Case Report
online © ML Comm
Korean J Otorhinolaryngol-Head Neck Surg 2012;55:728-31 / pISSN 2092-5859 / eISSN 2092-6529
http://dx.doi.org/10.3342/kjorl-hns.2012.55.11.728

Giant Ancient Schwannoma of the Lateral Neck Presenting with Preoperative Horner's Syndrome

Seung Woo Kim, Dae Jin Sah, Seul Gi Kwak and Jun Yeon Hwang
Department of Otolaryngology-Head and Neck Surgery, VHS Medical Center, Seoul, Korea

술전 호너증후군을 동반한 측경부의 거대한 고대신경초종

김승우 · 사대진 · 곽슬기 · 황준연
중앙보훈병원 이비인후과

Received July 31, 2012
Revised September 24, 2012
Accepted October 4, 2012
Address for correspondence
Jun Yeon Hwang, MD
Department of Otolaryngology-Head and Neck Surgery,
VHS Medical Center,
53 Jinhwangdo-ro 61-gil,
Gangdong-gu,
Seoul 134-791, Korea
Tel +82-2-2225-1880
Fax +82-2-2225-1385
E-mail djs0@hotmail.com

While ancient schwannoma (AS) from the cervical sympathetic chain is very rare. Preoperative Horner's syndrome resulting from cervical sympathetic chain schwannoma (CSCS) is extremely rare. A 58-year-old woman visited our clinic with a huge lateral neck mass that had been present for thirteen years. Ptosis and enophthalmos were observed on the left eye. After radiologic evaluation, we assessed the CSCS by performing surgical excision. Finally, pathologic examination revealed it to be AS. There has been no recurrence for 15 months after discharge, however Horner's syndrome has become more prominent. We report this unique case of ancient schwannoma with literature review. Korean J Otorhinolaryngol-Head Neck Surg 2012;55:728-31

Key Words Horner syndrome · Preoperative period · Schwannoma · Sympathetic nervous system.

Introduction

Schwannoma is a benign neoplasm of nerve sheath origin except olfactory and optic nerves. The incidence of the head and neck lesion is 20−45%, and it may also arise in the vagus, hypoglossal, lingual, cervical sympathetic chain and brachial plexus in the order of frequency.[1] In earlier reports, five variants of schwannomas have been described; common, plexiform, cellular, epithelioid, and ancient type.[2] The ancient schwannoma (AS) is characterized with degenerative changes, hyperchromatism and marked nuclear atypia, which differentiates itself from the typical type.

In cervical sympathetic chain schwannoma (CSCS), the most common post-operative complication is the Horner's syndrome. However, preoperative Horner's syndrome in CSCS is extremely rare. The following is a clinical report about a giant AS from cervical sympathetic chain (CSC), accompanied with preoperative Horner's syndrome. To the best of our knowledge, such case has not been reported earlier.

Case

A 58-year-old woman visited our clinic, presenting with a huge left lateral neck mass that had been present for thirteen years. She complained of pressure sensation on the left neck and slight dysphagia. On physical examination, the characteristics of mass were 8×7 cm sized, rubbery hard, slightly movable and non-tender. In ophthalmic examination, intra-ocular pressure on the left eye was 15 mm Hg, with the left eye showing enophthalmos of 2 mm in Hertel exophthalomometry. The patient showed miosis, ptosis and enophthalmos on the left eye and left double-eyelid seemed to be swollen compared to the right eye (Fig. 1). There was no another neurologic deficit.

Contrast-enhanced axial CT scans revealed a well-margin-

ed heterogeneously enhanced huge mass in the left carotid space. The common, internal and external carotid arteries were displaced to the anterior while the internal jugular vein was collapsed (Fig. 2A). Findings of MRI were equal to those of CT scan (Fig. 2B and C). There was no feeding artery in four vessels trans-femoral carotid angiography. Fine needle aspiration cytology showed no specific findings. We initially diagnosed with schwannoma from the CSC or vagus nerve, malignant peripheral nerve sheath tumor and sarcoma (least likely).

Under general anesthesia, we performed the surgical excision. The carotid sheath was exposed. The vagus nerve ran over the mass and was carefully dissected. Since the mass originated from the CSC, it was impossible to dissect the mass without nerve sacrification (Fig. 3).

The Horner's syndrome was more prominent in the postoperative period. There has been no recurrence for 15 months. The patient still shows similar preoperative ocular findings without complaining of symptoms. The surgical specimen was an encapsulated ovoid, pinkish gray to pale yellow and slightly firm, measuring 8.0×5.5×5.0 cm (Fig. 4A). The cut surface was myxoid, partly yellow, hemorrhagic and cystic structures (Fig. 4B). The microscopic findings were compatible with ancient schwannoma (Fig. 5).

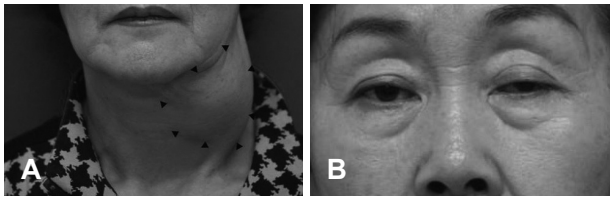

Fig. 1. The external gross photo shows huge bulging contour in the right lateral neck (black arrowheads)(A). The ptosis and enopthalmos is observed in the left eye (B).

Discussion

Ackerman and Taylor[3] suggested the terminology ''ancient'' schwannoma to connote the long duration and degenerative changes to the appearance of these lesions. This lesion is characterized with diffuse hypocellularity, focal infiltration of hyaline and fatty degenerations. Due to poor cellularity especially in the cystic degenerated lesions, AS may be difficult to diagnose from cytology.[4] The nuclear atypia and hyperchromatism are not a sign of malignancy but a regressive phenomenon. According to the size and duration of tumor, degeneration and atypia becomes more severe. AS are distinguished from malignant schwannoma by the absence of mitosis and the invasive pattern. Because the S-100 protein is rich in the supporting cell in the peripheral nerve, the Antoni A areas show strong positive reaction in this stain.

The head and neck, thorax, retroperitoneum, pelvis and extremities are the predilection sites of AS.[5] AS mainly occurs in the middle and elderly age, with no sexual difference. The asymptomatic mass is the most common clinical sign. According to the growing of mass, various symptoms such as cough, dyspnea, hoarseness, dysphagia, numbness, pain and paresthesia may be presented.

Preoperative knowledge of tumor origin is useful for surgical planning, prediction of postoperative complications and surgical outcomes. Before surgery, the CSCS must be differentially diagnosed with carotid body tumor (CBT) and paraganglioma. Usually CBT requires embolization, whereas not in CSCS. Generally, the MR image of CSCS shows high signal intensity on T2 weighted images, with relatively low signal intensity on T1 weighted images.[5] As the CSCS grow and expand, it has a tendency to displace the common, internal and external carotid arteries, anteriorly. The internal jugular vein

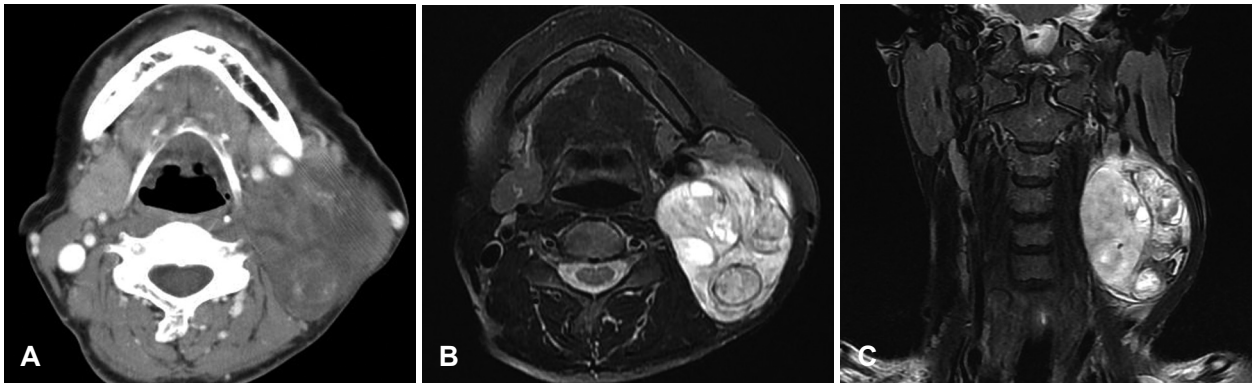

Fig. 2. Contrast-enhanced axial CT scan shows the well-margined heterogeneously enhanced mass with anterior displacement of internal carotid and external carotid arteries (A). T2 weighted MR image shows heterogenous high signal intensity mass with the same vascular findings of CT (B and C).

is therefore displaced anterolaterally. The separation between the internal jugular vein laterally and carotid arteries medially are not observed. The Lyre sign, which is spreading between internal carotid artery and external carotid artery in angiography, is observed in both CBT and CSCS.[6] The differential diagnosis with AS from CSC include paraganglioma, chemodectoma, parotid gland neoplasm, branchial cyst, ICA aneurysm, lipoma, meningioma, rhabdomyoma and thyroid carcinoma.[6]

The preoperative Horner's syndrome is not diagnostic as CSCS since it is possible that the effects may arise due to compression from mass of other origin. Patients with Horner's syndrome show miosis, ptosis, enophthalmos and facial anhydrosis. Ptosis results from paralysis of Müller's muscle, but usually the patient does not complain of any symptoms. Since the CSC runs in a relatively loose fascial compartment, the preoperative Horner's syndrome in CSCS has only been reported in six cases,[7] and AS from CSC has not been reported earlier. Horner's syndrome has been known to usually occur in the postoperative period.

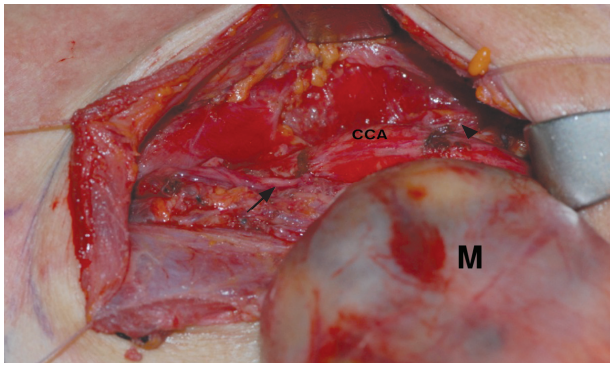

Fig. 3. Intraoperative findings. The mass is nearly extracted from the cervical sympathetic nerve. The vagus nerve is located over the mass. The internal jugular vein is not seen due to severe collapse. M: mass, CCA: common carotid artery, arrow: vagus nerve, arrowhead: cervical sympathetic nerve.

The treatment of choice in CSCS is complete excision of the mass while trying to keep the nerve intact. Generally, the nerve was excised with a superior and inferior cuff, because of its dense attachment to the mass.[7] Also, nerve graft is not usually performed. The vagus nerve runs superior to the mass, where meticulous and careful dissection is essential. Especially in large tumors, the nerve is located more superficial and also unusually displaced. Therefore, the surgeon must be very careful, so the vagus nerve is preserved which may be mistaken for ansa cervicalis. The recurrence and malignant change of AS are very rare, but some case have been reported.[8]

We have described the unique case of giant ancient schwannoma of the lateral neck presenting with preoperative Horner's syndrome with literature review. To the best of our knowled-

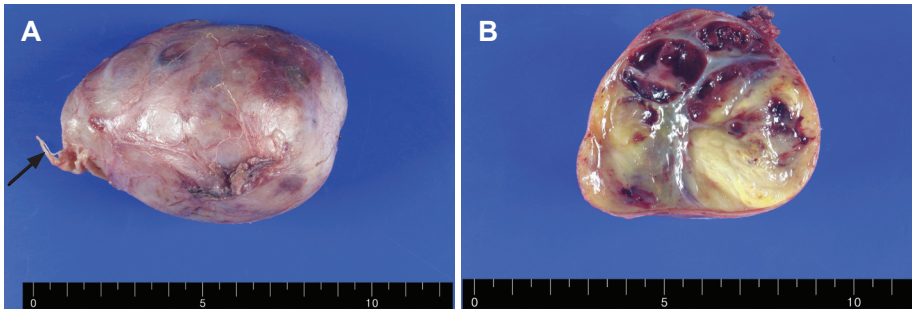

Fig. 4. The gross findings show an encapsulated ovoid, pinkish gray to pale yellow and slightly firm mass, measuring 8.0×5.5×5.0 cm. It is also observed the resected nerve (arrow)(A). The cut surface shows a myxoid, partly yellow, hemorrhagic and cystic structures (B).

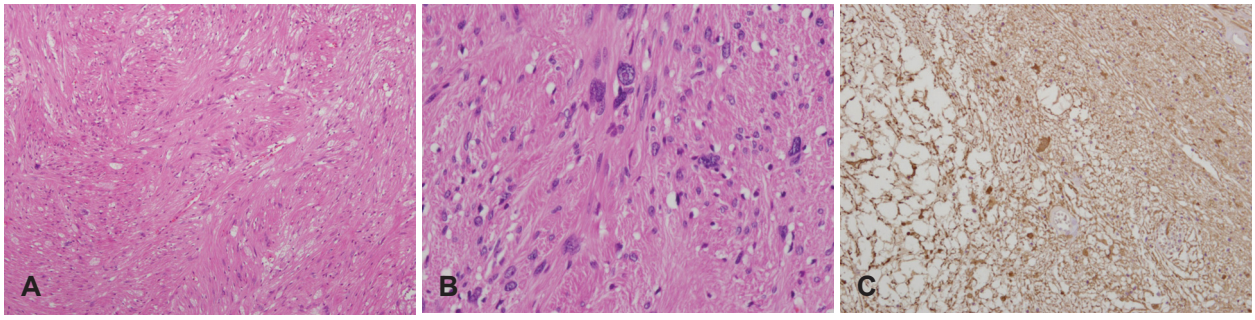

Fig. 5. The microscopic findings. It shows the hypercellular area with pallisading nuclei (A). Ancient schwannoma displays degenerative changes such as hyperchromatism and marked nuclear atypia. This shows hyalinization and cellular atypia with hyperchromatic bizarre nuclei, but no mitotic figure (B). The immunohistochemical stain shows strong and diffuse positivity for S-100 protein (Hematoxylin and Eosin stain, original magnification: A; ×100, B; ×400, immunohistochemical stain: C; ×200)(C).

ge, such disease entity has not been reported earlier.

REFERENCES

1) Guerrissi JO. Solitary benign schwannomas in major nerve systems of the head and neck. J Craniofac Surg 2009;20(3):957-61.
2) Weiss SW, Goldblum JR. Parotid schwannoma. In: Enzinger FM, Weiss SW, editors. Enzinger and Weiss's Soft Tissue Tumors. 4th ed. St Louis: Mosby;2001. p.1146-67.
3) Ackerman LV, Taylor FH. Neurogenous tumors within the thorax; a clinicopathological evaluation of forty-eight cases. Cancer 1951;4(4): 669-91.
4) Domanski HA, Akerman M, Engellau J, Gustafson P, Mertens F, Rydholm A. Fine-needle aspiration of neurilemoma (schwannoma). A clinicocytopathologic study of 116 patients. Diagn Cytopathol 2006;34(6):403-12.
5) Isobe K, Shimizu T, Akahane T, Kato H. Imaging of ancient schwannoma. AJR Am J Roentgenol 2004;183(2):331-6.
6) Casserly P, Kiely P, Fenton JE. Cervical sympathetic chain schwannoma masquerading as a carotid body tumour with a postoperative complication of first-bite syndrome. Eur Arch Otorhinolaryngol 2009;266(10):1659-62.
7) Wax MK, Shiley SG, Robinson JL, Weissman JL. Cervical sympathetic chain schwannoma. Laryngoscope 2004;114(12):2210-3.
8) Mikami Y, Hidaka T, Akisada T, Takemoto T, Irei I, Manabe T. Malignant peripheral nerve sheath tumor arising in benign ancient schwannoma: a case report with an immunohistochemical study. Pathol Int 2000;50(2):156-61.