SCHWANNOMA OF NECK: A CASE REPORT AND REVIEW

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Abstract

Schwannoma, a benign, slow-growing neoplasm arises from Schwann cells of the nerve sheath of peripheral, cranial or autonomic nerves. Most Schwannomas are asymptomatic at the time of presentation, with a neck mass of variable size being the most common sign. Secondary symptoms like Horner's syndrome, hoarseness of voice, dysphagia depending upon the location of the tumour. FNAC, CT scan, MRI, USG help in preoperative diagnosis of the lesion. Surgical excision with preservation of nerve of origin (NOO) is the main treatment protocol. Post surgical histopathological examination establishes final diagnosis.

Key Words Schwannoma, neurilemmoma, nerve of origin (NOO), benign nerve cell tumour, surgical excision

INTRODUCTION

Schwannoma, also known as neurilemmoma, is a benign, solitary, encapsulated, and peripheral nerve tumour that arises from the perineural Schwann cells. Approximately 25–45 % of all schwannoma variants occur in the head and neck, usually presenting in the fourth decade of age [1]. Schwannomas can occur along the pathway of any somatic or sympathetic nerve, with the exception of the olfactory and optic nerves [2]. Typically, these lesions are freely mobile, with a single attachment at the nerve of origin. The lesions are slow growing, and not commonly associated with pain or neurologic symptoms, except in the setting of the third form of neurofibromatosis termed "schwannomatosis" [3]. If pain or neurologic symptoms are present, they are usually associated with a mass effect.

The nerves most commonly involved in Schwannomas of the head and neck are the vagus and the cervical sympathetic chain [4]. The trigeminal nerve constitutes the second most frequent site for intracranial schwannoma occurrence, after the vestibular nerve [5, 6], literature on schwannoma is very scanty and thus forming the purpose of the article which is to analyse the clinic-radiological features, preoperative diagnosis and treatment outcome.

CASE REPORT

A 19years old male patient presented with painless, gradually increasing swelling in the right side of the neck for last 4-5 months without any other associated symptoms like dysphagia, dysphonia. There was no other significant positive history.

Local examination revealed a 3cm x 2cm firm, non tender swelling 1cm below the lower border of mandible of right side. Contrast CT showed an enhancing mass in the right carotid space projecting into adjacent oropharynx. MDCT angiogram of neck revealed a well defined hypodense heterogeneously enhancing soft tissue mass at the

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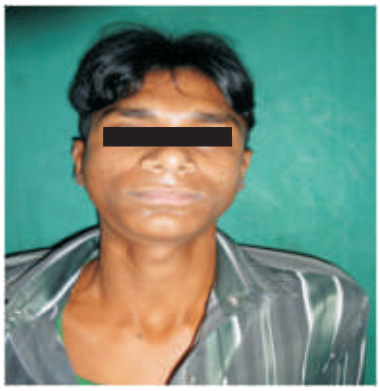
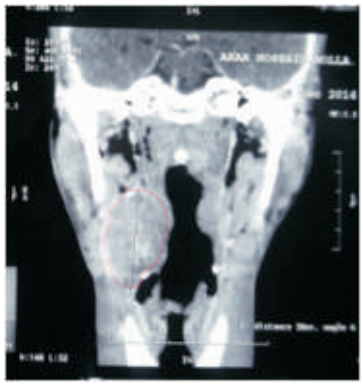
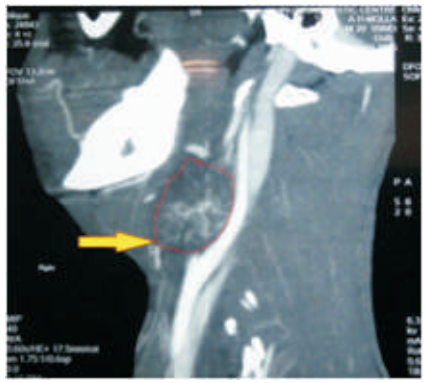


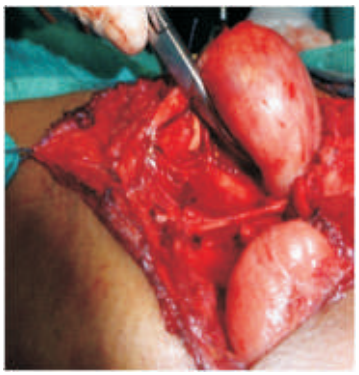
Preoperative photograph



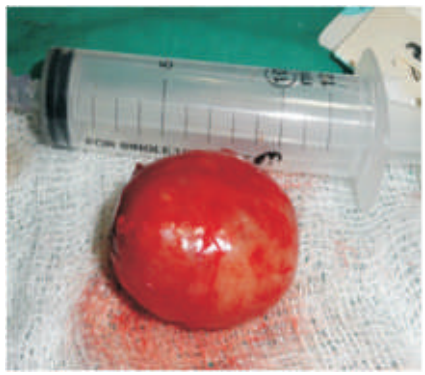
CT Scan showing the lesion



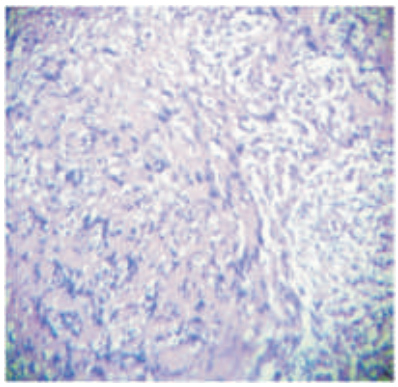
CT Angiogram showing tumour pushing ECA (Rt. side) outwards



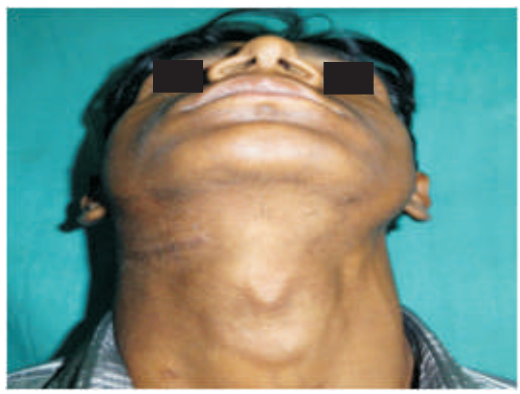
Intraoperative photograph of tumour



Excised tumour specimen



Histopathogical picture



1month follow up

level of carotid bifurcation with mild compression on right external carotid artery. USG guided FNAC finding was schwannoma or neurofibroma. Excision of the lesion was done and postoperative histopathology confirmed the diagnosis as schwannoma.

DISCUSSION

Verocay described a nerve sheath tumour in 1908 that he believed was derived from Schwann cells, which are responsible for the myelin sheath that surrounds and protects axons [7]. In 1910, he called it neurinoma. Later the term neurilemmoma was coined by Stout in 1935[8]. The Schwann cell is the parent cell of both the schwannoma and neurofibroma. However, there are significant differences between these tumours. In contrast, the schwannoma is encapsulated, rarely associated with Von Reklinghausen's disease very rarely undergoes malignant change (12% of neurofibromas do). Neurons do not traverse schwannomas, but do pass through neurofibromas.

Extra cranially, neurilemmomas are found most commonly in the neck, with the majority in the carotid triangle[9]. Two-thirds of the study population were female in line with a study by Torossian et al[10] observing female predominance in extracranial cephalic schwannomas, but at variance with Leu and

Chang's series [11] noting male predilection. More striking in the population was the preferential distribution of the tumour on the left side.

Tumors arising in the parapharyngeal space usually Present as an asymptomatic mass [12] until it becomes larger than 2.5 cm. Many of these neck masses were longstanding and have been manifested in the patient for as long as 3 to 9 years. Most schwannomas are asymptomatic at the time of presentation, with a neck mass of variable size being the most common sign. Upon palpation, they are slightly movable, except along the long axis of the nerve. Symptoms of damage of the various nerves, such as hoarseness or Horner's syndrome, may aid in the differentiation between schwannoma of the vagus nerve and schwannoma of the cervical sympathetic chain, but only a few patients show these symptoms before operation. Vagal schwannoma is typically characterized by dysphagia and hoarseness. Sympathetic schwannoma is characterized by Horner's syndrome. Dysfunction in the nerve of origin may not only result from axons being stretched over the tumour capsule, but also adjacent nerves may be affected by direct pressure.

Schwannomas almost always are diagnostic problems because the history and clinical examination are nonspecific and deceptive. Often the unilateral asymptomatic neck mass is diagnosed clinically incorrectly as an enlarged lymph node, a carotid body tumour, a brachial cyst, a thyroid cyst or nodule, even a parotid cyst or tumour in the case of parotid schwannomas. However, diagnostic modalities in the form of FNA cytological techniques as well as better imaging in the form of MRI or CT scans have lessened the problem of misdiagnosis to some degree [13,14] In addition to facilitating diagnosis; preoperative imaging provides information on tumour size, location, extent and surrounding anatomy, thereby aiding surgical planning.

The mainstay of treatment ideally remains complete surgical excision with preservation of the affected nerve. There was no recurrence for all the cases after complete surgical removal. Most authors have recommended careful intracapsular excision of the tumour to minimise postoperative neural deficit. Microneurosurgery to facilitate intraoperative microscopic diagnosis of the schwannoma and achieve more superior nerve preservation has been described. This involves microscopic enucleation of the tumour after opening epineurium without disrupting nerve continuity. The role of radio surgery as an alternative to surgical resection is a matter of ongoing debate.

The microscopic portrait of schwannoma is distinct and can hardly ever be confused with that of other lesions. In classical variant, the core of the tumour is composed of a mixture of two cellular patterns Antoni A and Antoni B [15–17]. Antoni A areas are collection of compact spindle cells with twisted nuclei arranged in bundles or fascicles. In extremely differentiated areas there may be nuclear palisading and formation of Verocay bodies, which are formed by arrangement of two rows of nuclei and cell processes which assume oval shape. Antoni B variant is less cellular and less organized, representing degenerated Antoni A areas composed of randomly arranged spindle or oval cells within myxoid, loosely textured, hypocellular matrix punctuated by microcyst, inflammatory cells and delicate collagen fibers [15–17].

A schwannoma does not recur if totally excised and the prognosis is usually outstanding. No recurrence was seen in any cases till date in the present study. Malignant transformation is rare [18]. These tumours are highly radioresistant and radiotherapy has no place in the therapeutic management [19].

CONCUSSION

Extracranial schwannoma are rare and most often present as asymptomatic masses. The preoperative diagnosis may be difficult. The definitive diagnosis relies on clinical suspicion and histopathological confirmation after operation. Local recurrence is rare. Complete surgical excision is the treatment of choice. The possibility of nerve injury should be kept in mind.

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