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Schwannomas—Atypical Presentation and Challenges

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Abstract

Introduction: Neurogenic tumors include schwannomas and neurofibromas. They occur often in the head and neck. However, their occurrence on the vagus is uncommon. A high index of suspicion is needed to order imaging in pelvic lesions that present atypically. **Discussion:** Schwannomas are hypointense on T1 and heterogeneously hyperintense on T2 on MRI. The histopathological appearance with Antoni type A and type B is typical of Schwannoma. **Conclusion:** Schwannomas are slow growing benign tumors that are separable from the parent nerve. Recurrence is uncommon after resection.

Keywords

Cervical, Presaccral, IHC

1. Introduction

Schwannomas are neural sheath tumors. They are commonly present in the neck, extremities and retro-peritoneum. The lesion grows gradually and superficial lesions are usually small at the time of diagnosis, but in case of retroperitoneum, large lesions present without any signs and symptoms. Although schwannomas are generally benign lesions, they are known to increase in size 2.5 to 3 mm/year. Among schwannomas, vestibular type is the common cranial nerve schwannoma followed by facialand trigeminal schwannomas and then the glossopharyngeal, vagus and spinal accessory nerve schwannomas.

2. Case Presentation

2.1. Case 1

A 71 years old male presented with slowly progressive swelling on the right side

of the neck for 1 year, which was painless. Examination revealed a well circumscribed oval (6 × 4 cm) swelling (firm in consistency), non pulsatile, located in the posterior triangle (Figure 1). Ultra-sonogram of neck showed a complex well encapsulated predominantly solid mass, with no significant lymphadenopathy. CECT of the neck showed a large well defined thick walled hypodense lesion in the right posterior cervical space, in the region of right jugular chain (5 × 4.6 × 7.5 cm) with ill-defined relative hypodensity. It superiorly extended upto C2 vetrebra, inferiorly up to end plate of C7, laterally displacing right sternocleidomastoid, medially abutting the vascular chain (Figure 2). USG guided FNAC showed spindle like cells arranged in a cellular and loose areas suggestive of nerve sheath tumor. Pre-operatively the vocal cords were mobile. Intra-operatively a firm/cystic tumor was identified arising from the vagus, posterior to the IJV and displacing the carotids medially (Figure 3(a)). Intraoperatively the patient had multiple episodes of bradycardia during dissection of the tumor. The anaesthetist was intimated and bradycardia was managed by intra venous doses of atropine. The tumor was mobilized and resection was done in toto and the specimen was sent for biopsy.

HPE: Histopathology showed cellular areas composed of spindle cells and Verocaybodies. Hypercellular area showed spindle cells in myxoid areas and congested blood vessels were seen.IHC was positive for S 100 and less than 3% for ki67 (a proliferative index marker) (Figure 3(b)).

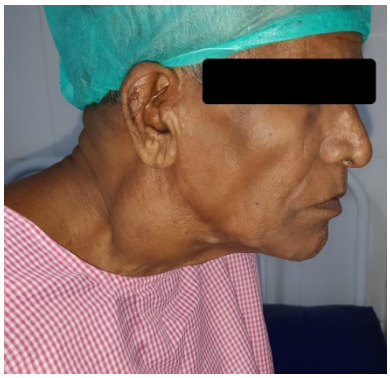


Figure 1. Swelling in the posterior triangle of neck.

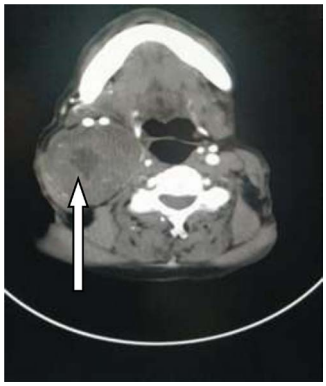
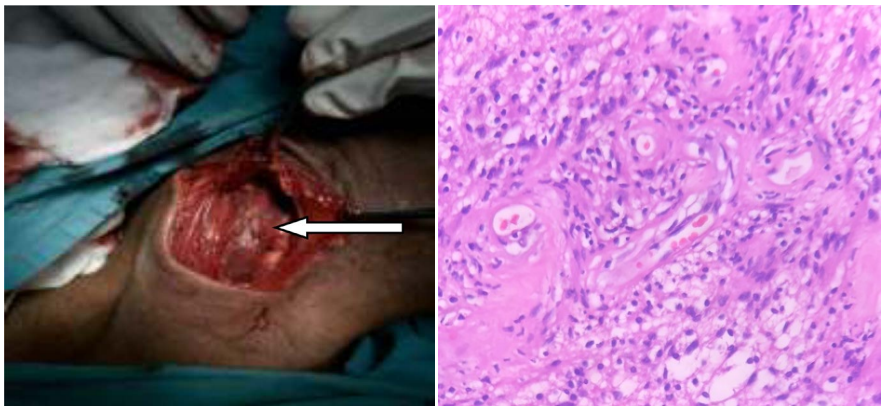


Figure 2. CECT showing the mass displacing the carotid.

2.2. Case 2

A 80 year old gentleman came with complaints of pain in the right thigh and leg for 2 months. He had history of difficulty in defecation and per rectal examination showed a extra-mucosal mass. A screening ultrasound showed a 8 × 7 × 8 cm hypoechoic lesion in the presacral region and CECT showed a large well defined heterogeneously enhancing lesion in the pre-sacral region with preserved fat plane with the adjacent structures. The lesion extended to the anterior sacral foramen and was suggestive of a neurogenic benign lesion. MRI showed a heterogeneous presacral lesion with internal hypo intense focus (likely cystic) in the pre-sacral (S1 - S4) with postero-inferior part in close contact with exiting nerve root (**Figure 4**). At laparotomy a 10 × 7 × 7 cm mass was found in the retro-peritoneum. The retro-peritoneum was entered and the vessels and ureters were identified. The mass was arising from the nerves of hypogastric plexus. The tumor was excised (**Figure 5(b)**), the troublesome pre-sacral ooze was controlled and the specimen sent for HPE.

Biopsy showed hypo-cellular areas with spindle cells having wavy nuclei and scanty eosinophilic cytoplasm. Hypercellular areas showed spindle cell with wavy



(a) (b)

Figure 3. (a) IJV splayed over the cystic lesion (Intraoperative); (b) Histopathology showing schwannoma (10× resolution).

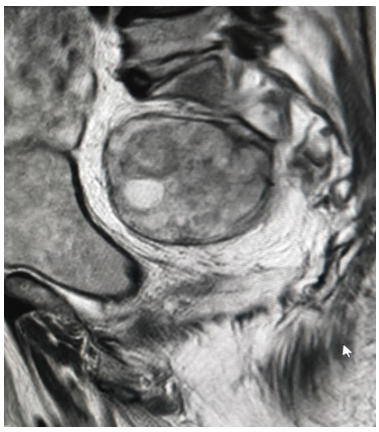
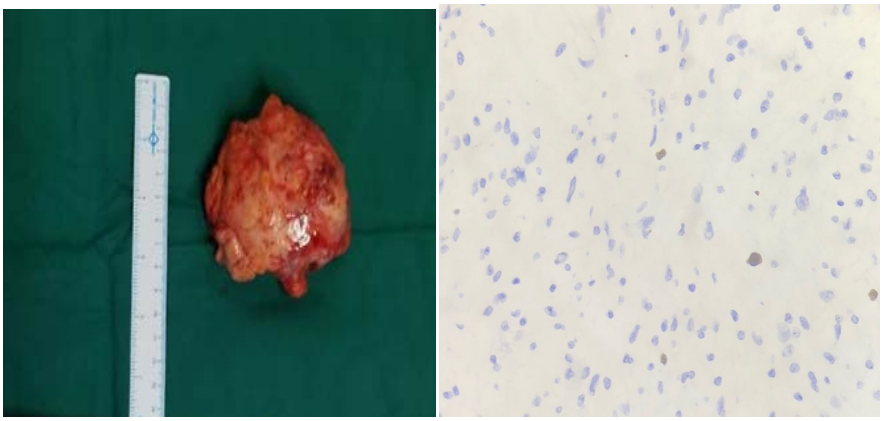


Figure 4. MRI showing presaccral mass.



(a) (b)

Figure 5. (a) Excised presaccral mass; (b) Slide shows IHC.

nuclei with formation of Verrocay bodies. Areas of focal foreign body giant cell reaction, foamy cells and dilated congested vessels were seen. Few vessels show myxoid change in the wall-consistent with schwannoma with secondary changes. IHC was positive for S-100 (**Figure 5(b)**).

Both patients were free of symptoms at the 2 month follow up and showed no residue/recurrence at imaging. Consent was taken from the patients preoperatively for use for research purposes.

3. Discussion

Schwannoma is generally defined as benign tumor of neural cells derived from nerve sheath composed of schwann cells [1]. Common sites include the head and neck, the flexor surfaces of the extremities, and the para vertebral area of retro-peritoneum. The nerve of origin is not often made until the time of surgery. Schwannoma originating from the vagus nerve (cervical), is very rare to occur in men between the 3rd and 6th decades of life. The most common presentation is a painless, slow-growing, lateral neck mass this appears in a large proportion of cases [2]. Rapidly growing tumors with either evidence of invasion or presenting with complete loss of nerve function should be treated by complete excision of the tumor.

About 25% - 33% of all the extracranial schwannomas occur in the head and neck. Chandramohan has described a case of vagal schwannoma, in which tumor was excised by intra capsular dissection thus sparing left vagus nerve [3]. Saini et al has reported intra operative bradycardia during dissection of the mass from its adjacent structure [4]. Samarakoon et al had described a 46-year-old man who was evaluated for chronic constipation due to a giant pre-sacral schwannoma (diagnosed on magnetic resonance imaging scan). Pre-operatively they came to diagnosis of schwannoma by trans-rectal ultrasound scan (TRUS)-guided biopsy and further proceeded with excision of mass [5]. A 32-year-old man with a painless right neck mass presented with dysphagia for six months. MRI is the gold standard to assess the origin and the extent of the tumor [6].

Histologically the cut surface appears solid, smooth, glistening and gray-white. It may show cystic and hemorrhagic areas with calcification. Histo-morphology of schwannoma characteristically shows two alternating patterns—Antoni A and Antoni B areas. Antoni A are cellular areas with compactly arranged spindle cells frequently arranged in interlacing fascicles, palisades, or in an organoid arrangement, with two compact parallel rows of well-aligned nuclei forming eosinophilic structures (Verocay bodies). Antoni B exhibits hypocellular areas consisting of a few tumor cells in loose myxomatousmatrix [7].

S100 is a marker for neural tumors and is extensively studied in schwannomas [8]. In a study Fausto J. Rodriguez has mentioned all neurogenic tumors are always strongly GFAPpositive, suggesting cross reactivity of cytokeratin antibodies with GFAP, rather than true protein expression. By immunohistochemistry, schwannomas typically show diffuse, strong expression of S100 protein and abundant peri-cellular collagen type IV, consistent with the presence of a continuous peri-cellular basal lamina [9]. Glial fibrillary acid protein (GFAP) is expressed in a subset of schwannomas. Recent markers frequently positive in schwannomas include, calretinin, SOX10 and podoplanin.

In a study conducted in Kyushu University in Japan all cervical tumors were resected through a trans-cervical approach. The nerve of origin was mainly determined by the postoperative neurological findings complete tumor resection was performed in 11 patients, and intra-capsular enucleation of the tumor was performed in 16 patients. Intra-capsular enucleation was an effective and feasible method for preserving the neurological functions [10].

Makni has said various techniques for excision of pre-sacral mass biopsy should be done before deciding either to start on neo-adjuvant treatment or excision [11].

4. Conclusions

As slow growing tumors of the head and neck, schwannomas very rarely presents as potentially morbid lesions. Although it is rare, clinicians should not forget the possibility of a nerve sheath tumor in presence of a neck mass. The preoperative bedside diagnosis is dependent on clinical suspicion and imaging modalities. Complete resection of the tumor is the treatment of choice for all benign schwannomas.

Schwannomas arising from vagus nerve cause bradycardia and the anaesthetist must be vigilant during surgical excision. Pelvic schwannomas may present with constipation. We present our cases for their challenges and uncommon presentations.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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