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A rare case of cervical nerve root C2, C3, C4 ancient schwannomas

¹Pavan T. Lanjewar, Junior Resident, Department of General Surgery, Government Medical College, Aurangabad-431001.
²Anitha Kandi, Associate Professor, Department of General Surgery, Government Medical College, Aurangabad-431001.
³Sarojini Jadhav, Professor and Head, Department of General Surgery, Government Medical College, Aurangabad-431001.
⁴Sachin Jangle, Lecturer (Plastic surgeon), Department of General Surgery, Government Medical College, Aurangabad-431001.

Corresponding Author: Pavan T. Lanjewar, Junior Resident, Department of General Surgery, Government Medical College, Aurangabad-431001.

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Abstract

Introduction: Schwannoma is benign а mesenchymatous tumor developing exclusively from the cells of the Schwann sheath of peripheral nervous system nerve fibers. Extracranial schwannomas in the head and neck region are rare neoplasms occurring in 25-45% cases, and further those arising from nerve root are extremely rare. Diagnosis is established by imaging studies such as Magnetic Resonance Imaging (MRI) or Computed Tomography (CT) and Fine needle aspiration cytology (FNAC), while histopathology gives definitive diagnosis. The management of these tumors is surgical resection with preservation of the neural pathway. Ancient schwannoma is histopathological finding showing relative loss of Antoni A areas with irregular nuclei and areas of hyalinization with hyperchromatism suggestive of degenerative changes.

Case report: The present is a rare case of cervical (C2, C3, C4) nerve root schwannoma in a 70 year old lady who presented with left lateral neck swelling with associated intermittent dull aching local pain managed

by enucleation with histopathology report suggestive of ancient schwannomas.

Conclusion: Extracranial schwannomas in the head and neck region, especially arising from nerve roots are rare neoplasm, diagnosed with the help of imaging techniques like CT, MRI and definite diagnosis by histopathology. The accepted treatment for these tumors is surgical resection with preservation of the neurological function.

Keywords: Schwannoma, nerve root, ancient.

Introduction

Schwannomas also known as neurilemmomas or neurinomas are benign nerve sheath tumors deriving from Schwann cells that occur in the head and neck region in 25-45% of cases (1). Extracranial schwannomas in the head and neck region are rare neoplasms (2). About 10% of schwannoma that occur in the head and neck region generally originate from the vagus or sympathetic nervous system whereas those arising from nerve root are extremely rare, as in this case report (3). Preoperative imaging studies such as MRI and CT are

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used to distinguish its location and origin. (4). FNAC aids in diagnosis. The treatment of schwannoma is primarily surgical resection with preservation of neurological function. The histopathology in ancient schwannoma, as in the case we report, shows relative loss of Antoni A areas with irregular nuclei and areas of hyalinization with hyperchromatism suggestive of degenerative changes (5).

Case report

A 70 year old lady presented with chief complaints of slow growing swelling over left side of neck since 6 years with intermittent dull aching pain at the same site. There was no past history of trauma or any surgical intervention. On examination, partially mobile firm lobulated non-pulsatile swelling of size 6 x 3x 1cm over left side of neck posterior to sternocleidomastoid muscle was present, without associated skin lesion, discharge or local signs of inflammation. Patient had no associated difficulty in breathing or deglutition or mobility of neck. Per oral examination was normal. Cervical spine examination showed no abnormalities with no neurological deficit. Patient was investigated with contrast CT scan, showing evidence of three mildly enhancing lobulated soft tissue density masses within left posterior triangle of neck with perineural spread with lesions extending approximately till neural foraminae of left C2, C3, C4 vertebra suggestive of nerve sheath tumor most probably schwannomas (Figure 1). FNAC was performed suggestive of schwannoma. Based on the clinical history, imaging and FNAC, a diagnosis of left C2, C3, C4 nerve root schwannoma was considered. Patient was operated by enucleation of tumors in following steps: Left neck transverse skin crease incision taken over the most bulging part of the swelling, incision deepened, sternocleidomastoid muscle retracted and underlying three encapsulated tumors with accompanying

nerves identified. All three tumors were dissected from surrounding structures and excised with associated nerves spared. (Figure 2,3). Surgery was uneventful and patient tolerated the procedure well with no postoperative complications. Histopathology report of all three tumors was benign spindle cell lesion suggestive of ancient schwannomas (Figure 4). Patient was discharged uneventfully. Patient is followed up at 6 months after the procedure with no evidence of any recurrence.

Figures



Figure 1: Computed tomography scan with contrast image suggesting left cervical schwannoma tumors



Figure 2: A. Intraoperative evidence of two cervical schwannoma tumors, B. Intraoperative evidence of third cervical schwannoma tumor with associated cervical nerve (after two schwannoma tumors as in Part A of image removed)



Figure 3: Specimen of excised three schwannoma tumors (A: One encapsulated tumor and B, C: Another two tumors showing substance of tumor)



Figure 4: Histopathological view of excised tumours suggestive of Ancient Schwannomas

Discussion

Schwannomas are benign, slow-growing, solitary neoplasms of Schwann cell origin. Most of the schwannomas arise in the cranial vault in the cerebellopontine angle (acoustic neuroma). About 25% to 45% of extracranial tumors arise in the head and neck area, in any point of the neuron axon from the skull base or spinal column down to skin, mucosal or end-organ structures (6). Schwannomas arising from nerve roots are extremely rare, as is our case from C2, C3, C4 nerve

roots (2). Schwannomas arising from particular cranial nerve or plexus may present with the corresponding peculiar symptoms related to the nerves like vestibular with tinnitus, vagal with dysphagia or hoarseness, cervical sympathetic chain with Horner's syndrome, etc. However, in most cases, there are no or vague symptoms, thus there is difficulty to identify the neurological origin based on the physical examination (7). Imaging modalities like USG, CT and MRI are helpful in identifying the tumor and its correlations with surrounding vascular structures, muscles and nerves. FNAC aids in diagnosis, however the diagnostic accuracy of FNAC depends strongly on the specimen quality and the experience of the cytopathologist (3). For surgical treatment of schwannomas, various techniques with their neurological function preservation rates are as: excision plus nerve reconstruction (9%), enucleation with nerve sparing (31%), emptied or shelled-out tumors (71%), etc. (8). Our case is operated by enucleation with preservation of nerve fibres with no postoperative loss of any neurological function. There are two histological types of schwannomas i.e. Type A Antoni and type B Antoni. Type A Antoni are characterized by centrifugally compacted arrangements of spindle cells and Verocay bodies while type B Antoni are characterized by the lack of tissue cellularity and myxoid with loosely arranged spindle cells (9). The histopathological features of ancient schwannoma are relative loss of Antoni A areas with irregular nuclei and areas of hyalinization with hyperchromatism suggestive of degenerative changes, as found in this case report (10).

Conclusion: Extracranial schwannomas in the head and neck region, especially arising from nerve roots are rare neoplasm managed by excision with preservation of the neurological function and definitive diagnosis being established by histopathology.

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Abbreviations

CT: Computed tomography

MRI: Magnetic Resonance Imaging

FNAC: Fine needle aspiration cytology

USG: Ultrasonography

"C" in C2,C3.C4: Cervical

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