CASE REPORT

Cervical sympathetic chain schwannoma

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Summary
Introduction: Cervical schwannoma is a benign peripheral nerve tumor specifically developing from Schwann cells. Cervical sympathetic chain schwannoma is rare. Following a case report, the authors describe its specific radiological and histological characteristics. Treatment is surgical.
Clinical case: A 56-year-old woman consulted for an isolated left lateral cervical mass of several years’ standing, but with recently associated pharyngeal discomfort. Cervical CT revealed a vascularized retrostyloid mass with venous-time enhancement, inducing anterior displacement of the jugulo-carotid axis. The tumor could not be identified on fine-needle aspiration cytology, and surgical resection was performed by cervicotomy. Surgical exploration found a tumor developing from the cervical sympathetic nerve, posterior to the jugular vein and carotid sheath. Histopathologic examination diagnosed schwannoma. Postoperative outcome featured Horner’s syndrome.
Conclusion: Cervical sympathetic chain schwannoma is a rare benign tumor, to be suspected in the presence of an isolated lateral cervical mass. Preoperative CT is mandatory to guide diagnosis; treatment is surgical, to confirm histologic diagnosis. Postoperative Horner’s syndrome often confirms cervical sympathetic chain involvement.

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Introduction

Schwannoma is a benign mesenchymatous tumor developing exclusively from the cells of the Schwann sheath of peripheral nervous system nerve fibers. In 25% of cases, the location is cervical, mainly involving the pneumogastric nerve (X) [1]. Other locations, and notably cervical sympathetic chain schwannoma (CSCS) are rarer. The attitude of choice is surgical, but the option of abstention should always be discussed with the patient in the light of the functional risk of definitive sequelae inherent in the surgical procedure.

Based on one recent case of CSCS, we review the clinical, radiological and therapeutic aspects of this rare anatomo-clinical entity.
Case study

A 56-year-old woman with no particular medical or surgical history presented with a left cervical mass, known to have been present for 2 years but which had recently increased in size.

Physical examination found an isolated superior left lateral cervical mass which was oblong, firm, insensitive and mobile. There was no associated dysphagia, dysphonia or neurologic deficit.

Cervical CT found a rounded, well-contoured, unilocular mass occupying the left-side sectors IIa and III, of heterogeneous tissue density in the center and pushing the jugulo-carotid axis forward and outward. Contrast injection showed late density enhancement of the peripheral part of the mass while the center remained isodense (Fig. 1).

Fine-needle aspiration cytology proved non-contributive. Exploratory cervicotomy was decided on in a multidisciplinary coordination meeting, without other complementary examinations as the clinical presentation and CT findings were considered sufficiently diagnostic and there were no risk factors related to alcohol or smoking.

Surgical exploration found a homogeneous 4 × 3 cm mass causing anterior displacement of the internal jugular vein and Xth nerve and medial displacement of the carotid vessels (Fig. 2). It appeared to develop from the deep branches of the cervical plexus; extemporaneous diagnosis pointed to benign schwannoma (Fig. 3). In the immediate postoperative course, Horner’s syndrome developed, pointing diagnosis toward schwannoma of the cervical sympathetic chain.

Discussion

Cervical schwannoma was first reported by Ritter in 1899. Twenty-five percent to 45% of locations are head and neck [2,3], most frequently intracranial (vestibular nerve) [4]; cervical locations are less common. The tumor may develop in the parapharyngeal space from the last four cranial nerves and cervical sympathetic chain, with Xth nerve most frequently involved [5–8].

CSCS is generally of adult onset (20–70 years of age), with a balanced sex ratio [6]. The clinical presentation is non-pathognomonic. The aspect is usually of an isolated

Figure 1  Contrast-enhanced head and neck CT, C4 slice: cervical sympathetic chain schwannoma. 1: styloid; 2: external carotid; 3: internal carotid; 4: internal jugular vein.

Figure 2  Left lateral cervicotomy, cervical sympathetic chain schwannoma. 1: common carotid artery; 2: external carotid artery; 3: pneumogastric nerve; 4: internal jugular vein.

Figure 3  C4 CT slice. Cervical sympathetic chain schwannoma (S) pushing the jugular and carotid vessels forward. Pneumogastric (X) schwannoma enlarging the inter-jugulo-carotid space. 1: styloid; 2: external carotid artery; 3: pneumogastric nerve; 4: internal carotid artery; 5: internal jugular vein.
asymptomatic lateral cervical mass of progressively increasing volume [6]. The size of the mass may induce pharyngeal compression and non-specific signs such as pharyngeal discomfort or odynophagia [5]. CSCS is easier to diagnose if Horner's syndrome is associated, but this is rarely the case at the time of diagnosis, being related to nerve compression. Unlike neurofibroma, schwannoma is an encapsulated tumor which never infiltrates the perineural sheath or nerve fibers [6]. Imaging is essential to management [6,8]. CT and nuclear MRI determine tumor size, sector-II parapharyngeal location, extension and vascular relation to the internal and external carotids [8]. In case of Xth nerve involvement, the tumor tends to enlarge the space between the internal or common carotid and the internal jugular vein, while sympathetic involvement pushes the jugulo-carotid axis forward; in both these cases, location is retrostyloid (Fig. 3). CT can rule out non-vascularized adenopathy, congenital cyst and paraganglioma of X or the carotid glomus, which shows intense contrast medium uptake as of the arterial injection time, with a classic aspect of a tumor superior to and in contact with the carotid bifurcation, enlarging the space between the internal and external carotids [3]. Schwannoma shows lower density than the neighboring muscles; after injection, late enhancement is generally homogeneous, as vascularization is basically by intra-tumoral capillaries. CT is the most frequent means of first-intention cervical mass assessment after ultrasound. When schwanna is suspected, MRI is not indispensable, but may refine location and rule out X schwannoma, which is the first differential diagnosis. Fine-needle aspiration cytology is often non-contributive [2]. Macroscopically, anatomopathology diagnoses a spherical tumor with a definite capsule formed from the sheath; microscopically, the aspect is pleomorphic, with two architectural types according to Antoni: type A, fascicular, and type B, myxomatous, reticulated and more richly vascularized (Fig. 4). The proliferation index is low. On immunohistochemistry, S-100 protein is positive in all schwannomas and neurofibromas [9].

Surgery is the attitude of choice in schwannoma [3,5,7,10]. During cervicotomy, total CSCS excision usually involves sacrifice of the nerve. Horner's syndrome is then the most frequent postoperative complication [8]; dysphonia caused by trauma to X is rarer. Evolution is uncomplicated, and recurrence is rare if enucleation was complete. Peroperative neuro-monitoring may be useful in cervical nerve tumor, allowing partial tumor resection if need be—as in case of peroperative diagnosis of schwannoma of X.

Small weakly evolutive cervical schwannoma when detected raises the question of surgery again, especially when developing from the pneumogastric or greater hypoglossal nerve or a branch of the cervical plexus.

Conclusion

CSCS is a rare benign slow-growing tumor; when the volume is large, however, and no cytological diagnosis can be made, surgical exploration may be recommended. Contrast-enhanced head and neck CT is indispensable for preoperative assessment. Imaging shows a well-contoured retrostyloid tumor, vascularized in venous-time, pushing the carotid axis and internal jugular vein forward without enlarging the inter-jugulo-carotid space. The patient should be systematically alerted to the risk of postoperative Horner's syndrome, for which there is no effective form of treatment.

References