Center*

[3]*Department of Otorhinolaryngology Head and Neck Surgery The Medical City*

[4]*Department of Otorhinolaryngology Head and Neck Surgery Rizal Medical Center*

[5]*Department of Otorhinolaryngology Head and Neck Surgery East Avenue Medical Center*

Intraparotid Facial Nerve Schwannoma with Temporal Bone Extension

ABSTRACT

Objective: To present a rare case of facial schwannoma manifesting as a parotid mass and discuss its diagnosis and treatment.

Methods:

Design: Case Report
Setting: Tertiary Government Hospital
Patient: One

Results: A 48-year-old female was seen for a 2-year progressive left hemifacial paralysis and a 5-month gradually enlarging left infraauricular mass with episodes of tinnitus but intact hearing and balance. Physical examination showed a left-sided House-Brackmann grade VI facial paralysis and a 5 x 4 x 3 cm soft, ill-defined, slightly movable, nontender, left infraauricular mass. Gadolinium-enhanced magnetic resonance imaging revealed a 5 cm heterogeneously-enhancing lobulated mass centered within the deep lobe of the left parotid gland extending to the left mastoid, with facial nerve involvement. A diagnosis of a facial nerve tumor, probably a schwannoma, was entertained. Pure tone audiometry revealed normal hearing thresholds for both ears with dips at 6-8 KHz on the left. The patient underwent total parotidectomy with facial nerve tumor resection via transmastoid approach, with simultaneous facial – hypoglossal nerve anastomosis reconstruction. Histopathologic findings confirmed the diagnosis of a schwannoma.

Postoperative facial function was Grade VI. Hearing and hypoglossal nerve function were preserved.

Conclusion: A progressive hemifacial paralysis of chronic duration with or without the presence of an infra-auricular mass should raise the suspicion of a facial nerve tumor. Gadolinium-enhanced magnetic resonance imaging is valuable since intraparotid facial nerve schwannomas are mostly diagnosed intraoperatively when the neoplasm and the nerve are exposed and determined to be contiguous. The clinician should be aware that not all parotid masses are salivary gland in origin.

Keywords: *intraparotid facial nerve schwannoma, facial nerve paralysis, parotid mass*

Correspondence: Dr. Alexander T. Laoag
Department of Otorhinolaryngology
Jose R. Reyes Memorial Medical Center
San Lazaro Compound, Rizal Ave., Sta. Cruz, Manila 1003
Philippines
Phone: (632) 711 9491 local 320
Fax: (632) 743 6921
E-mail: alexanderlaoag_md@yahoo.com
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Gradual facial paralysis in a patient with infra-auricular mass most often indicates a parotid malignancy.[1] However, if the tumor grows toward the facial canal, facial nerve schwannoma should be suspected.[2]

Schwannomas are benign, slow growing neoplasms of ectodermal origin that arise from Schwann cells. These tumors are usually solitary, well encapsulated and tend to splay the nerve of origin. Although approximately one third of all of these lesions occur in the head and neck, facial nerve schwannomas are rare with less than 500 cases reported in literature — mostly intratemporal and less than 70 with parotid involvement. [3]

The estimated frequency of parotid tumors originating in the facial nerve ranges from 0.2% to 1.5%. Preoperative diagnosis of this tumor in the parotid gland is usually difficult because of the low occurrence of the disease and few distinctive signs associated with it. [1, 2] Patients may present with a painless facial mass, progressive facial weakness or paralysis and audiovestibular symptoms (hearing loss, tinnitus, and instability). Despite a characteristic microscopic appearance, the firmly attached cells in tumors of neurogenic origin make obtaining positive cytology difficult; therefore, fine needle aspiration may not be a useful diagnostic modality in the evaluation of parotid schwannoma. A gadolinium-enhanced magnetic resonance imaging might only be indicative but not pathognomonic. Therefore, intraparotid facial nerve schwannomas are mostly diagnosed intraoperatively when the neoplasm and the nerve are exposed and directly visualized.[4] Management is quite difficult considering the possible need for resection resulting in severe facial nerve paralysis with important aesthetic and functional consequences.

The aim of this article is to discuss a rare case of a facial nerve schwannoma initially presenting as an infraauricular mass preceded by a long-standing facial nerve paralysis.

CASE REPORT

A 48-year-old female consulted with a 2-year history of progressive left hemifacial paralysis and a 5-month history of a gradually enlarging left infraauricular mass. She complained of episodes of tinnitus but hearing and balance were intact. She was initially treated by a general practitioner for acute cerebrovascular infarct versus Bell's palsy. Gadolinium-enhanced magnetic resonance imaging (MRI) of the brain revealed a 5 cm heterogeneously-enhancing lobulated mass centered within the deep lobe of the left parotid gland extending to the left mastoid with facial nerve involvement. *(Figure1)*

She was referred to our department. Physical examination findings showed a left-sided House-Brackmann grade VI facial paralysis and a 5 x 4 x 3 cm soft, ill-defined, slightly movable, nontender, left infraauricular mass. Otoscopy was unremarkable. Tympanic membranes were both intact. There was no bulging or medialization of the left tonsillar area. A parotid malignancy was initially suspected. Upon review of the MRI, a facial nerve tumor, probably a schwannoma was entertained. Pure tone audiometry (PTA) revealed normal hearing threshold in both ears, with

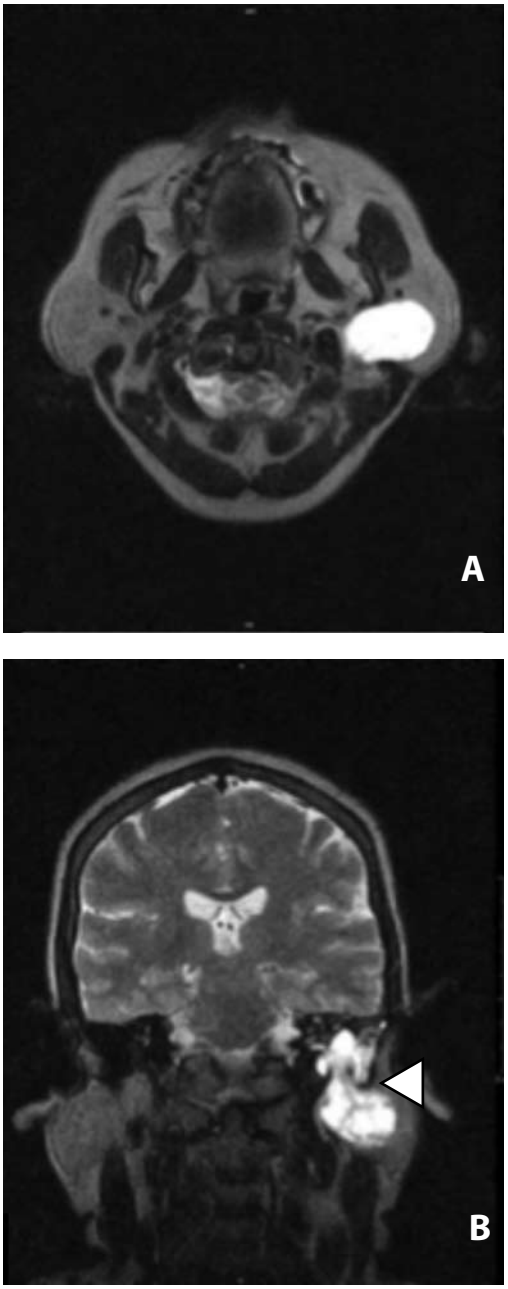
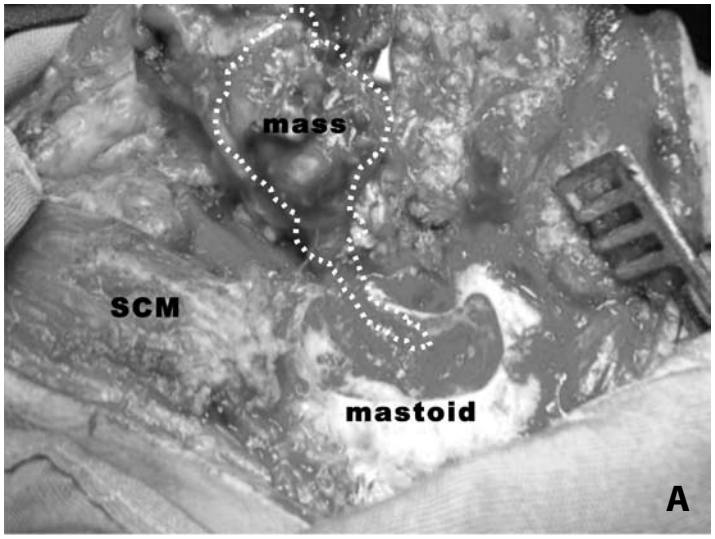
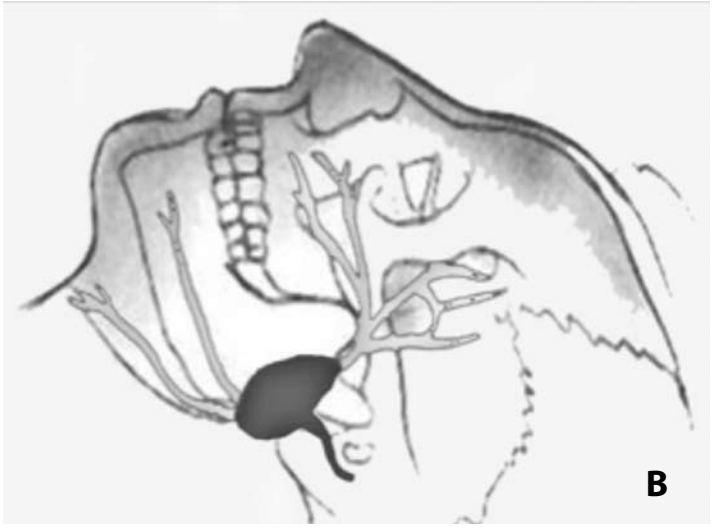
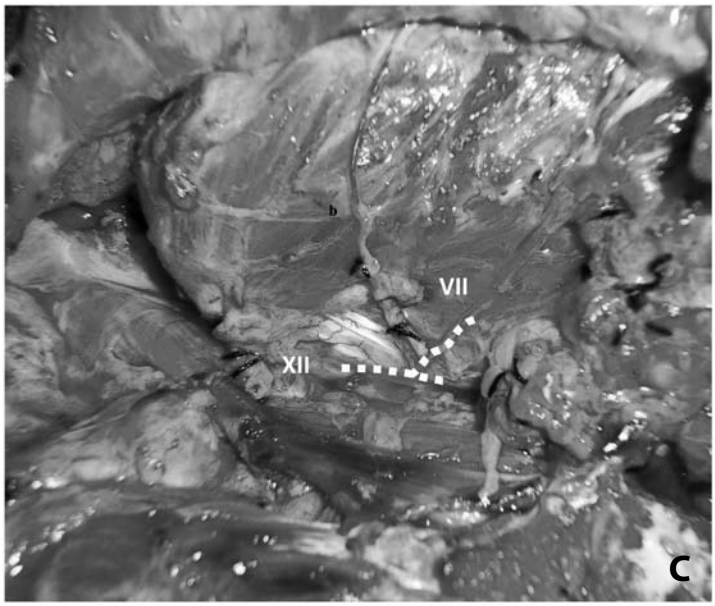


Figure 1. T2-weighted Magnetic Resonance Images showing the 5 cm heterogeneously enhancing lobulated mass centered within the deep lobe of the left parotid gland **A.** Extending to the left mastoid with facial nerve involvement **B.** Arrowhead.







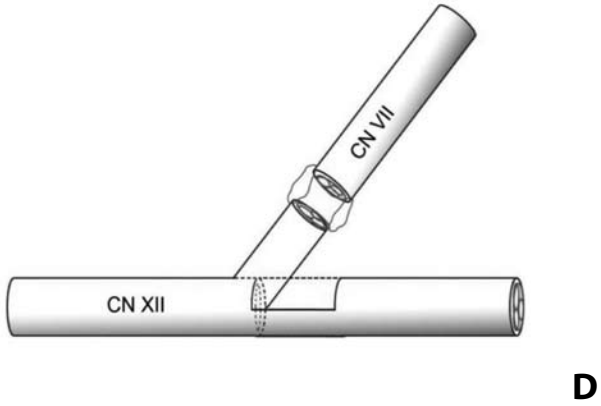


Figure 2. Intraoperative Findings
A. The dilated nerve trunk was followed as it passed through the stylomastoid foramen. Involved segments were resected and access to the intratemporal segment was done via transmastoid approach (top photo). **B.** A schematic diagram showing the tumor. **C.** Primary anastomosis of the facial and section of half of CN XII using interrupted epineural sutures with 9-0 monofilament nylon was performed (left). **D.** A schematic diagram of the procedure (right).

dips at 6000-8000Hz on the left.

The patient underwent total parotidectomy with facial nerve resection. A well-encapsulated, slightly-adherent, tan-brown mass was seen continuous with the facial nerve at the *pes anserinus*. The capsule contained yellowish, caseous material with light-brown, slightly-turbid fluid amounting to 3 ml. The mass, which seemed more like a dilated trunk of the facial nerve was followed as it passed through the stylomastoid foramen through the mastoid area with minimal erosion of the posterior canal wall. *(Figure 2A)* The nerve was resected via transmastoid approach from the *pes anserinus* up to the vertical segment before it entered the middle ear at the level of the tympanic membrane. *(Figure 2B)* The frozen section margins of resection of both proximal and distal ends of CN VII were void of tumor. A primary anastomosis of the facial nerve trunk and section of half of cranial nerve XII was performed using interrupted epineural sutures with 9-0 monofilament nylon. *(Figure 2C)* The use of a hemihypoglossal – facial nerve anastomosis instead of the classic technique prevented total denervation of the tongue to avoid post-operative hemi-tongue atrophy. Histopathologic findings were

consistent with schwannoma. Postoperative facial function was House-Brackmann Grade VI. Speech and hearing remained intact. *(Figure 3)*

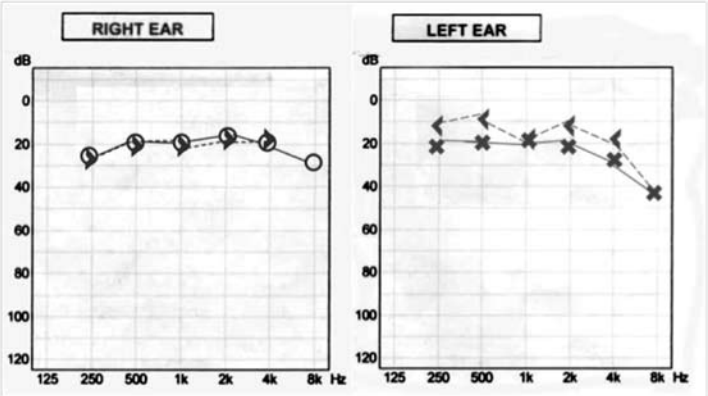


Figure 3. Post-operative Audiogram:

DISCUSSION

Parotid masses are commonly seen in the practice of head and neck surgery and almost 80% of them are benign. The presence of facial nerve paralysis may indicate compression and/or raise the suspicion of malignancy that warrants radiographic imaging and fine-needle aspiration biopsy. However, not all cases point to parotid tissue pathology.

Our patient initially presented with non-specific, gradual onset facial nerve paralysis. It was only after some time that an infra-auricular mass was noted warranting referral to an ENT specialist. We initially suspected a parotid malignancy most probably an adenoid cystic carcinoma due to its tendency for perineural invasion.

However, the peculiar presentation of paralysis preceding a parotid mass was a cause for concern. Was it really a parotid malignancy invading the facial nerve causing the facial paralysis? Did the neoplasm eventually spread via the nerve, through the stylomastoid foramen, hence the mastoid involvement? If so, the parotid mass had manifested quite late – two years after the facial paralysis.

Parotid malignancies such as adenoid cystic carcinoma typically present with skip lesions and would not involve the entire facial nerve on imaging.[2] Moreover, malignancies tend to contiguously spread to areas of least resistance. If it were a primary parotid lesion, a soft-tissue or parapharyngeal extension would have been more probable. The MRI findings in our patient of an intraparotid mass with temporal bone extension following the course of a well-delineated and enlarged facial nerve led to a seemingly more logical impression of a facial nerve schwannoma. If this were the case, then the lesion may have primarily occurred on the main trunk and followed a proximal course. A high-resolution temporal bone CT-scan together with a facial nerve MRI would have been beneficial for surgical planning but the financial resources for these were not available.

The surgical management of schwannomas is challenging. The aim of surgery is complete tumor excision with preservation of the facial and hearing nerve functions.[4] Many authors insist that it is beneficial to operate as early as possible but some prefer to delay surgery until the facial nerve function deteriorates to at least a HB Grade III. Resection is favored in nearly all cases and nerve decompression for a selected few. The final decision of whether to resect facial nerve schwannomas remains contentious because of the benign and slow growing nature of the tumor.[3]

Intraparotid facial nerve schwannomas are classified for management options. Type A (41.3%) and B neoplasms can be resected without apparent aesthetic and functional compromise. Type C neoplasms involve the main trunk of the nerve and type D involve both the main trunk and its main divisions. In type C and D tumors, patients with HB $\leq$ 3 warrant biopsy and physicians may adopt conservative management and follow-up after ruling-out malignancy. Patients with HB $\geq$ 4 require resection +/- reconstruction. Reconstruction options include end-to-end anastomosis, nerve grafting and facial – hypoglossal nerve anastomosis.[5]

Our case involved three issues: First was the timing and extent of surgery. Resection of the affected segment of the facial nerve was based on the extent of the lesion and preoperative facial function. The lesion was classified as type D and facial function was HB VI. The gradual-onset facial paralysis of chronic duration should have triggered a higher index of suspicion for a tumor, and additional diagnostic procedures such as imaging could have been done earlier.[6] Early identification and intervention would have provided a higher chance of recovering satisfactory facial nerve function.[1]

Second was the type of facial reanimation surgery. A temporalis muscle facial sling could have been appropriate.[7] However, the decision for hemihypoglossal – facial nerve anastomosis was a consensus of the surgical team lead by a neurootologist and a microvascular surgeon. *(Figure 2)* Casas[1] and Roland[8] report satisfactory outcomes for facial to hypoglossal nerve anastomosis even for patients with longstanding paralysis of more than two years. There is no advantage for any particular type of reconstruction, with the best recovery being HB grade III function.[5] The impact of a facial nerve disorder can be dramatic. Disabilities encountered include corneal exposure of the affected

eye, oral incompetence and articulation difficulties, and functional nasal obstruction. It would have been better if we had incorporated adjunctive reanimation techniques to augment the results of our facial to hemihypoglossal neurorraphy. Eyelid gold weights, tarsorraphy, endoscopic browlift and facial sling could have been offered as interim therapies in conjunction with the nerve anastomosis.[7]

During the six or more month - waiting period for nerve regeneration/ recovery, eye rehabilitation should be instituted to prevent excessive corneal exposure leading to ophthalmologic complications. The patient was advised to apply lubricating ophthalmic drops, occlusive dressing and cross taping of the eyelids to narrow the lid fissure and decrease drying. If incomplete lid closure persists, surgical reanimation can be performed.[9]

The third issue concerned the surgical approach and preservation of hearing. Pre-operatively, the patient did not complain of audiovestibular symptoms other than infrequent episodes of tinnitus. Most of the surgical approaches to the intratemporal segment of the facial nerve may result in a sensorineural or conductive hearing loss. It is therefore prudent to inform the patient of the risks and possible consequences of surgery. Fortunately, our patient's lesion was sufficiently exposed via a transmastoid approach. Because the ossicular and labyrinthine structures were virtually untouched, our patient's hearing and balance remained intact. *(Figure 3)*

A progressive hemifacial paralysis of chronic duration with or without the presence of an infra-auricular mass should raise the suspicion of a facial nerve tumor. Gadolinium-enhanced magnetic resonance imaging is valuable since intraparotid facial nerve schwannomas are mostly diagnosed intraoperatively when the neoplasm and the nerve are exposed and determined to be contiguous. The clinician should be aware that not all parotid masses are salivary gland in origin.

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