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International Journal of Histopathological Interpretation

Case Report

ANCIENT SCHWANNOMA MASQUERADING AS CAROTID BODY TUMOR-A RARE CASE REPORT WITH REVIEW OF LITERATURE

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How to cite: *Gowthami N et al, Ancient schwannoma masquerading as carotid body tumor-a rare case report with review of literature 2023; 12(1):27-33. DOI:* ***https://doi.org/10.56501/intjhistopatholinterpret.v12i1.829***

Received :09/04/2023 *Accepted: 20/04/2023* *Web Published: 05/05/2023*

Abstract

The most common tumor at carotid artery bifurcation is a carotid body tumor causing widening of bifurcation, usually seen on conventional arteriography that can also be produced by schwannoma of the cervical sympathetic chain. Ancient schwannoma is the uncommon variety of schwannoma, a benign tumor that arises from the nerve sheath. It usually arises from any nerve except the optic and olfactory. Here we present a rare case of a male in his early 50s with lateral neck swelling diagnosed radiologically and excised as a carotid body tumor diagnosed as ancient schwannoma on histopathological examination. Although quite rare, cervical sympathetic chain schwannoma should be considered a differential diagnosis for tumors at the carotid bifurcation. Insight into morphological features of ancient schwannoma avoids misdiagnosis as a malignant nerve sheath tumor.

Keywords: Ancient schwannoma, cervical sympathetic chain, carotid body tumor.

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INTRODUCTION

Schwannomas also known as neurilemmomas; neuromas are uncommon benign nerve sheath neoplasms. (1,2) Ancient schwannoma describes the group of neural tumors showing degenerative changes, diffuse hypocellular areas, nuclear hyperchromatic, and moderate to marked nuclear atypia. (2) Almost 25% to 45% of head and neck schwannomas pose diagnostic and management challenges. (1) Schwannomas arising from the cervical sympathetic chain are rare, and approximately 57 cases have been reported. (4) Splaying of carotid bifurcation on radiology is due to some lesion at that level and is called Lyre's sign. It is a definitive sign of a carotid body tumor. A schwannoma arising from the cervical sympathetic chain also produces similar signs. (3) Here we present a case of ancient schwannoma of the cervical sympathetic chain that was misdiagnosed and excised as a carotid body tumor preoperatively, with a brief review of the literature.

CASE REPORT:

A male patient in his early 50s presented to surgical OPD with a history of swelling over the right side of the neck for 2-3 years. There was no history of tooth infection, sore throat, oral ulcer, or fever. The patient had no difficulty chewing and there was no change in facial appearance. Systemic examination was within normal limits. Local examination showed a 5x3 cm swelling over the right side of the neck. USG neck which revealed a large well defined heterogeneous predominantly isoechoic solid lesion with few small cystic areas with mild tapering of the lower end involving the upper part of the right side of the neck. Subsequent MRI and Contrast-enhanced MRI showed a well-defined longitudinally elongated altered signal intensity lesion noted in the right carotid space which appears heterogeneous predominantly hyperintense on T2 and PD sequences showing heterogenous post-contrast enhancement with extension. (Figure-1) Fine needle aspiration cytology (FNAC) revealed a benign spindle cell lesion in favor of a peripheral nerve sheath tumor(figure-2). The patient underwent upper cervical mass excision and the specimen was sent to histopathology for further eva

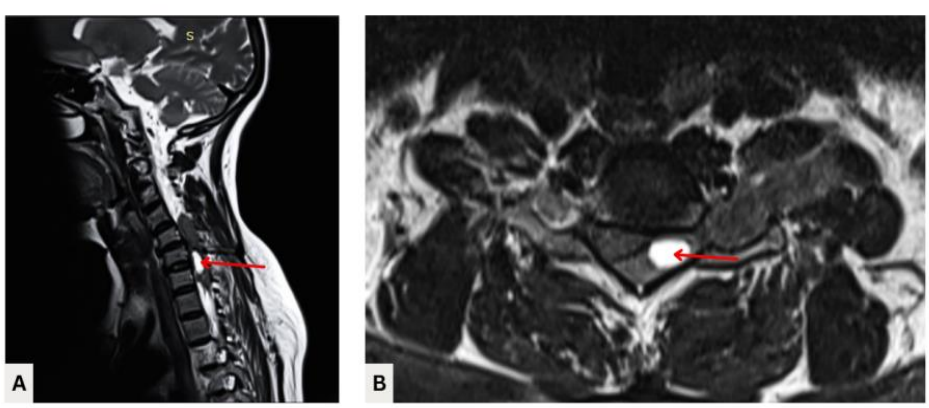

Figure-1: A&B-Sagittal and axial view of 12 weighed MRI showing a well circumscribed hyperintense mass (arrow) impinging on adjacent structures.

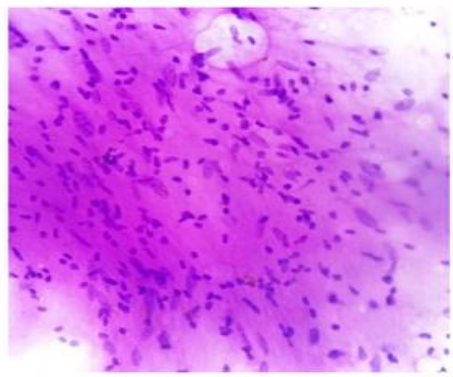

Figure-2: Fine needle aspiration cytology showing benign spindle cells (MGG-40X)

Grossly a globular whitish firm tissue piece measuring 4.5x 3x 1.5 cm was received. The Cut section showed homogenous yellowish-white areas with specs of hemorrhage. One complete cross-section was taken and the tissue was processed for examination(figure-3). Microscopy showed a collection of parasympathetic ganglia and adjacent to it was a well-circumscribed partially encapsulated growth showing moderately cellular to hypocellular areas comprising of spindle cells in nodules and broad fascicles showing bland to atypical nuclear features, smudged, hyperchromatic and clubbed nuclei. There were foci of myxoid degenerative changes and hyalinization. Mitoses were infrequent. Tumor necrosis was not seen in the sections examined(figure-4). A Panel of Immunohistochemistry including SOX10, S100, CD31, CD34, SMA, Desmin, and HMB45 was done among which SOX10 showed diffuse nuclear positivity(figure-5); S100 showed strong cytoplasmic and nuclear positivity; Ki67 for the proliferative index was between 1-2%(figure-6). Rest IHCs were negative. So based on histopathology and IHCs the diagnosis of benign nerve sheath tumor- Ancient Schwannoma was rendered.

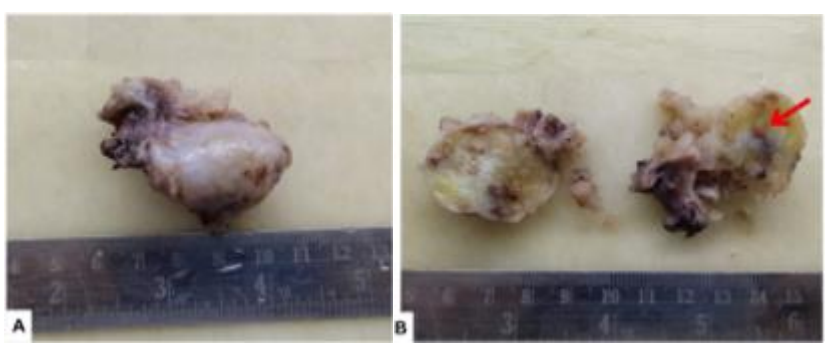

Figure-3: A-Gross: Globular whitish firm mass; B-Cut section showing homogenous yellowish white areas along with myxoid and microcystic areas (arrow).

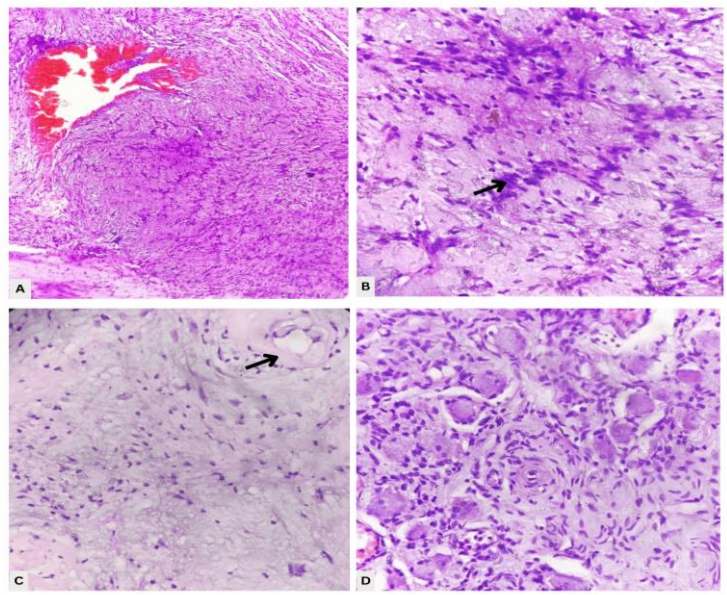

Figure-4: Microscopy-Moderately cellular areas of spindle cells with Clubbed nuclei showing palisading-Verocay body(arrow) and myxoid degenerative changes(A-H&E:10X&B-H&E:40X) Areas showing hyalinization of blood vessels (arrow), and myxoid changes (C-H&E:40X). Normal unaffected ganglion cells of carvical sympathetic Chain attached to the tumor(D-H&E-40X)

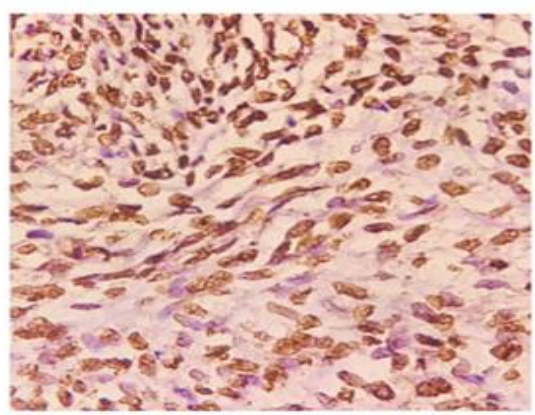

Figure-5: SOX1O showing diffuse nuclear positivity of tumor cells (IHC-40X)

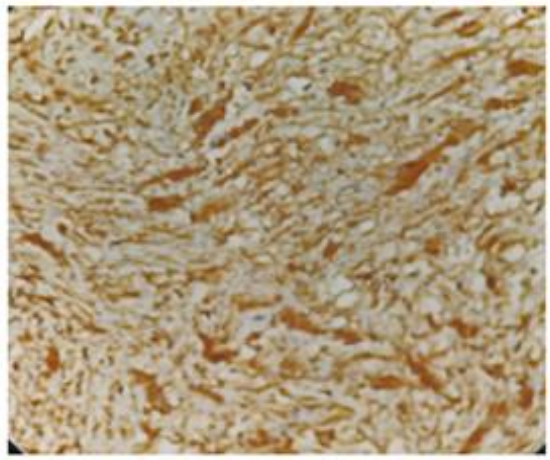

Figure-6: 5100 showing cytoplasmic and nuclear positivity of tumor cells (IHC-40X)

DISCUSSION:

Schwannomas are uncommon nerve sheath tumors that arise from Schwann cells of any peripheral, cranial, or autonomic nerves of the body except olfactory and optic nerves which lack Schwann cell sheath. (1,4) Extracranial Schwannomas arise in the head and neck accounting for 25-45%. (1,3) Generally, the neurogenic tumors of this region are slow growing asymptomatic. CT and MRI imaging are useful not only to determine tumor size and extension but also to clearly delineate the relative position of the tumor to its surrounding vasculature. This is less useful in distinguishing a carotid body tumor from a cervical sympathetic chain schwannoma since splaying of carotid at carotid bifurcation occurs in both the tumor. (1,3)

Microscopically, schwannomas have a distinct pattern of compact cellularity arranged with a palisading nucleus (Antoni A) alternating with a more loosely hypocellular pattern (Antoni B). Occasionally schwannomas may present with hyperchromatic areas and bizarre nuclei. These changes reflecting degeneration are typical of ancient schwannoma. (2,4) Average age of occurrence of schwannomas is between 30 to 60 years of age. Schwannomas of the cervical sympathetic chain is an extremely rare and usually present as para-pharyngeal masses. (3,4)

Complete surgical excision of the tumor is the treatment of choice. (3,4) During the excision of the tumor efforts for the preservation of the nerve should be made although it is difficult because of the dense attachment to the tumor. Incomplete excision of the tumor may result in slow recurrence over months to years. Recurrence and malignant transformation of ancient schwannomas are very rare. (4) Diagnosis is confirmed after histopathological examination showing two distinct areas. Tumor cells are usually positive for SOX10 and S100 antigens in the immunohistochemical examination. (1-4)

The differential diagnosis for neck schwannomas is carotid body tumor, neurofibroma, and benign fibrous histiocytoma. Carotid body tumor arises from chemoreceptor cells of the carotid bulb. Microscopically, a tumor is composed of nests of fairly uniform epithelioid cells with granular eosinophilic cytoplasm, and a round nucleus surrounded by vascular stroma. IHC shows positivity for Chromogranin, Synaptophysin, S100,

and GATA3. (5) Neurofibromas are benign nerve sheath tumors with a tan-white, glistening cut surface apparent grossly. Their growth pattern is either well-demarcated intraneural or diffuse infiltration of soft tissue at extraneural sites. Histologically, Schwann cells with wire-like collagen fibrils, stromal mucosubstances, mast cells, Wagner Meissner corpuscles, Pacinian corpuscles, fibroblasts, and collagen. No verocay bodies, and no nuclear palisading are seen. IHC shows positivity for S100, SOX10, and CD34.(6)

Benign fibrous histiocytoma shows short spindle and ovoid cells with slightly irregularly shaped nuclei arranged in a storiform fascicular pattern along with an admixture of inflammatory cells, and foamy macrophages. Entrapped thickened, hyaline collagen bundles are seen at the peripheries of the lesion. Immunohistochemically, the tumor cells are often positive for factor XIIIa, SMA, CD68, desmin, and CD34 are often expressed and negative for SOX10 and S100. (7)

In 2011, Sathish et al reported a case of neck schwannoma in a 50-year-old female mimicking a carotid body tumor. (1) Styajith et al in 2015 reported a case of ansa cervicalis, but the origin of schwannoma from ansa cervicalis was confirmed intraoperatively by contraction of strap muscles. (2) Ancient schwannoma of the cervical sympathetic chain is a rare disease and only a few cases have been reported. In 2016, Primuharsa et al reported a case of ancient schwannoma of the cervical sympathetic chain that presented as a mass in the right parapharyngeal space. (4)

CONCLUSION:

Clinically ancient schwannoma is often mistaken as tumors of the thyroid, enlarged lymph nodes, paraganglioma/ carotid body tumor or branchial cyst. Moreover, preoperative diagnosis of ancient schwannoma is difficult. Thus, we intend to alert the reader to consider the rare scenario in investigating a lateral neck mass by considering all possible differential diagnoses. So, surgeons should consider that schwannomas of the cervical region can arise from any nerve and can be misdiagnosed as carotid body tumors. It is necessary to preoperatively rule out carotid body tumors from ancient schwannoma as the management of both cases varies.

Funding

None

Acknowledgements

Not applicable

Competing interests

None

Contributorship statement

GN, NA, MA and VP have equally contributed towards the preparation of the manuscript.

Consent of patient- Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review

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