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Case Report

Schwannoma of Accessory Nerve – A Differential of Neck Swellings

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Abstract

Introduction: Intracranial schwannomas comprise approximately 8% of primary intracranial tumours, and more than 90% of intracranial schwannomas originate from vestibular nerves. Schwannomas originating from non-vestibular cranial nerves are very rare, particularly arising from accessory nerve. Accessory nerve schwannomas constitute only 6.66% of all non-vestibular intracranial schwannomas. The most recent substantial review of literature reports only 44 total cases of accessory nerve schwannoma.

Case Report: We report a rare case of right accessory nerve schwannoma in a young male (genetic predisposition-unknown), not associated with neural deficit, presented with visible neck swelling and discomfort, assessed clinically and evaluated by CT scan, managed by surgical resection without residual weakness of muscle, diagnosis confirmed by histopathological examination.

Discussion: Schwannomas, arising from accessory nerve, are relatively uncommon, and only a few of cases have been reported so far. Patients with accessory nerve schwannoma locating in extracranial site present with a gradually increasing neck mass that draw less attention. It is challenging for surgeons on how to evaluate extracranial/cervical spinal accessory nerve schwannomas by radiography. However, it is insensitive to make a definite diagnosis for schwannoma, and to identify the nerve origins of tumours. Enhanced CT scans can help us to diagnose schwannomas.

Conclusion: Though a rare diagnosis, peripheral nerve Schwannoma should be included in differential diagnosis of neck swellings having ruled out other conditions, easily evaluated by CT/MRI and successfully managed by surgical resection.

Keywords: sleep disorders; restlessness; sleep interruption of activity; type of encephalitis; circadian rhythm; restive poles disease; polysomnography; cognitive-behavior therapy; CPAP; sleep well-being

Introduction

Intracranial schwannomas comprise approximately 8% of primary intracranial tumours, and more than 90% of intracranial schwannomas originate from vestibular nerves[1]. Schwannomas originating from non vestibular cranial nerves are very rare[2], particularly arising from accessory nerve[3]. Accessory nerve schwannomas constitute only 6.66% of all non-vestibular intracranial schwannomas[2]. The most recent substantial review of literature reports only 44 total cases of accessory nerve schwannoma[4].

Case Report

A 27year/male, ECOG-0, cigarette smoker and occasional alcohol drinker, BMI 22.03kg/m2, having no significant family history, resident of Nashik, presented to General Surgery OPD with chief complaint of visible swelling on right side of neck since past 18 months associated with discomfort, starting smaller in size when noticed and gradually increased to present size of 4X4 cms not having any signs of inflammation and having no constitutional symptoms of tuberculosis or malignancy. Having skin tattoo over the same site can be a mere coincidence as it was done much earlier. On clinical examination the swelling was a single, limited to right carotid triangle in retro muscular (Right SCM) plane, 4X4 cms, globular, well defined margins and regular surface, tense cystic consistency, transmitted pulsations present without expansile pulsations, non tender, non mobile in vertical and having limited mobility in horizontal planes. Both sides trapezius and SCM were equal in tone and

power. Patient evaluated by preliminary investigation like Ultrasound and FNA Cytology suggestive of Spindle Cell Tumour having isoechoic picture, situated in upper part of right side of neck posterior to right SCM medial to internal jugular vein and lateral to right carotid arteries. Findings were consistent on CT scan neck with isodense 4X4cms mass in right upper neck having hypo-dense areas (of necrosis) related to internal jugular vein and carotid arteries as described. Though MRI is advisable in supra- hyoid swellings[5], CT was done due to financial constraint. Surgical resection was done uneventfully under General Anaesthesia without postoperative right trapezius and right SCM muscle weakness or vocal paralysis. Diagnosis of schwannoma was confirmed on Histopathological examination described as a well circumscribed tumour with compact hyper cellular (Antoni A) and myxoid hypo cellular (Antoni B). The hyper cellular show nuclear palisading around fibrillary processes (Verocay bodies). The hypo cellular areas are myxoid and show thick walled blood vessels. Cells are elongated, narrow, wavy, with tapered edges. Tumour cells have ill defined cytoplasm and wavy nuclei. There are few collagen fibres and thick walled blood vessels. No atypia/malignancy in the material studied. Immuno-histo-chemical analysis for positivity for S-100 [6] was not done due to unavailability at our hospital and financial constraint.

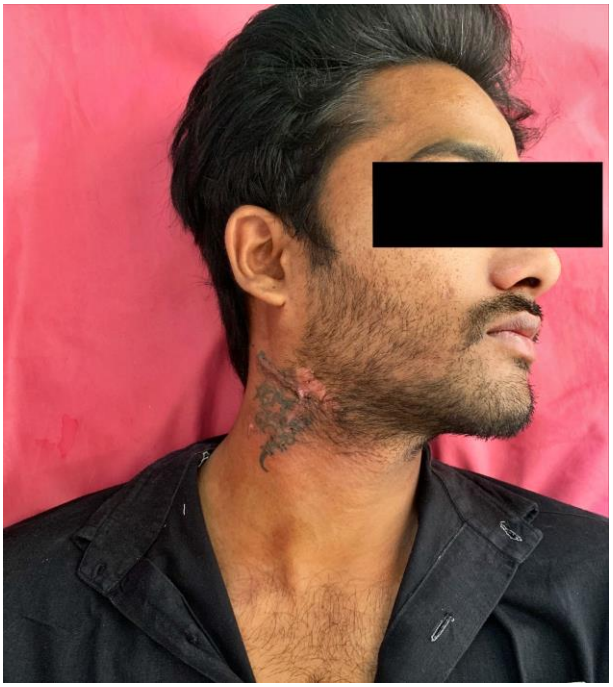

Figure 1: Clinical picture.

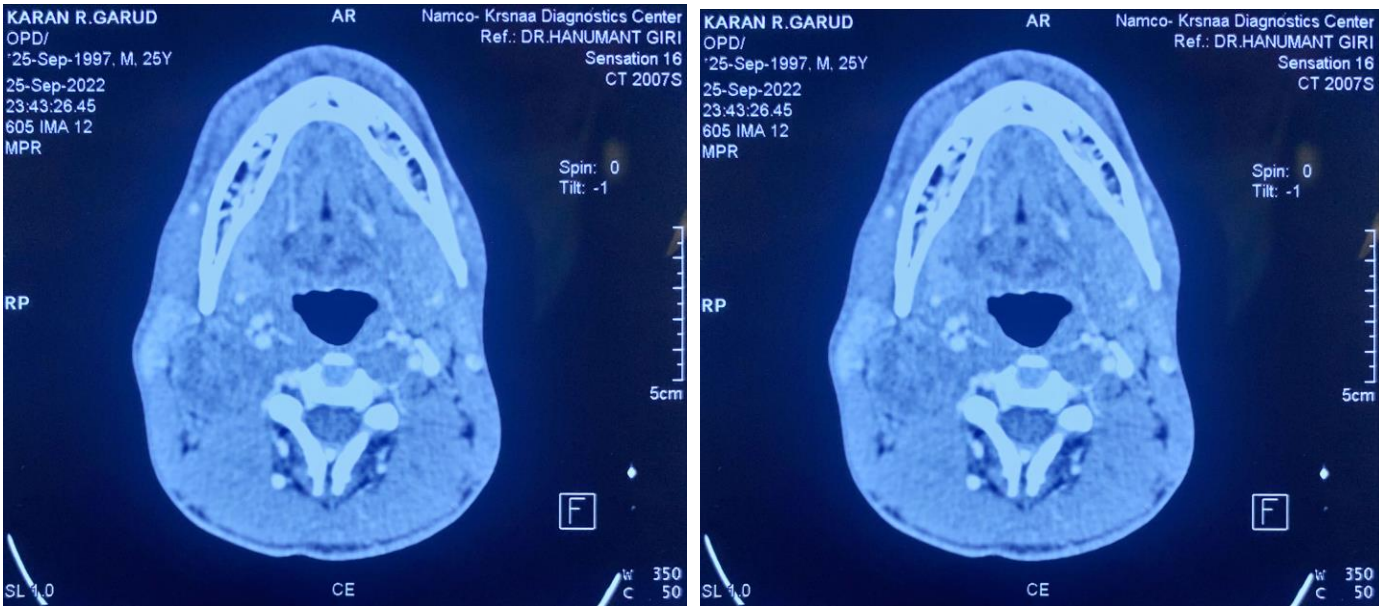

Figure 2: CT images depicting the mass with hypodense areas.

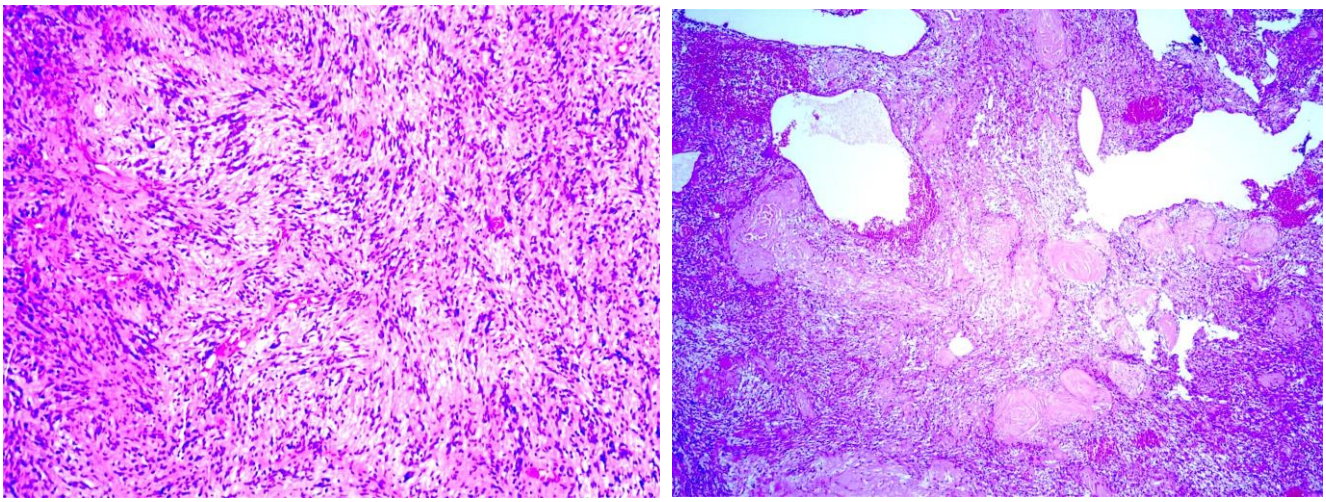

Figure 3: Histopathology images showing Antoni A and Antoni B pattern with Verocay bodies.

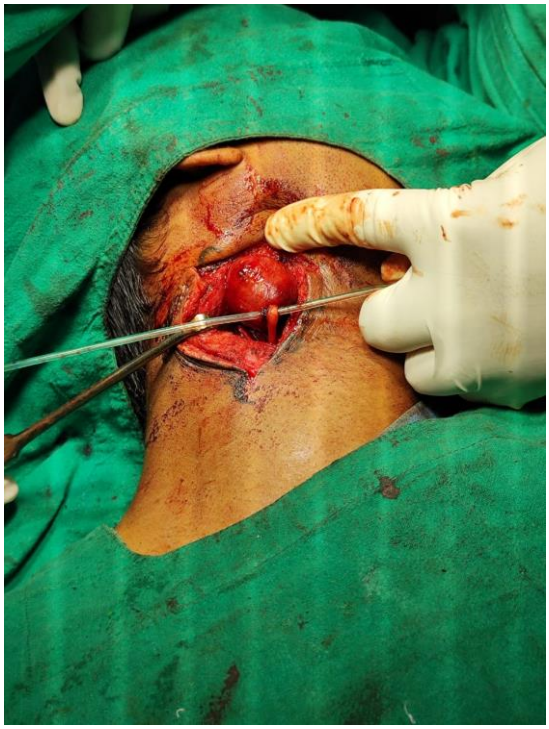

Figure 4: Intraoperative image showing Schwanomma (pointed by the finger) over Accessory nerve (structure over the probe)

Discussion

A schwannoma is a relatively slow growing tumour of the nerve sheath, which is composed of the schwann cells, which normally produce the insulating myelin sheath which covers the peripheral nerves. They are homogenous tumours and they always stay on the outside of the nerve and may push the nerve aside or against the bony structure[7]. The schwannomas which occur in the head and neck region mostly originate from the vagus nerve or from the sympathetic nervous system[8]. Schwannomas, arising from accessory nerve, are relatively uncommon, and only a few of cases during operation have been reported so far. The accessory nerve consists of a cisternal,a foramen jugular, and an extracranial section. Tumors can grow from anywhere along the path of this nerve. It is challenging for surgeons how to evaluate extracranial/cervical spinal accessory nerve schwannomas by radiography[9]. However, it is insensitive to make a definite diagnosis for schwannoma, and to identify the nerve origins of tumors[10]. Enhanced CT (ECT)scans can help us to diagnose of schwannomas[11].

The diagnosis of schwannoma is confirmed by combining clinical suspicious, observation in surgery and pathologic evaluation. Accessory nerve schwannomas are histologically classified as two distinct alternating structure types, named as Antoni A and Antoni B. Antoni A tissue is characterized by compactly arrayed spindle cells, which have long oval nuclei with their long axes being parallel to each other. Antoni B tissue consists of spindle cells arranged by a loose way without forming a distinct pattern. Although an entire tumor may be composed of either type of tissue, both types are more commonly intermixed[12]. S-100 protein, a neural create marker antigen, is specific as a marker of schwannomas, broadly used as immunohistochemical marker[13]. A gross total resection remains the treatment of choice for these tumours, because they are radio resistant[14]. The other treatment option is sterotactic radiosurgery (SRS). SRS is gaining popularity, but it depends on many factors, which include the size and the location of the Schwanoma and the age of the patient. Whenever it is possible, SRS should be avoided in young patients[15].

Conclusion

Though a rare diagnosis, peripheral nerve Schwannoma should be included in differential diagnosis of neck swellings have ruled out other conditions, easily evaluated by CT/MRI and successfully managed by surgical resection

Conflicts of Interest: None

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