

Case Report and Literature Review –A Rare Case of Schwannoma of Accessory Nerve

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Abstract: A rare case of schwannoma originating from cervical portion of the accessory nerve is reported. The tumor was diagnosed by C.T. Scan and confirmed by Surgery. Tumor was small and located in left posterior triangle of neck without causing any detectable neurological deficit.

Keywords; Accessory nerve, Schwannoma

I. Introduction

Schwannomas are usually slow growing benign tumors arising from schwann cells. Vestibular schwannomas are the most common cranial nerve schwannomas followed by trigeminal and facial schwannomas and then the glossopharyngeal, vagus and spinal accessory nerve schwannomas. Schwannomas involving the oculomotor, trochlear, abducent and hypoglossal are rare. A review of literature revealed only 28 cases (1) of spinal accessory nerve schwannoma, 4 in the craniocervical region extending from C1 or C2 level to vermis (7,14,15), with only two located in cervical region (2,8). Here, we report a rare case of accessory nerve schwannoma.

II. Case Report

A 16 year old girl was admitted in the surgery department with complaint of painless lump on left side of neck for the last 1 year. It was slowly growing, soft, cystic, mobile, 6 × 5 cm oval mass palpable in the posterior triangle of neck. She was neurologically intact. On examination, neck muscles on the two sides were symmetric. In particular, muscle power, tone and trophism of trapezius and sternocleidomastoid muscle were similar on both sides. C.T. SCAN REPORT –Left sided posterior triangle mass---? Schwannoma ?? Mesenchymal tumor (Fig:1)

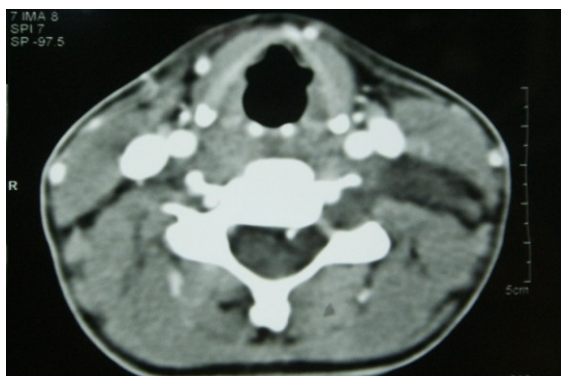


Fig :1 CT scan showing solid hypodense mass lesion with widening of neural foramen

We used the lateral cervical approach incision for resection. During surgery, the tumor was well circumscribed, encapsulated, yellowish white, shiny mass. The tumor was observed to be growing within the substance of the spinal accessory nerve, displacing uninvolved fascicles running between the leaves of capsule but not entering the tumor itself. There was extension of schwannoma into the neural canal but was easily separated with blunt dissection and mild traction to the tumor itself. The total excision of tumor along with entire capsule was done (Fig:2). Histopathologically, it showed typical schwannoma having both Antoni type A and Antoni type B cells. Postoperatively patient developed weakness of the trapezius muscle and patient was unable to abduct the left arm. Patient was advised physiotherapy and after one month of discharge, the patient was able to abduct her left arm and there was no other neurological deficit seen.



Fig:2 Showing preoperative extension of schwannoma into neural canal.

III. Discussion-

Schwannomas and neurofibromas comprise the group of neoplasms referred to as nerve sheath tumors. They are so designated because they are thought to arise from neuroectodermal elements of the nerve sheath, such as schwann cells, perineural fibroblasts or their more primitive precursors (4). Approximately 40% of neurogenic tumors occur in head and neck and most of these are benign schwannomas (4). Schwannomas are usually slowly growing benign tumors that arise from schwann cells (9). Of all schwannomas, 60% arise from the cranial nerves. Among cranial nerves, sensory cranial nerves are most frequently involved. Incidence of schwannomas arising from the trigeminal nerve, the facial nerve and lower cranial nerves are 40%, 23% and 20% respectively (5,6). The lower cranial nerves, including the glossopharyngeal nerve, the vagus and the accessory nerve form a complex at the jugular foramen. Therefore, a lower cranial nerve schwannomas at the jugular foramen is generally defined as a jugular foramen schwannoma without identification of the individual cranial nerve. On the other hand, the accessory nerve consists of two portions, the cranial root of the accessory nerve combines with the vagus nerve and it is difficult to recognize at this location. Based on anatomy, intracranial accessory nerve schwannoma are divided into two types by Julows (7) the jugular foramen type, which is the majority of the cases and the intracisternal type, which occupies the cisterna magna. In the case of a cervical schwannoma of the spinal accessory nerve, the symptoms consists of an 11th cranial nerve palsy and myelopathy i.e. trapezius atrophy (8). The spinal accessory nerve arises on a group of cells in the ventromedial part of the anterior horn of segments C1 to C4 (13). Clinical symptoms depend on the location and size of tumor. Preoperatively it is not possible to determine the exact nerve of origin without the aid of M.R.I.

IV. Histopathology-

Schwannomas consists of Schwann cells in a collagenous matrix. Histologically the term Antoni type A neurilemoma and Antoni type B neurilemoma are used to describe varying growth patterns in schwannomas. Antoni type A tissue has elongated spindle cells arranged in irregular streams and is compact in nature. Antoni type B tissues has a looser organization, often with a cystic spaces intermixed within the tissue (Fig:3).

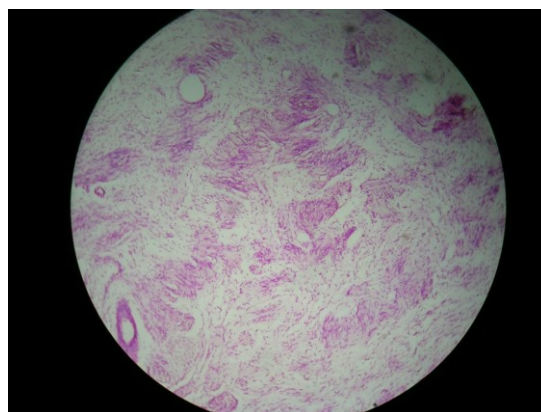


Fig:3 Histological section of schwannoma showing Antoni type A & type B cells.

Investigations-M.R.I. with the use of gadolinium based contrast medium is the technique of choice for imaging of cranial nerves. C.T. Scan is ideal for evaluating the secondary effects on the neural foramen. Ultrasonography may also be helpful in establishing whether the tumor lies between the jugular vein and carotid artery (11).

TREATMENT-There are two types of treatment options. Open surgery or stereotactic radiosurgery(SRS).The latter is gaining popularity but depends on many factors including the size and location of Schwannoma and the age of the patient. Whenever possible SRS should be avoided in young patients.

V. Conclusion-

These extremely rare slowly growing tumors were divided according to their location into intrajugular and intracisternal accessory nerve Schwannoma (7) by Julow. The brief anatomy of accessory nerve shows clearly that classifying Schwannoma according to their location may be more appropriate as follows (a) Intrajugular (b)Intracranial or intracisternal (c) Cervical(9).

The initial evaluation of the patient with a suspected cervical nerve sheath tumor should include history ,clinical assessment ,focusing on stigmata on a family history of neurocutaneous disease. A complete examination of head and neck should be performed, to rule out other neck masses. Gross total resection remains the treatment of choice for these tumors, because they are radioresistant (12) and tumor is confirmed on histopathologically by the presence of Antoni type A and type B cells.

Usually no postoperative dysfunction of the accessory cranial nerve occurs. The tumor growth can be compensated functionally by the rest of the intracranial nerve fibres because of the rootlets of the spinal part of accessory nerve originate from C1-C6 level and unite to form a common trunk. So, total tumor with sacrifice of the involved nerve is the primary treatment of schwannoma originating from cervical portion of spinal accessory nerve (14).

In our review of the literature we find only 28 cases of undoubted accessory nerve schwannoma of which only two were located in the cervical region (2,8) and we present an additional accessory nerve schwannoma of cervical region.

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