Case Report

An Unusual Parapharyngeal Tumor: Cervical Sympathetic Chain Schwannoma

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Abstract Schwannomas are benign, solitary, and well-differentiated tumors originating from Schwann cells. They may originate from any of the peripheral, cranial, or autonomic nerves of the body with the exception of the olfactory and the optic nerves. Schwannomas arising from the cervical sympathetic chain are very rare. Only < 65 cases have been reported in the literature to date. Here we report a case of cervical sympathetic chain schwannoma in a 41-year-old lady presenting as a parapharyngeal mass which was excised by a transcervical approach without any postoperative Horner’s syndrome.

Keywords parapharyngeal tumor; cervical sympathetic chain; Schwannoma

1. Introduction

Schwannomas were first described by Verocay in 1908 [18]. Since then, they have been called as neurilemmomas, solitary nerve sheath tumors, perineural fibroblast tumors, and recently as schwannomas according to the WHO classification [11]. Nearly 45% of all schwannomas occur in the head and neck area, and the commonest site in the head and neck area is the parapharyngeal space. In the parapharyngeal space, schwannomas may arise from the last four cranial nerves or the autonomic nerves, the vagus being the most common site. Cervical sympathetic chain schwannomas are uncommon [2,12,15] and most often appear as an asymptomatic, slow-growing, solitary neck mass. Horner’s syndrome is rarely apparent on clinical examination [8]. We are reporting a case of cervical sympathetic chain schwannoma presenting as a parapharyngeal mass in a 41-year-old lady.

2. Case report

A 41-year-old lady presented with an asymptomatic swelling in the left upper neck of 5 years duration. There was no history of hoarseness, nasal regurgitation, aspiration, syncopal attacks, associated pain, fever, or trauma. Clinical examination revealed a firm, 7 × 4 cm swelling in the left carotid triangle (Figure 1). It was mobile, non-tender, and non-pulsatile, with no associated bruit. There were no palpable neck nodes. Oropharyngeal examination revealed a visible bulge in the left tonsil and posterior pillar. On indirect laryngoscopy, both vocal cords were mobile. Cranial nerve examination was normal.

Ultrasoundography showed a 7 × 4 cm mass displacing the carotid arteries slightly forward. Contrast-enhanced CT scan of the neck revealed a well-defined contrast-enhanced mass of 7.2 × 4.2 cm in left carotid triangle pushing the carotid artery anteriorly and compressing the internal jugular vein. FNAC was done and was suggestive of lymphoepithelial cyst.

A provisional diagnosis of left parapharyngeal mass was made and was taken up for excision of the mass through a transcervical approach under GA. An upper transverse neck incision was put (Figure 2). Deep to the upper portion of the sternocleidomastoid muscle, an encapsulated mass was found. The internal jugular vein was stretched over the mass and so was forced to sacrifice. The common carotid artery with its external and internal branches and the vagus nerve
Figure 2: An upper transverse neck incision is put.

Figure 3: Intraoperative photograph after tumor excision showing the carotid bifurcation, vagus nerve, and hypoglossal nerve.

Figure 4: A pear shaped $7 \times 4$ cm tumor is excised.

Figure 5: Intraoperative photograph shows the intact cervical sympathetic chain after tumor excision, which appears thickened.

were displaced anteriorly. The tumor was found to originate from the cervical sympathetic chain and did not involve the vagus, hypoglossal, spinal accessory, glossopharyngeal, or lingual nerves. The upper part of the mass was extending to the parapharyngeal region, and it was easy to recognize the cervical sympathetic chain inferior to the tumor. The mass was eccentric to the nerve and was surrounded by a well-defined capsule. Complete surgical removal of the mass, without sacrificing nerve fiber was possible as the capsule was easily separable from the underlying fibers (Figures 3 and 4). The cervical sympathetic chain was found thick after the complete excision of the mass (Figure 5).

Surprisingly, there was no Horner’s syndrome in the post-operative period. Histopathological examination of the specimen confirmed the tumor to be a benign schwannoma originating from the cervical sympathetic chain (Figure 6).
3. Discussion

Cervical sympathetic chain schwannomas (CSCS) are rare, benign tumours originating from the superior or middle part of the cervical chain [5] and typically located in the retrostyloid compartment of the parapharyngeal space. Most of the cervical sympathetic chain schwannomas present as asymptomatic solitary neck masses. They grow slowly, approximately 3 mm per year [6]. They are usually seen in patients between 20 and 50 years of age. Frequency is the same in both sexes, and malignant change is rare [12]. Features of nerve compression are rare because the cervical sympathetic trunk lies in a relatively loose fascial compartment. Preoperative Horner’s syndrome is unusual and has only been reported in six cases of CSCS [8]. Pulsation is an atypical finding and suggests the presence of a carotid body tumor at initial workup. Here angiography may be necessary for the differential diagnosis [14]. Pulsation may be due to reflection of the carotid artery system, or it may be true pulsation caused by the hypervascularity of the schwannoma [3].

CT with contrast medium or magnetic resonance imaging is essential to the initial workup for CSCS. A mass pushing the internal carotid artery or common carotid artery anteriorly is suggestive of a schwannoma originating from the sympathetic chain or vagus nerve [16]. Schwannomas typically have well-delineated margins. They commonly show higher attenuation than adjacent muscle on contrast-enhanced CT but may be isodense or, less commonly, of lower attenuation than the adjacent muscle [16]. On MRI, schwannoma appears to have intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images [20].

Replacement of vascular structures due to the mass effect of the schwannoma on radiological imaging may give an idea of the origin of the lesion. Schwannomas originating from the vagus nerve cause separation of artery and vein on Doppler US since they grow between the common carotid artery and internal jugular vein or internal carotid artery and internal jugular vein. No separation is seen between the artery and the vein in CSCS [7]. In addition, the course of the vagus nerve can be seen with ultrasound. In CSCS, the vagus nerve courses superficial to CSCS, whereas there is a connection with the tumor in vagal schwannomas [7].

If the tumor presents as a pulsatile mass, carotid body tumor must be considered. A “salt and pepper” pattern on postgadolinium MRI sequences is commonly seen in carotid body tumor, but it is not pathognomonic as this may be found in other hypervascular lesions and in CSCS [4,9,19]. In carotid body tumor, CT and MRI display a homogeneous and intense pattern of enhancement following intravenous contrast, while enhancement in schwannoma is less intense and dishomogeneous. Splaying of the carotid bifurcation, typical of carotid body tumor, can also be found in schwannoma arising from the lower four cranial nerves or the sympathetic chain [17]; the main imaging criterion to differentiate CBT and a nerve sheath tumor is hypervascularity. This can be demonstrated by USS, contrast CT, MRA and conventional angiography. Demonstration of contrast agent accumulation, the absence of arteriovenous shunts, and low-degree vascularity in DSA should suggest a schwannoma [1].

Treatment of CSCS is total excision of the lesion. During surgery, the appearance of the tumor can suggest the right diagnosis if the lesion presents as a fusiform mass, eccentrically to the nerve and surrounded by a capsule. Complete surgical removal of the mass, without sacrificing nerve fiber, is possible only when the capsule is easily separable from the underlying fibers. In our case, we could remove the mass without sacrificing cervical sympathetic chain, as the capsule was easily separable from the underlying fibers. When dissection of the capsule from the nerve is not easy and there are no signs of malignancy, functional loss can be minimized by opening the capsule longitudinally and removing the tumor from inside [13]. Most of the time CSCS cannot be removed without sacrifice of some nervous fibers or section of the sympathetic trunk. Since cervical sympathetic chain damage is well tolerated, restoration of the nerve has only been rarely performed, while in vagal schwannomas, the practice of nerve reconstruction is recommended [10,13].

4. Conclusion

Cervical sympathetic chain schwannomas are rare tumors. These lesions are commonly asymptomatic or present with non-specific symptoms, and accurate preoperative diagnosis is not always easy. Imaging examinations cannot reveal the exact origin of the tumor. Only surgical observation of the lesion and the nerve from where it originates, and histologic examination of the specimen, can lead to a correct diagnosis. An accurate preoperative workup is useful for surgical...
planning and informing the patient about any possible complications. In our patient, we could remove the tumor without sacrificing cervical sympathetic chain, as the capsule was easily separable from the underlying fibers and so, there was no Horner’s syndrome in the postoperative period.

References


