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CASE REPORT

Cervical sympathetic chain schwannoma: A case report

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Abstract Nerve tumors arising from the sympathetic chain are uncommon slow-growing tumors and represent a diagnosis challenge. Their malignant degeneration is rare. Definitive pre-operative diagnosis may be difficult as investigations are not usually helpful. We report the case of a 23-year old woman who presented with an asymptomatic solitary left cervical swelling. She was evaluated with sonography and computed tomography. Complete surgical excision of the lesion was carried out and histologic examination revealed a schwannoma. Post-operatively, the patient showed clinical findings of Horner's syndrome. Pathologic and radiological evaluation, differential diagnosis of this neoplasm and its management are discussed.

1. Introduction

Schwannomas are benign nerve sheath tumors deriving from Schwann cells that occur in the head and neck region in 25–45% of cases.[1] Cervical lesions originate from spinal nerves, the last four cranial nerve roots, or occasionally from the sympathetic chain. The latter are extremely rare. 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 (CSCS) are uncommon and most often appear as an asymptomatic, slow-growing, solitary neck mass.[2]

We report a case of schwannoma arising from the cervical sympathetic chain presenting as a swelling in the upper neck. Clinical findings, surgical management and outcomes of this tumor are reviewed.

2. Case report

A 23-year-old woman who had no past medical history, presented with a painless swelling of the left upper lateral neck lasting for 2 months and slowly increasing in size. There was no history of hoarseness, compression complaints, nasopharyngeal discomfort, otalgia, syncopal attacks or associated pain. Physical examination revealed a visible bulge of the left tonsil and the left posterior pillar. Neck examination showed a firm oblong mass measuring approximately 4×3 cm located in the left mid jugular chain area. The mass was painless, non-pulsatile, non-expansive and movable. The overlying skin was normal and there were no palpable neck nodes. Nasal

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endoscopy was normal and indirect laryngoscopy excluded any vocal cord paralysis. Fine needle aspiration of the mass produced insufficient material for cytological examination. Ultrasonography showed a hypoechoic homogeneous well-circumscribed mass located in the upper internal jugular area with no fatty hilum (Fig. 1).

Computed tomography revealed a 30×22 mm mass displacing the carotid artery and the jugular vein antero-medially, with no neck nodes observed (Fig 2).

The patient underwent surgical treatment with a provisional diagnosis of lymphadenopathy, schwannoma or paraganglioma. A transcervical approach was performed (Fig 3). Deep into the upper portion of the sternocleidomastoid muscle, an encapsulated 3×2 cm tumor was found (Fig 4). The carotid artery and the internal jugular vein were displaced medially but not compressed. The mass appeared to originate from the cervical sympathetic chain and left vagus was intact. It was found to be well encapsulated and was carefully dissected from the nerve. The capsule was incised longitudinally and tumor excised completely from inside. The frozen section of the surgical specimen revealed a benign mesenchymal tumor.

Post operatively, a left side Horner's syndrome occurred but without facial anhidrosis. This complication was well tolerated and the patient put under corticosteroids systemically for one week (Fig 5). She was discharged on the third post-operative day.

Histopathological examination of the specimen confirmed the tumor to be a benign schwannoma originating from the sympathetic chain. It showed spindle shaped cells associated with nerve cells.

3. Discussion

Schwannomas are slow-growing, encapsulated tumors that arise from the peripheral cells located in the peripheral nerve sheath.[4] The retrostyloid compartment of the parapharyngeal space is the most common site of origin.[1,4,7] In the neck they tend to arise either medially (glossopharyngeal, vagus, accessory, or hypoglossal nerves) or, as in our case, from the sympathetic chain.[4] Langer [10] found that among 21 patients suspected to have cervical schwannomas, sympathetic chain was affected in 4 cases (19%).

Though benign, they can undergo malignant transformation.[5,6] They occur more frequently in adults 20–50 years old but are even observed in patients aged 5–77, without sex preference.[1,2]

Majority of schwannomas are asymptomatic but may cause dysphagia or sore throat by exerting pressure on the surrounding structures.[7] Features of nerve compression are rare because the cervical sympathetic trunk lies in a relatively loose fascial compartment and schwannomas, by nature, are noninfiltrative.[11]

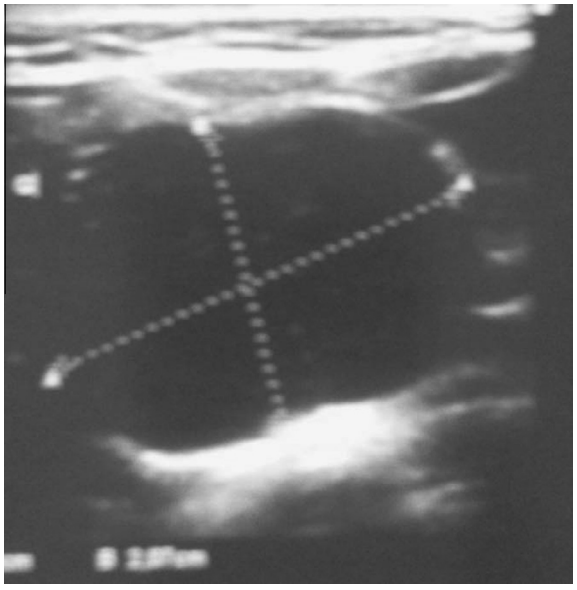

Figure 1 Ultrasonography of the neck showing an oval mass of the upper internal jugular chain.

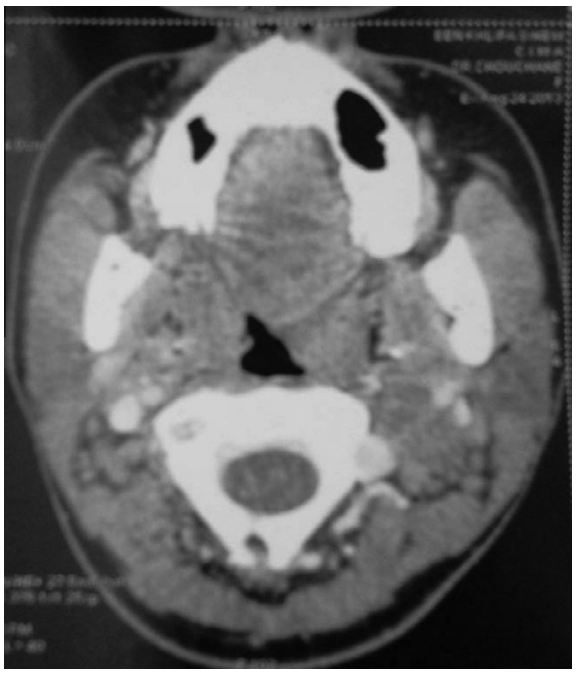

Figure 2 Axial section revealing the hypodense oval-shaped well-defined 30×22 mm mass.

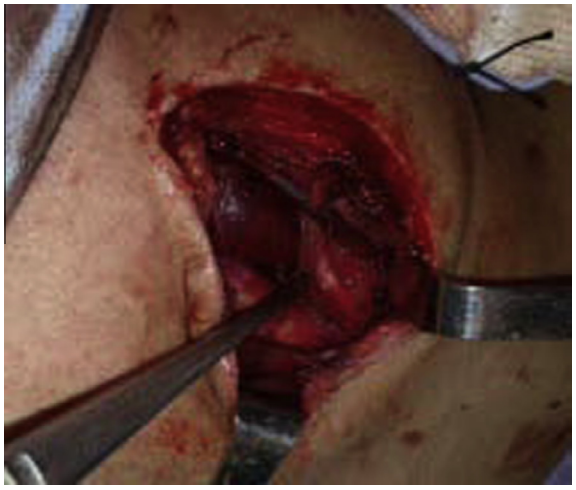

Figure 3 Resection of the schwannoma.

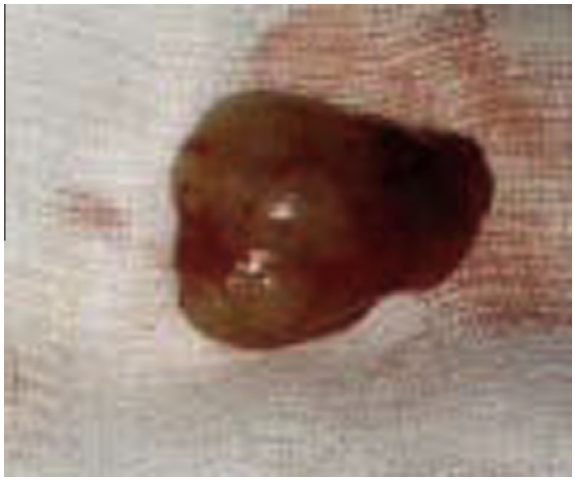

Figure 4 Excised specimen.

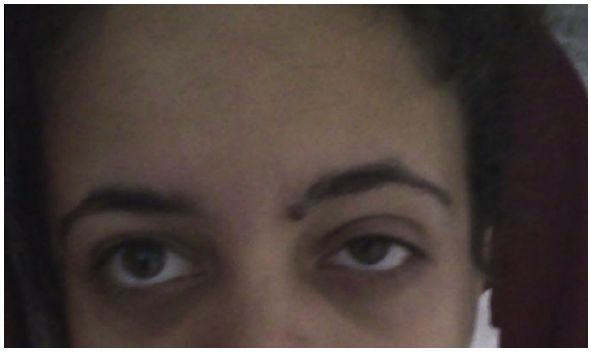

Figure 5 Horner's syndrome occurring post operatively.

Pulsation is an atypical finding for CSCS and may be due to schwannoma hyper-vascularity or reflection of the carotid artery system.[3]

Pre-operative diagnosis is difficult and further investigations are necessary such as US, CT, MRI and angiography[2] but CSCS do not present with specific symptoms or imaging signs. Our patient was examined only by US and CT.

Contrast enhanced CT is the best initial diagnostic study to determine the size and extent of tumor, to demonstrate degree of tumor vascularity and to help the surgical approach.[7] The mass appears hypodense with some degree of enhancement[2] while paraganglioma is classically isodense. A mass on contrast-CT pushing the internal carotid artery or common carotid artery anteriorly is suggestive of schwannoma originating from the sympathetic chain or vagus nerve.[1] MRI has become the routine imaging study for these tumors and the schwannoma is generally hypointense on T1 and hyperintense on T2-weighted images, depending on its cellularity.[1,2] A normal ganglion is usually hypointense on T2, a finding that may help distinguish it from a schwannoma or enlarged retropharyngeal node.[11] Anil[11] found that MR imaging describes the internal heterogeneity better than CT. The delineation of the surrounding fat planes and the relationship with soft tissue and vascular structures are also better demonstrated on it.[11]

This radiological examination has a great value in the pre-operative work-up as it is helpful in defining diagnosis and evaluating the extent and the relationship of the tumor with the jugular vein and the carotid artery.[8,13] It can be useful to determinate the nerve of origin.[8] In most cases (67%), the internal carotid artery is displaced anteriorly with a component of lateral displacement but can be in a neutral position when the tumor is much smaller than average (1.5×1 cm).[11] The anterior displacement of parapharyngeal fat and the medial displacement of the visceral space are typical of a carotid space lesion.[11] A marked enhancement of the solid component is seen after the administration of Gadolinium and the random distribution of Antoni A and B within the tumor is responsible for the dishomogeneous signal at MRI.[1,11] Schwannomas can appear as hypovascular or moderately hypervascular tumours on angiography which are indicated if the mass is contrast enhancing and if vessels appear to be compressed by the tumor.[1]

Fine-needle aspiration cytology (FNAC) has revolutionized the diagnosis of head and neck masses. However the ability of FNAC to reveal diagnosis is poorly defined in neurogenic tumors as it was seen in our case. It can lead to correct diagnosis in only 25% of cervical schwannomas.[1,8] Schwann cell is common to both schwannoma and neuro fibroma, which can only be distinguished microscopically.[7]

The main differential diagnosis for this tumor is paragangliomas and other pathologies of the retrostyloid parapharyngeal space.[1,7,11,12] Preoperative diagnosis is very important in these retrostyloid parapharyngeal masses as management of carotid body tumors varies from surgery to radiation to observation, while complete surgical excision is the therapy of choice in vagal and cervical sympathetic schwannomas.[12]

Paragangliomas show early arterial enhancement on CT; they are hypervascular and show certain characteristic MRI appearances like scattered flow voids.[11,12]

Carotid body tumor (CBT) must be considered, especially if the mass is pulsatile and hypervascular.[3,12] This can be demonstrated by US, contrast CT, MRA and conventional angiography.[1] In some cases, the schwannoma separates the internal and the external carotid artery mimicking a carotid body tumor but the vessels are never encased by the mass with less than 180° of contact between them.[11] Schwannoma of the vagus nerve grows between the internal or common carotid artery and the internal jugular vein; which is not seen in CSCS.[1,8,11–13] In fact, vagal schwannomas displace the internal jugular vein laterally and the carotid artery medially, whereas schwannomas arising from the cervical sympathetic chain displace both the carotid artery and jugular vein anteriorly without separating them.[8,11,13] In our case, there were no radiologic signs of separation between the common carotid artery and the internal jugular vein on the CT and no clinical signs of vagal involvement were evident. During surgery, the tumor was fusiform and surrounded by a capsule eccentrically to the nerve, thus suggesting the right diagnosis.

Schwannomas being well encapsulated tumors can be completely removed during surgery.[7] This requires an adequate exposure of the tumor and the neurovascular structures. Transcervical, transmandibular and transparotid are the main approaches described for the parapharyngeal space.[1,7] Complete surgical removal of the mass, without sacrificing nerve fiber, is possible only if the capsule is easily separable from the underlying fibers.[13]

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 often recommended.[1]

Cautious surgical treatment include extracapsular 'peeling' or even intracapsular enucleation of the tumor from the nerve in an effort to preserve its function.[13] If the lesion is known to be a schwannoma, it is possible to open the capsule and shell out the tumor, thereby leaving the capsular nerve fibers undisturbed and possibly avoiding functional deficits.[9,13] The neurological functions can also be monitored using a nerve stimulator or under a microscope in performing the intracapsular enucleation.[13] According to Zbären[14] there was no significant difference in the recurrence rate between the total tumor resection including nerve fibers and the intracapsular enucleation.

On histological examination, schwannomas are encapsulated, solid or cystic tumors. They can be composed of two cellular zones: Antony type A, densely arranged with spindle-shaped Schwann cells and areas of palisading nuclei, and Antony B, which tend to be hypocellular with a large quantity of myxoid tissue.[1,2,8] Other typical features include necrosis, hemorrhage and cystic degeneration.[8] Malignant schwannoma tumors differ from the benign type in their higher mitotic rate, the presence of necrosis, their infiltrative appearance and irregular positivity for the S-100 protein.[10]

Postoperative complications involve Horner's syndrome with miosis, anhydrosis, enopthalmos and ptosis. It's a common postoperative sequela associated with the close relationship between nerves and the tumor mass.[4] Its frequency ranges in the literature from 23% to 50%.[8,10] In our case, a partial well tolerated Horner's syndrome occurred on the left side of the face because the tumor was too intimately associated with the cervical sympathetic chain to preserve the integral nerve function.

4. Conclusion

SCSC, although rare, should be considered as a differential diagnosis in patients presenting with a lateral neck mass. FNAC is usually inconclusive, but radiological examination has a definite role in diagnosis thus excluding lymphadenopathies and salivary gland tumors.[12,13] Total resection is the treatment of choice with recurrence being rare, since radiotherapy and chemotherapy are of limited effectiveness[10] and only histologic examination can lead to a correct diagnosis.[2] Horner's syndrome is a common post-operative neurological consequence which is relatively well tolerated and should be discussed with the patient during preoperative counseling.[2] A close follow-up is mandatory.

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Conflict of interest

No.

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