ABSTRACT

Introduction. Schwannomas of the head and neck develop from the sheaths of cranial, peripheral or autonomic nerves. Between 25% and 45% of schwannomas occur in this region. Schwannomas of the sympathetic chain are very rare, and the literature describes just over 40 cases, localised mostly in the neck or retroperitoneal areas.

Case report. We present the case of a 38 year-old woman with a large schwannoma of the cervical sympathetic chain. At its widest, the tumour reached a maximum diameter of 130 mm. Since the mediastinal component was so large, the tumour was operatively excised via a right posterolateral thoracotomy. Microscopic analysis confirmed the diagnosis of benign schwannoma. A computed tomography scan six months after resection showed no evidence of local recurrence.

Conclusion. Schwannomas of the cervical sympathetic chain are extremely rare tumours. Proper diagnosis requires microscopic tumour tissue analysis. Although these tumours are benign, they can cause a variety of symptoms while growing, necessitating urgent operative treatment. Key words: schwannoma, sympathetic chain, head and neck neoplasms, surgery, pathology

INTRODUCTION

Schwannoma (also known as “neurilemmoma”) is a rare tumour arising from the nerve sheaths of cranial, peripheral and autonomic nerves. Between 25% and 45% of these tumours are located in the cranial and cervical regions, and schwannomas most commonly arise from cranial nerves and their branches. Clinically, they present as solitary, painless, slow-growing tumours, sometimes followed by neuralgia and paresthesia in the corresponding region. The treatment of choice is complete surgical excision to prevent neurological deficits and recurrence. Since these tumours rarely undergo malignant transformation, the primary goals of surgery should be preserving and repairing nerve function.

Schwannomas in this location often imitate other conditions such as infections or metastatic tumours. Although their incidence is quite low, surgeons must consider the potential neurogenic origin of these tumours. When a schwannoma is incompletely resected, permanent nerve damage may occur. Schwannomas are diagnosed using computed tomography (CT) and magnetic resonance imaging (MRI) imaging in addition to angiography, and histological examination of resected tissue is required to confirm the diagnosis.

ABBREVIATIONS

α-SMA - smooth muscle actin; CT - computerised tomography; GFAP - glial fibrillary acidic protein; EMA - epithelial membrane antigen; MRI - magnetic resonance imaging; US - ultrasound.


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CASE REPORT

A 38 year-old woman was admitted to our hospital for swelling in the right half of her neck. She described a one-year history of occasional aching in the right half of her chest and difficulty swallowing. Physical examination revealed a painless, movable mass located in subcutaneous tissue in the right supraclavicular region. No neurological deficit was detected. Laboratory tests and spirometry parameters were within normal limits. Ultrasound (US) scan of the neck revealed a hypoechoicogenic mass in the right supraclavicular area, inferior to the main blood vessels. Radiographic, posterior-anterior films of the chest revealed a clearly defined shadow that increased the pressure on the trachea, causing leftward tracheal deviation. CT scans revealed an expansive mass, sized 98 x 78 x 70 mm, located in the upper mediastinum, with the intensity of the non-homogenous soft tissues (the attenuation of 40HU). No pathological changes were observed in the surrounding lung parenchyma. The tumour compressed trachea and brachiocephalic vein, but there was no clear evidence of infiltration (Figure 1).

Based on these imaging procedures, the patient was given a clinical diagnosis of a tumour in the upper mediastinum. A multidisciplinary medical team recommended a right postero-lateral approach for surgical excision because of the bulky mediastinal portion of the tumour, despite expansion of the tumour into the neck.

After thoracotomy, a rounded mass was visualised, with a maximum diameter of 130 mm, below the upper thoracic aperture. The root originated over the upper edge of the first rib. The mass was identified as a cervical sympathetic chain tumour. To safely resect the tumour, we first reduced the tissue and then removed it together with the tumour capsule.

The patient experienced Horner’s syndrome after the operation, but the postoperative period was otherwise uncomplicated. The patient was released from the hospital after two weeks. A CT scan performed six months after the resection showed no signs of a local recurrence.

Three tissue samples of irregular shape (maximum diameter of 23, 52 and 61 mm), with total weight of 210 grams, were sent for pathological examination (Figure 2). Macroscopically, they had smooth and shiny surfaces, and the cross-sections were white-yellowish in colour with a glassy and firm consistency. Microscopic analysis revealed an encapsulated tumour with a clear border and spindle-shaped cells grouped in short and long fascicles oriented in different directions (Figure 3a). The tumour was uniformly cellular, though there were a few hypercellular zones with palisade nuclei forming Verocay’s bodies and areas of myxoid degeneration (Antoni A component). Pathologists found no evidence of cytological atypia, mitosis or necrosis. The tumour had scant macrophage infiltration, focal aggregates of lymphocytes, and zones of cystic degeneration together with hyaline in the blood vessel walls. Immunohistochemically, tumour cells diffusely expressed vimentin (Figure 3b), S-100 protein (Figure 3c) and glial fibrillary acidic protein (GFAP) (Figure 3d), but lacked epithelial membrane antigen (EMA), desmin, CD 31, CD 34, smooth muscle actin (α-SMA) and CD 68 (Figure 3e).

DISCUSSION

Schwannomas were first described in 1910 by Verocay.2 They have alternatively been called neurilemmomas, solitary tumours of nerve sheaths, and peripheral fibroblastic tumours but the WHO has recommended the term schwannoma.3, 4 In general, schwannomas are solitary tumours, whereas the presence of multiple schwannomas is a component of von Recklinghausen disease. In the head and neck, schwannomas can originate from any peripheral, spinal or cranial nerves except the optic or olfactory nerves.5

Schwannomas of the sympathetic chain are very rare. Approximately 40 cases have been described in the literature, and these are most often localised to the cervical and lumbar retroperitoneal region.6-11, 12-16 These tumours usually affect patients from 20 to 50 years old, and men are affected three times more often than women.16 Schwannomas can grow to 20 cm in diameter.17 These large tumours are often associated with focal haemorrhage, calcifications or cystic degeneration of the lesion.18

Most schwannomas of the cervical region are asymptomatic. Depending on their exact location, cervical schwann-
nomas can cause tonsillitis, hoarseness, dysphagia and pain. Although schwannomas infrequently compress or infiltrate surrounding organs, in these rare cases, the sympathetic chain is usually affected, resulting in pain and paresthesias. Several cases have been associated with Horner’s syndrome.6, 8, 10, 11, 13, 14

Sympathetic hyperactivity can also occur, and one report describes a cervical sympathetic chain schwannoma that caused ipsilateral lacrimation, palmar hyperhidrosis, conjunctival injection, and nasal congestion.9

The challenge in the treatment of these tumours is to differentiate a benign schwannoma of the sympathetic chain from other pathological processes with similar presentations. Depending on the location of the tumour, the differential diagnosis could include lesions of the carotid artery, paragangliomas, sarcomas and malignant schwannomas. Careful clinical examination is important, but histopathological and immunohistochemical analyses are required to confirm the diagnosis and exclude malignancies.

Preoperative imaging procedures including MRI, CT and angiography are particularly helpful. On an MRI scan, schwannomas show a high intensity signal on T2-weighted imaging and a low intensity signal on T1-weighted scans. In comparison to paragangliomas, schwannomas are not vascularised. Non-contrast CT shows a mass less dense than surrounding muscle tissue. CT scans with contrast show heterogeneous distribution of the contrast agent.

US can help to distinguish a vagal nerve schwannoma from a schwannoma of the sympathetic chain. Carotid angiogram can also be important in the diagnosis of cervical tumours. Carotid body tumours are characterised by hypervascularity while schwannomas lack this distinguishing feature.12, 15

Cervical anatomy can give insight into the appropriate differential diagnosis for a cervical tumour. Paragangliomas have a cranial origin and spread laterally. Schwannomas mostly occur in the parapharyngeal region, affecting cranial nerves IX, X, XI and XII. If they include the vagal nerve, the schwannomas divide the common carotid artery and internal jugular vein, while schwannomas of the sympathetic chain do not divide these vessels.7

Microscopic and immunohistochemical analysis are key to establishing a definite diagnosis. According to the WHO classification of central nervous system tumours, schwannomas are defined as the tumours originating from nerve sheaths.3, 4 They are further classified into cellular, plexiform and melanotic types.3, 4 Immunohistochemically, they are characterised by expression of S-100 protein and GFAP.19

In our case, the presence of Antoni A and Antoni B zones, microscopic patterns pathognomonic for schwannomas, confirmed our diagnosis.20 We observed nuclear palisades in several tumour specimens, which formed Verocay’s bodies. In addition, schwannomas of cranial nerve VIII characteristically lack nuclear palisades.20 Schwannomas are typified by cystic and hyaline degeneration of blood vessel walls, and they have macrophages and lymphocytic aggregates.19 Ki-67 is expressed only in some tumour cells, which aligns with reports that mitosis is rarely observed in schwannomas.21

The appropriate schedule for surgical resection and post-operative follow-up remains controversial.22 In our case, there were several indications for immediate intervention. Our patient was young, and she had a large sympathetic chain schwannoma that was likely to grow quickly and/or convey novel symptoms during the disease course. Furthermore, our differential diagnosis included other tumour types that require urgent surgical intervention. Since some schwannomas are malignant and benign schwannomas can undergo malignant transformation, young patients may have a higher risk of having cancerous schwannomas.23 Surgical intervention was indicated since only microscopic examination of biopsy specimens could yield a precise diagnosis.

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Figure 3. Histological features of a schwannoma: a) the tumour is composed of spindle cells grouped in shorter and longer fascicles (HE staining technique, x200). The tumour cells express: b) vimentin (x100), c) S-100 protein (x200), and d) GFAP (x200). e) The tumour cells are weakly, focally positive for CD 68 (x200), and f) the Ki-67 proliferation index is very low, less than 1% (x200).
In our patient, the surgeon had to resect the tumour through a right posterolateral thoracotomy. Direct visualisation revealed that the tumour originated from the sympathetic chain. After the schwannoma and cervical sympathetic chain were removed, the patient experienced Horner’s syndrome postoperatively. Previous studies reported similar neurological deficit in patients with this kind of tumour. 10, 11, 13, 16, 23, 24 Classical Horner’s syndrome is characterised by lesions of the oculosympathetic pathway at any point between the hypothalamus and eye. It presents as pupillary miosis, ptosis, enophthalmos and facial anhydrosis. Ptoxis is a consequence of paralysis of Müller’s muscle, and it can be corrected by strengthening the levator aponeurosis or resecting the adjacent conjunctiva and muscles. 7, 25 Aside from the obvious aesthetic consequences, Horner’s syndrome does not cause functional deficits.

Schwannomas of the cervical sympathetic chain are extremely rare tumours. They are difficult to diagnose preoperatively, and clinical diagnosis must be confirmed by microscopic analysis of resected tumour tissue. Complete surgical excision is the treatment of choice and recurrence is rare. Since most schwannomas are benign, long term follow-up is usually not required.

REFERENCES