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Pathology in Focus

Horner's syndrome: a rare presentation of cervical sympathetic chain schwannoma

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Abstract
We describe a cervical sympathetic chain schwannoma in a 77-year-old woman who presented with a neck mass and Horner's syndrome. Such schwannomas are rare and this is the first documented case of a Horner's syndrome at presentation. The mass was excised via a cervical approach and her post-operative course was uneventful. The prognosis is excellent, with recurrence being rare. A brief discussion of the pathology, presentation, diagnosis, and treatment of this condition is made in this paper. The relevance of the uncertainty in diagnosis is discussed with the message that a pre-operative Horner's syndrome may guide the surgeon in the care of the patient but we suggest that in all cases proper counselling of the possible neurological consequences of this surgery be conducted.

Key words: Schwannoma; Neurilemmoma; Horner's syndrome

Introduction

Schwannomas are benign neoplasms originating in the Schwann cell of the nerve sheath which surrounds, but is anatomically distinct from, the nerve axis. They are the most frequent neoplasms arising in peripheral nerves, but are nevertheless uncommon (Brandenburg, 1972). Schwannomas have a wide distribution of origin but have a predilection for certain sites such as the roots of spinal nerves and the auditory nerve, and many arise in the head and neck. In the parapharyngeal space, schwannomas may arise from the last four cranial nerves or the autonomic nerves, and the vagus is the most common site. Cervical sympathetic chain schwannomas are rare and we were only able to find thirteen clearly documented cases (Kragh *et al.*, 1960; Daly and Roesler, 1963; Brandenburg, 1972; Clifton, 1977; Clairmount and Conley, 1978; Myssiorek *et al.*, 1988). A post-operative ipsilateral Horner's syndrome is to be expected following removal of such a tumour, due to the necessary division of the cervical sympathetic chain. We describe the first case in which a Horner's syndrome had developed pre-operatively, apparently as a result of the presence of the tumour.

Case history

A 77-year-old woman presented with a nine-month history of a painless swelling in the left anterior triangle of the neck. This had been progressively increasing in size but was not associated with any other symptoms such as dysphonia, dysphagia, pain or weight loss. Examination revealed a firm, non-tender ovoid mass about three by four cm situated just below the left submandibular gland. It felt pulsatile, possibly due to tranmission from the adjacent carotid artery. The patient had a left-sided Horner's syndrome, the symptoms of which she had not noticed. Examination of her larynx and pharynx, including post-nasal space, was normal. An ultrasound scan suggested an enlarged lymph node. Her chest radiograph and haematological indices were normal. She underwent excision of

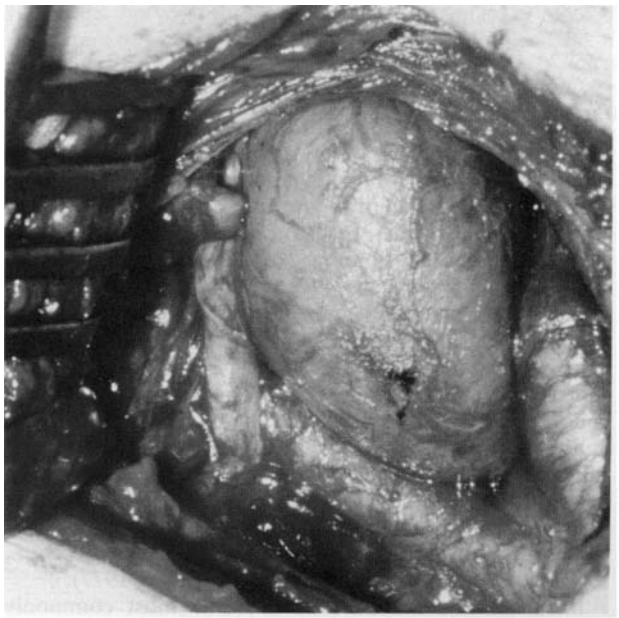

FIG. 1
The pre-operative appearance of the schwannoma arising between the internal and external carotid arteries.

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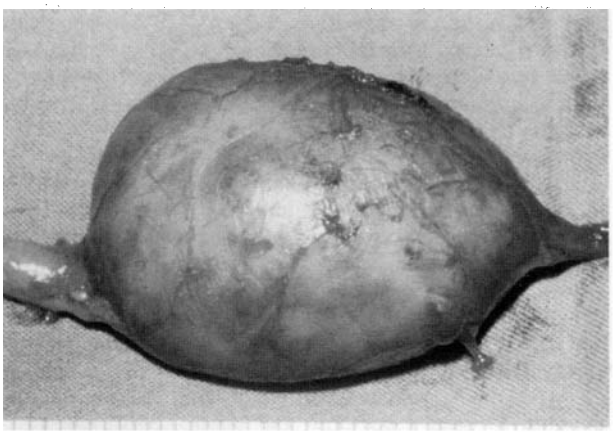

FIG. 2
Macroscopic appearance of the schwannoma after removal with stalks of cervical sympathetic chain shown.

the lump through an incision parallel with the anterior border of the sternomastoid muscle. A mass was seen lying on the prevertebral fascia between the internal and external carotid arteries, with the hypoglossal nerve superficial to it (Figure 1). The tumour was clearly arising from the cervical sympathetic chain. It was dissected out intact together with superior and inferior stalks of the cervical sympathetic chain (Figure 2). She made an uneventful recovery and was discharged on the fifth post-operative day. Histology showed the tumour to be a benign schwannoma arising within the cervical sympathetic chain (Figure 3).

Discussion

Peripheral nerve tumours, originally classified by Virchow (Virchow, 1863) may arise either from the neuron proper, such as the traumatic neuroma, or from the nerve sheath, the more common and important group – which includes truly neoplastic cases such as the schwannoma and neurofibroma. Schwannomas and neurofibromas derive from the Schwann cell, a cell that normally provides mechanical protection for the axon, produces and maintains the myelin sheath, and serves as a guide to regenerating nerve fibres. Although schwannoma and neurofibroma are closely related neoplasms, there are important reasons for separating them (Geschickter, 1935; Stout, 1935; Stout, 1946).

The schwannoma (neurilemmoma, solitary benign nerve sheath tumour, neurinoma or perineural fibroblastoma) is a slowly growing tumour which generally presents between 20 and 50 years of age (Geschickter, 1935; Sharaki *et al.*, 1982) and affects both sexes equally (Clifton, 1977).

Intracranially, the acoustic nerve is most commonly affected. Extracranial schwannomas occur in the head and neck region in 25 to 40 per cent of all cases (Gore *et al.*, 1956; Das Gupta *et al.*, 1969; Katz *et al.*, 1971) although they may be seen in the flexor surfaces of the upper and lower extremities (Stout, 1935) and deeply situated schwannomas may also be seen in the posterior mediastinum and the retroperitoneum.

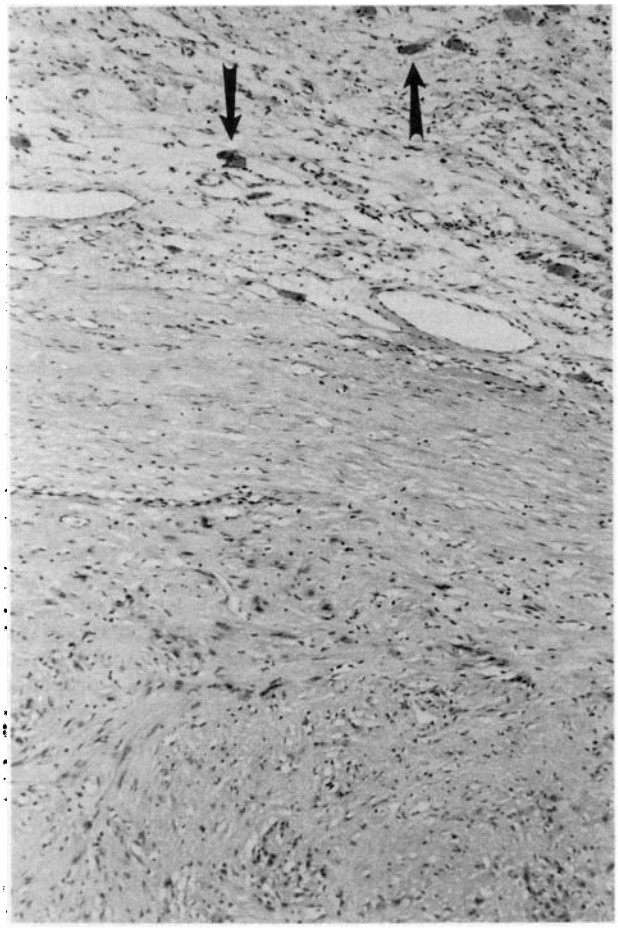

FIG. 3
Histological appearance of periphery of schwannoma (Antoni A area) with contiguous autonomic ganglion cells visible. (arrow) (H & E: × 175)

Neck schwannomas may be divided into the lateral group, arising from the cervical and brachial plexuses and the medial (parapharyngeal) group, arising from the last four cranial nerves (particularly the vagus) or the cervical sympathetic chain (Gore *et al.*, 1956; Daly and Roesler, 1963). Schwannomas are usually solitary lesions except in von Recklinghausen's disease (neurofibromatosis type 1), a disease characterized by multiple neurofibromas rather than schwannomas. When involving a small nerve there is very little restriction of mobility but in a larger nerve, movement along the nerve axis is restricted. Degenerative cysts within the tumour may give it an elastic feel, and occasionally the patient is aware that the swelling varies in size, a phenomenon thought to be related to fluctuations in the amount of cystic change.

Pain and neurological symptoms are uncommon unless the lesion becomes large. Dysfunction in the nerve of origin may result from axons being stretched over the tumour capsule but because adjacent nerves may also be affected by direct pressure, importantly therefore, a specific nerve dysfunction does not indicate with certainty the exact site of origin of the tumour.

Schwannomas very rarely arise from the cervical sympathetic chain. Myssiorek found only 11 clearly documented cases of cervical sympathetic chain schwannoma and added two further cases. All these cases presented as neck masses and, depending on size, were associated with dysphagia, lower cranial nerve palsies, conductive hearing loss and trismus (Myssiorek *et al.*, 1988). A pre-operative Horner's syndrome was not

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reported in any of these cases. Moreover, in our literature search we were unable to find any recorded cases of a pre-operative Horner's syndrome associated with a cervical sympathetic chain schwannoma. We therefore believe this to be the first documented case.

At operation, because the schwannoma arises within the nerve sheath, it is surrounded by a true epineural capsule. The appearance will vary from a small fusiform swelling to a larger eccentric mass, usually less than five cm maximum diameter, over which the fibres of the nerve of origin are splayed. A relatively large tumour may arise from the sheath of a very small nerve, hence the frequent failure to identify the nerve of origin. The cut surface is pink, white, or yellow, and larger lesions may show foci of haemorrhage and cyst formation. Microscopically, the fibrous capsule consists of epineurium with residual peripherally displaced nerve fibres, but true neural elements are absent. The tumour shows the hallmark Antoni A and B areas (Antoni, 1920). Antoni A areas are highly ordered and densely populated with cells arranged in short bundles or interlacing fascicles. Antoni B areas are myxoid and far less orderly or cellular. Electron microscopy suggests they may be degenerate Antoni A areas. The large irregularly spaced blood vessels, often with lumina filled with thrombus, that are very characteristic of schwannoma become more conspicuous in the hypocellular Antoni B areas.

Schwannomas are histologically benign. Following simple excision, enucleation, or even incomplete excision (if a more extensive procedure would have caused unacceptable functional results) recurrence is rare (Stout, 1935). Malignant change is exceedingly rare, and from a practical point of view can be discounted.

The pre-operative diagnosis of a cervical sympathetic chain schwannoma is difficult and investigations are usually unhelpful. The carotid artery may be displaced either medially or laterally but usually anteriorly (Guggenheim, 1953; Cutler and Gross, 1936). The differential diagnosis is vast and includes lymphadenopathies, thyroid neoplasms, paraganglioma, branchial cyst, deep lobe parotid neoplasms, minor salivary gland neoplasms, lipoma, carotid artery aneurysm, neurofibroma, teratoma, rhabdomyoma, and meningioma. Usually, the site of origin can be first identified only at biopsy, or if not then after histological analysis. Clinically it is very important to conduct a thorough pre-operative examination of the local neurology including actively looking for a Horner's syndrome because this will guide the surgeon and may prevent problems with post-operative complications. When discussing the operation with the patient, because of the uncertain diagnosis as indicated above one should advise the patient of the possible neurological consequences including a Horner's syndrome.

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