TÜBİTAK

CASE REPORT

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Horner's Syndrome Post-Excision of a Huge Cervical Sympathetic Chain Schwannoma

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Abstract: Schwannoma of the cervical sympathetic chain is a rare nerve tumor. These lesions typically present as an asymptomatic neck mass and are easily mistaken for a carotid body tumor during the initial work-up. In this report, a rarely seen huge cervical sympathetic chain schwannoma case, who experienced partial Horner's syndrome postoperatively, is presented. We report a case of schwannoma on the cervical sympathetic chain, which to our knowledge is the largest reported in the current literature.

Key Words: Schwannoma, Horner's syndrome, cervical sympathetic chain

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Büyük Boyutlu Servikal Sempatik Zincir Schwannomasının Eksizyonu Sonrasında Gelişen Horner Sendromu

Özet: Servikal sempatik zincir schwannomaları nadir görülen bir tümör olup tipik olarak boyunda asemptomatik bir kitle şeklinde ortaya çıkar. İlk muayene sırasında kolaylıkla karotis cisimciği tümörü ile karışabilir. Bu vaka takdiminde; nadir görülen büyük boyutlu postoperatif parsiyel Horner sendromuna yol açan servikal sempatik zincir schwannoması sunuldu. Bu sebeple, şimdiye kadar literatürde sunulan en büyük boyutlu servikal sempatik zincir kaynaklı schwannoma vakasını sunmayı uygun bulduk.

Anahtar Sözcükler: Schwannoma, Horner Sendromu, Servikal Sempatik zincir

Introduction

Schwannomas are painless, benign, and slow-growing solitary tumors. Verocay (1) was the first to describe these lesions in 1908. Microscopically, the tumor consists of two characteristic patterns as described in 1920 by Antoni. Schwannomas can arise from any cranial (IX, X, XI, XII), sympathetic, or peripheral nerve and most commonly occur in the neck. These tumors are well encapsulated, and nerve fibers often splay out on the surface but never penetrate the capsule. They have a rare malignant degeneration (2). Whereas cervical schwannomas are uncommon, those arising from the cervical sympathetic chain (CSC) are extremely rare, with less than 60 cases reported in the English literature (3-36). Diagnosis of the nerve that is the origin of the tumor and differentiation among the various histologic processes that occur in the parapharyngeal space are important for preoperative planning. They have a similar incidence in both sexes. Schwannomas, by their nature, rarely compress or destroy the nerve. Because the CSC runs in a relatively loose facial compartment, compression, such as seen with acoustic neuromas or facial nerve schwannomas, is exceedingly rare. In this respect, patients commonly present asymptomatic neck mass, but this syndrome is frequently observed after the tumor of the CSC is removed (4-7). Herein, we present a schwannoma case of the CSC, which we think is the largest reported in the current literature thus far.

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Case Report

A 74-year-old woman initially underwent an incisional biopsy of the non-tender right-sided neck mass by a general surgeon. The pathologic report was necrotizing granulomatosis lymphadenitis. The patient then received anti-tuberculosis treatment for four months. Because the patient observed no benefit from this treatment and the mass had been increasing in size for 12 months, she was referred to our otolaryngology service with an asymptomatic right-sided neck mass. Measuring approximately 7.5 x 5.5 cm, this mass lay medial to all of the sternocleidomastoid (SCM) muscle, at the level of the carotid bifurcation, and it was mobile, non-tender, and nonpulsatile, with no associated bruit. Laryngeal endoscopy revealed normal movement of her vocal cords and no ocular sign was present. Chest radiograph and PPD were negative. The remainder of the head and neck examination yielded normal findings. Fine needle aspiration, while conclusive in many cases of neck masses, is much less valuable for the compact neural tumor, and failed to reveal the diagnosis in our patient. Diagnostic studies included computed tomography (CT) and magnetic resonance imaging (MRI), which confirmed a well-circumscribed solid mass that was hypointense on T1-weighted imaging, 7.5 x 5.5 x 4.5 cm in size, showing necrotic component centrally, displaying extension from the ramus of the mandible to the thyroid cartilage on the right side of the neck, and displacing the SCM muscle and external carotid artery posterolaterally and the internal carotid artery and internal jugular vein anteromedially. Also noted was splaying of the internal and external carotid arteries (Figure 1a,b). The patient did not accept angiography due to her old age.

In the preoperative counseling, we discussed with the patient neurologic problems such as vocal cord palsy and Horner's syndrome. She accepted all the risks of the operation. The patient was taken to the operating room. Since the mass was large enough to influence the right side of the neck, it was exposed through a vertical incision along the anterior border of the SCM muscle. The mass was located deep into the SCM muscle, which was thinned and displaced posteriorly, inferior to the styloid process, superficial to the prevertebral muscle, and posterior to the internal carotid artery and internal jugular vein. External carotid artery was displaced posterolaterally. In the operation, hypoglossal nerve was displaced highly superiorly and the vagal nerve was identified and uninvolved. The tumor was encapsulated and separated from the surrounding vascular structures, but appeared to arise from the CSC. The tumor could not be resected without sacrificing a portion of the chain (Figure 2). A section of the chain was resected. Schwannoma was confirmed by pathology (Figure 3). The patient did well postoperatively and the drain was removed on the second

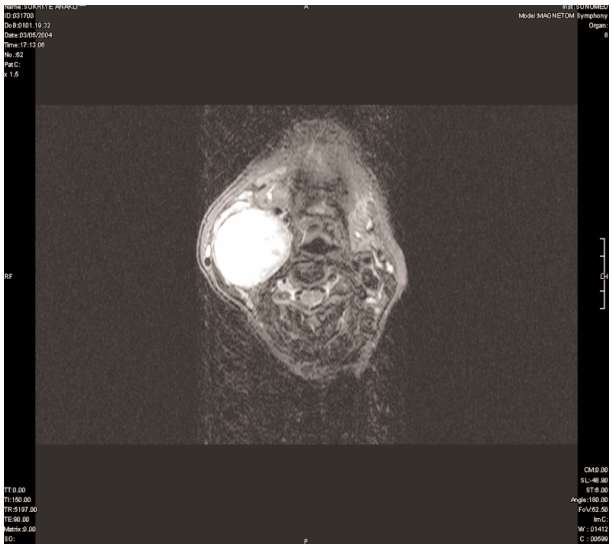

Figure1a. Axial magnetic resonance image of right cervical sympathetic chain schwannoma.

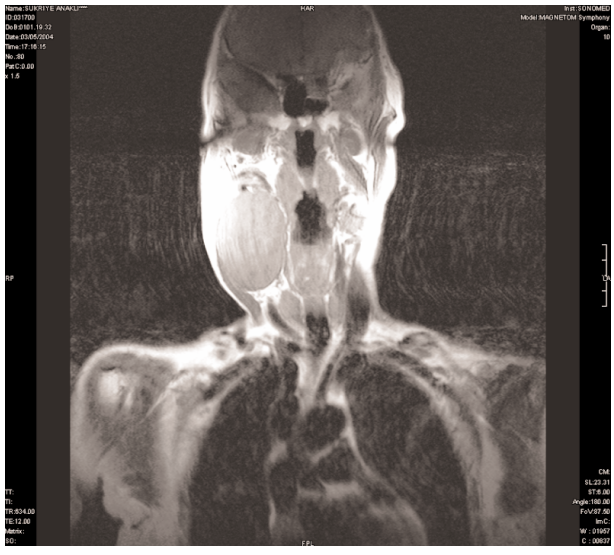

Figure1b. Coronal magnetic resonance image of right cervical sympathetic chain schwannoma.

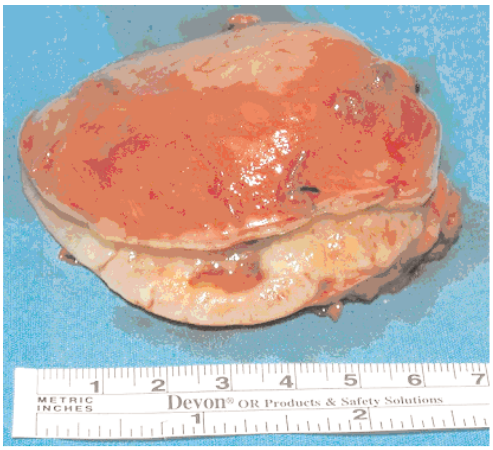

Figure 2. Surgical specimen of the huge cervical sympathetic chain schwannoma.

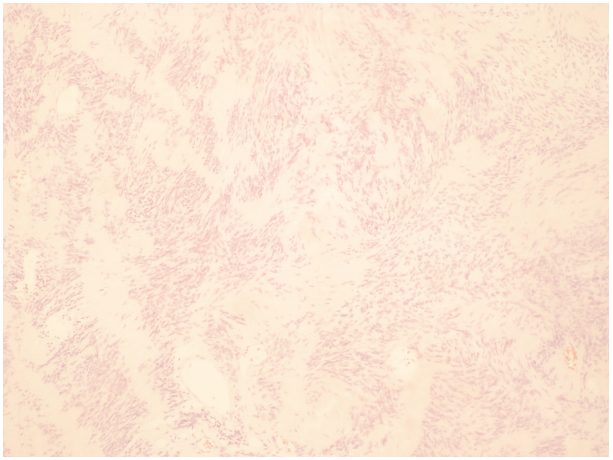

Figure 3. Antoni A configuration with Verocay bodies (H & E x100).

postoperative day. Pathology revealed a typical schwannoma. Other than a right miosis and slight ptosis (partial Horner's syndrome), the patient recovered well. After a 22-month follow-up, partial Horner's syndrome persisted. She did not have recurrence, and she had no complaints about anhidrosis or vasodilatation of the ipsilateral face; in fact, such clinical findings were absent (Figure 4).

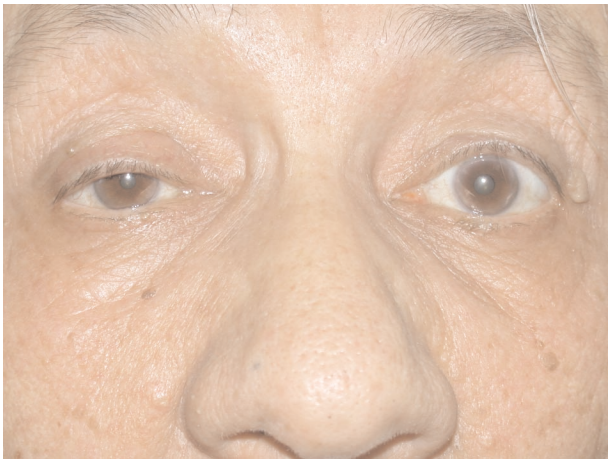

Figure 4. A view of patient with right miosis, ptosis, and enophthalmos after 22-month follow-up.

Discussion

Schwannomas are uncommon tumors that arise from any peripheral, cranial, or autonomic nerve. They have a wide distribution, but approximately 25 to 45% occur in the head and neck region; thus, care is usually in the domain of the otorhinolaryngologist. The nerves most commonly involved in post-styloid parapharyngeal space are the vagus and the CSC. The frequency is nearly equal, with less than 70 CSC schwannomas having been reported in the English literature (Table 1). Schwannomas of the CSC are reported to occur between the ages of 5 and 77 years. The male to female ratio has been reported as 1:1. No other etiologic factors or predispositions have been delineated from the small series or the case reports that have been published (3-36). Our patient's age was between the limits previously published.

The majority of schwannomas are asymptomatic at the time of presentation. Some patients may present nonspecific symptoms. Typically, further direct questioning of the patient reveals the presence of an indistinct neck mass that has been present for approximately six months. This painless neck mass brings the patient to the physician. However, if pain and neurological deficit are present, malignancy must also be considered. Our patient confirms this typical presentation. The incidence of Horner's syndrome before excision has been recorded in only 10 previously reported cases of cervical schwannomas (6,8,9,10,11), but in most cases that had some degree of Horner's syndrome developing in the postoperative period, the symptoms decreased over time. Although our patient was observed 22 months postoperatively, her partial syndrome did not

Table 1. Case reports of CSCS published in the English literature.

Authors	Publication date-number of case(s)	Age/sex	Size (cm)	Presentation (painless mass +)	Preoperative diagnosis
Politi[11]	2005-1	46/F	7 x 5 x 5	Horner syndrome	?
Uzun[22]	2005-1	25/F	4 x 5		CSCS
Wa x[4]	2004-4	68/M	3	partial Horner	CSCS
Wang[14]	2004-1	26	5 x 5		CBT
Benzoni[26]	2003-1	?	?		paraganglioma
Kara[25]	2002-1	20	2 x 2		?
Aygenc[21]	2002-1	?	?		?
Rosner[16]	2001-1	47/M	2 x 4		CBT OR VS
Hood[20]	2000-4	60/M	4 x 5	partial Horner	CSCS, CBT OR VS
Colreavy[15]	2000-4	19-77	5.5cm	partial Horner	CBT
Panneton[18]	2000-2	33M/49M	4 x 3.5cm and 2 x 2.5cm		CBT/CBT
Souza[8]	2000-1	35/M	3 x 4		CBT
Valentino[7]	1998-5	?	?		?
Sheridan[19]	1997-1	?	?		?
Ganesan[9]	1997-5	?	?	partial Horner	
Furukawa[23]	1996-9	?	?		?
Takimoto[17]	1989-1	5/M	?	partial Horner	?
Myssiorek[3]	1988-2	56F/59M	4.5 x 4.5cm and 3 x 3cm		CBT OR CSCS/CBT
Sharaki[27]	1982-2	?	?		?
Clairmont[28]	1978-1	62/F	2		CBT
Clifton2[4]	1977-2	53F/45F	4cm and 6cm		THYROID/CBT
Brandenburg[29]	1972-1	21/F	4cm		BRONCHIAL CYST
Daly[30]	1963-2	36M/66M	4.5cm and 2.5cm		CSCS/CBT OR CSCS
Kragh[31]	1960-7	?	?		?
Brandt[32]	1953-1	24/F	?		?
Rogers[33]	1953-1	42/F	?		ABERRANT THYROID
Cullen[12]	1952-2	40F/42M	6cm and 5cm		CBT/CBT
Callum[10]	1950-1	31/F	?		LYMPH NODE
Mayo[34]	1934-1	38/F	5		?
Amano[35]	1934-2	?	?		?
Gibberd[36]	1924-2	37M/32M	?		?/CSCS
our case		72/F	7.5 x 5.5		CSCS

CSCS: Cervical sympathetic chain schwannoma. CBT: Carotid body tumor. VS: Vagal schwannoma

regress over this period. Currently, this patient is asymptomatic and unaware of her syndrome.

The differential diagnosis for patients presenting a lateral neck mass is quite varied. The most demanding aspect of treatment of these lesions is distinguishing the benign cervical sympathetic chain schwannoma (CSCS) from other pathologies in the parapharyngeal space. The differential diagnosis of a parapharyngeal space mass is based on the division of the space into pre- and post-styloid compartments. Masses arising in the pre-styloid compartment may include deep lobe parotid tumors, lipomas, lymphadenopathy, and rare neurogenic tumors. In the post-styloid compartment, possible causes include carotid artery aneurysms, neurogenic tumors involving cranial nerves IX through XII, as well as the sympathetic chain and paragangliomas arising from the vagus nerve or carotid body (10,12,13). Fine needle aspiration, while conclusive in many cases of neck masses (14,15), is much less valuable for the compact neural tumor. It was performed but was not diagnostic in our patient. Because

the pathology report revealed the necrotizing granulomatosis lymphadenitis, the patient was treated for four months with antituberculosis drugs.

Imaging studies play a central role in diagnosis of these tumors. CT has until recently been the standard investigation, but has been superseded by MRI. With MRI, one can rule out other tumors that can present in a similar manner and determine the relation of the tumor mass to the carotid artery system. The most important goal is to distinguish between a vagal or CSCS and carotid body tumor (CBT). Splaying of the carotid bifurcation with hypervascularity usually indicates carotid body tumor, which, without hypervascularity, may suggest schwannoma. However, most of the CSCS with splaying of the carotid bifurcation and lack of rich vascularity are still thought preoperatively to be carotid body tumors (3,8,9,16,17). Furthermore, MRI of a CSCS can show a salt-pepper appearance similar to that of a CBT because of some flow voiding and an area of hemorrhage or cystic degeneration within the tumor. MR angiography is useful to essentially differentiate CBT and CSCS by the appearance of tumor vascularity. However, conventional angiography is still necessary, because some schwannomas have significant vascularity within the tumor or around the capsule (18,19,20,21,22). As for vagal schwannoma, there seem to be no reports of a vagal schwannoma with splaying of the carotid bifurcation in the literature. Because the vagus nerve is located between the internal carotid artery and the internal jugular vein, it always runs laterally to the internal carotid artery above the bifurcation (14). In their experience with nine cervical nerve sheath tumors, Furukawa et al. (23) found that vagal nerve tumors typically resulted in an increased distance between the internal carotid or common carotid artery (anteromedially) and the internal jugular vein (posterolaterally), but they could not separate the internal and external carotid arteries. Such separation is not noted in CSCS.

A review of the English literature to date (4,3,36) determined two cases of CSCS 6 cm in size (12,24). One of the cases, presented by Cullen and Monro, was 40 years old and the other, presented by Clifton, was 45 years old. We can say that the CSCS in our case, 7.5 x 5.5 x 4.5 cm in size, is the largest one reported thus far. The larger size in our case can be partially explained by our patient's older age of 74 years.

Surgical resection is recommended for CSCS. Excision of the tumor gives excellent results with no local recurrence. Resection of the CSCS is well tolerated, with a transient or permanent Horner's syndrome being the expected complication (7,18). Partial Horner's syndrome (ptosis and miosis without anhidrosis) was encountered on the right side of the face in the postoperative period in our patient. This meant that only the fibers branching to the internal carotid artery of the CSC were injured during surgery (22). Although mention is made in the literature of performing a neural graft with reanastomosis, it was not performed in our patient. To date, there has been no data that shows this procedure to be efficacious in these types of cases (4).

Consequently, schwannomas that originate from the CSC are rarely encountered by head and neck surgeons. They usually present as asymptomatic neck mass that, with proper investigation, can be diagnosed with relative accuracy after preoperative imaging. Surgical resection almost inevitably leaves the patient with a partial Horner's syndrome, which is relatively asymptomatic and which should be discussed with the patient in the preoperative counseling.

References

1. Verocay J. Multiple geschwulste als systemerkrank ung am nervosen. In: Chiari FF. Wien and Leipzig: W. Braunmiller; 1908. pp. 378-415.
2. Bailet JW, Abemayor E, Andrews JC, Rowland JP, Fu YS, Dawson DE. Malignant nerve sheath tumors of the head and neck: a combined experience from two university hospitals. Laryngoscope 1991; 101: 1044-1049.
3. Myssiorek DJ, Silver CE, Valdes ME. Schwannoma of the cervical sympathetic chain. J Laryngol Otol 1988; 102: 962-965.
4. Wax MK, Shiley SG, Robinson JL, Weissman JL. Cervical sympathetic chain Schwannoma. Laryngoscope 2004; 114: 2210-2213.
5. Mikaelian DO, Holmes WF, Simonian SK. Parapharyngeal schwannomas. Otolaryngol Head Neck Surg 1981; 89: 77-81.

6. Das Gupta TK, Brassfield RD, Strong EW, Hadju SI. Benign solitary Schwannomas (neurilemomas). Cancer 1969; 24: 355-366.

7. Valentino J, Boggess MA, Ellis JL, Hester TO, Jones RO. Expected neurologic outcomes for surgical treatment of cervical neurilemomas. Laryngoscope 1998; 108: 1009-1013.

8. Souza JW, Williams JT, Dalton ML, Solis MM. Schwannoma of the cervical sympathetic chain: It's not a carotid body tumor. Am Surg 2000; 66: 52-55.

9. Ganesan S, Harar RP, Owen RA, Dawkins RS, Prior AJ. Horner's syndrome: a rare presentation of cervical sympathetic chain schwannoma. J Laryngol Otol 1997 May; 111(5): 493-495.

10. Callum EN. Neurinoma of cervical sympathetic chain. Br J Surg 1950; 37: l 17.

11. Politi M, Toro C, Cian R, Costa F, Robiony M. Horner's syndrome due to a large schwannoma of the cervical sympathetic chain: report of a case. J Oral Maxillofac Surg 2005; 63: 707-710.

12. Cullen TH, Monro RS. Cervical neurofibroma in the differential diagnosis of carotid body tumor. Br J Surg 1952; 39: 454-457.

13. Rogers L. Neurinomas of the cervical sympathetic system with report of a case. Br J Surg 1953; 40: 579-580.

14. Wang CP, Hsiao JK, Ko JY. Splaying of the carotid bifurcation caused by a cervical sympathetic chain schwannoma. Ann Otol Rhinol Laryngol 2004; 113(9): 696-699.

15. Colreavy MP, Lacy PD, Hughes J, Bouchier-Hayes D, Brennan P, O'Dwyer AJ et al. Head and neck schwannomas--a 10 year review. J Laryngol Otol 2000; 114(2): 119-124.

16. Rosner M, Fisher W, Mulligan L. Cervical sympathetic schwannoma: case report. Neurosurgery 2001; 49(6): 1452-1454.

17. Takimoto T, Katoh H, Umeda R. Parapharyngeal schwannoma of the cervical sympathetic chain in a child. Int J Pediatr Otorhinolaryngol 1989; 18: 53-58.

18. Panneton JM, Rusnak BW. Cervical sympathetic chain schwannomas masquerading as carotid body tumors. Ann Vasc Surg 2000; 14(5): 519-524.

19. Sheridan MF, Yim DWS. Cervical sympathetic schwannoma: a case report and review of the English literature. Otolaryngol Head Neck Surg 1997; 117: 206-210.

20. Hood RJ, Reibel JF, Jensen ME, Levine PA. Schwannoma of the cervical sympathetic chain. The Virginia experience. Ann Otol Rhinol Laryngol 2000; 109(1): 48-51.

21. Aygenc E, Selcuk A, Ozdem C. Hypervascular parapharyngeal schwannoma: an unusual case. Auris Nasus Larynx 2002; 29(2): 215-217.

22. Uzun L, Ugur MB, Ozdemir H. Cervical sympathetic chain schwannoma mimicking a carotid body tumor: a case report. Tumori 2005 Jan-Feb; 91(1): 84-86.

23. Furukawa M, Furukawa MK, Katoh K, Tsukuda M. Differentiation between schwannoma of the vagus nerve and schwannoma of the cervical sympathetic chain by imaging diagnosis. Laryngoscope 1996 Dec; 106(12 Pt 1): 1548-1552.

24. Clifton MA. Sympathetic neurilemmoma: an uncommon cause of solitary cervical swelling. J R Coll Surg Edinb 1977; 22: 351-354.

25. Kara CO, Topuz B. Horner's syndrome after excision of cervical sympathetic chain schwannoma. Otolaryngol Head Neck Surg 2002; 127(1): 127-128.

26. Benzoni E, Cojutti A, Intini S, Uzzau A, Bresadola F. Schwannoma of the sympathetic cervical chain as a lateral neck mass. Tumori 2003; 89: 211-212.

27. Sharaki MM, Talaat M, Hamam SM. Schwannoma of the neck. Clin Otolaryngol Allied Sci 1982; 7: 245-251.

28. Clairmont AA, Conley JJ. Schwannoma of the cervical sympathetic nerve. Ear Nose Throat J 1978; 57: 336-339.

29. Brandenburg JH. Symposium on malignancy. IV. Neuro-genic minors of the parapharyngeal space. Laryngoscope 1972; 82: 1292-1305.

30. Daly JF, Roesler HK. Neurilemmomas of the cervical sympa-thetic chain. Arch Ololaryngol 1963; 77: 42-47.

31. Kragh LV, Soule EH, Masson JK. Benign and malignant neurilemmomas of the head and neck. Surg Gynecol Obstet 1960; 111: 211-218.

32. Brandt U. Neurinome du sympalhique cervical. Acta Otolaryngol 1953; 43: 503-505.

33. Rogers L. Neurinomas of the cervical sympathetic system with report of a case. Br J Surg 1953; 40: 579-580.

34. Mayo CW. Cervical neurofibroma. Surg Gynecol Obstet J 1934; 59: 671-677.

35. Amano M. Ueber zwei Faelle von Neurinom des Halssym-pathicus. Zentralbl Chir 1934; 61: 7S.

36. Gibberd GF. Two cases of neurofibroma of the cervical sympathetic. Guys Hosp Rep 1924; 74: 367-368.