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Original Research Article

Analysis of surgical outcome and review of literature of schwannomas arising from the extremities

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ABSTRACT

Background: Schwannoma is a benign peripheral nerve sheath tumour derived from Schwann cells. Also known as Neurilemoma, it can affect any nerve in the body. They usually present as a painless swelling or paresthesia over the sensory distribution of the affected nerve. Although it is classically described that schwannomas are well encapsulated and can be completely enucleated during excision, many of them have fascicular involvement and could not be completely shelled out. The aim of this work is to present our experience in operative management of schwannomas located in extremities.
Methods: Authors conducted a retrospective review for 18 adult patients with schwannoma, from June 2012 to June 2018. There were 10 men and 8 women, ranging from 20 to 68 years of age, with a mean age of 46 years old. All patients had excision done for the tumour and histopathological examination confirmed schwannoma. All patients were preoperatively evaluated both clinically and radiologically. FNAC was also done to confirm the origin of the swelling.
Results: The mean follow up period has been 2 years. Complete excision with preservation of nerve was done in all cases except for one case in which nerve graft was used.
Conclusions: Use of preoperative MRI, magnification and good surgical technique will help to enucleate the tumour completely without any collateral damage or recurrence. The possibility and option of nerve graft should be discussed with patients prior to schwannoma excision, so that nerve grafting could be directly proceeded with patient consent in case there is fascicular involvement of tumour found intraoperatively.

Keywords: Nerve sheath tumours, Neurilemoma treatment, Schwannoma excision**,** Schwannoma

INTRODUCTION

Schwannoma or neurilemmoma is benign peripheral nerve sheath tumour which can affect any nerve in the body.[1] They were first described by Verocay in 1908.[2] They are the commonest tumour of peripheral nerves, although the incidence in adults is only 5%.[3] The growth of these tumour is slow, so they can remain as painless swellings for a few years before other symptoms appear. [4] There is a higher incidence in the flexor surface of the upper limb, since the concentration of nerve fibers is higher over that region.[5] Most of the schwannomas could be diagnosed clinically. Schwannomas are mobile in the longitudinal plane along the course of the involved nerve but not the transverse plane.[6] There is Tinel's sign (shooting paresthesia in the distribution of the involved nerve) upon percussion of the tumour if the affected nerve is a sensory nerve or a mixed nerve.[7] Magnetic resonance imaging (MRI) is a preferred imaging technique in the diagnosis of tumour of peripheral nervous system. On imaging scans, benign tumour of peripheral nerve presents as well-defined mass, usually

fusiform in shape located within a nerve, isointense to surrounding muscles on T1-weighted images, and hyperintense on T2- weighted images.[8-10]

With regard to histopathology, schwannomas contain varying proportion of two different areas.[11,12] Antoni type A areas are highly cellular and are composed of closely packed spindle cells which form a palisade and produce Verocay bodies. Antoni type B areas are composed of loosely arranged Schwann cells in a mucinous-like matrix.[12] One area is usually predominant over the other in every tumour.[5]

Treatment of choice for schwannomas is microsurgical resection.[4,5,13,14] Careful resection and preservation of the nerve is necessary. The aim of this work is to present this experience in operative management of schwannomas located in extremities.

METHODS

Authors conducted a retrospective review for 18 adult patients with schwannoma, from June 2012 to June 2018. There were 10 men and 8 women, ranging from 20 to 68 years of age, with a mean age of 46 years old. All patients had excision done for the tumour and histopathological examination confirmed schwannoma. 27.8% (5 patients) had involvement of sensory nerve, and 66% (12 patients) had involvement of mixed nerve, 0.05% (1 patient) had involvement of motor nerve. All patients were preoperatively evaluated both clinically and radiologically. FNAC was also done to confirm the origin of the swelling. Patients who were diagnosed as having nerve sheath tumour clinically, radiologically and after FNAC were included in the study. Complete excision with preservation of nerve was done in all cases except for one case in which nerve graft was used. The mean follow-up period has been 2 years.

RESULTS

Schwannomas were located in the upper limb in 16 patients and in the lower limb in 2 patients. The tumour were located in the right side of the body in 10 patients and in the left side of the body in 8 patients. In total, 22 tumour in 18 patients were excised. Schwannomas originated from major peripheral nerves and digital nerves (18 tumour) or small nerve branches (4 tumour). 16 tumour arising from major peripheral nerves were located in the upper extremity (Fig. 1 and Fig. 2 A, B, C) and 2 tumour in the lower extremity. 4 tumour originating from small nerve branches were located only in the upper extremity.

Multiple tumours were removed in 3 patients. Two plexiform neurilemmomas arising from small nerve branches from the forearm area and one tumour from peroneal nerve were excised in a 46-year-old female patient. 2 tumours were excised from ulnar nerve in a 26-year-old female and in another 22-year-old female patient 2 tumours originating from superficial radial nerve in the forearm were removed.

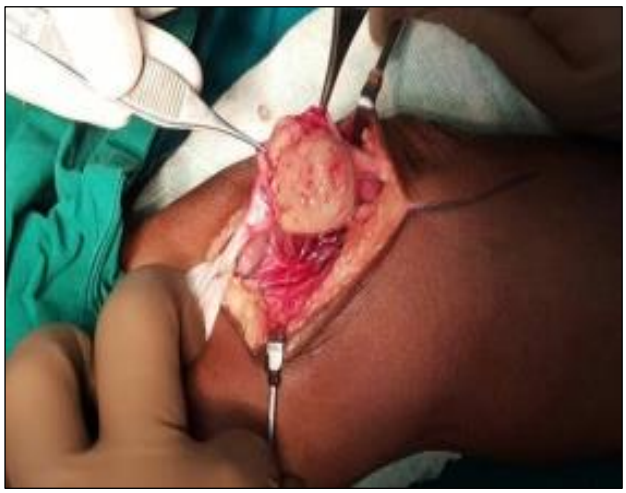

Figure 1: Schwannoma of ulnar nerve at elbow.

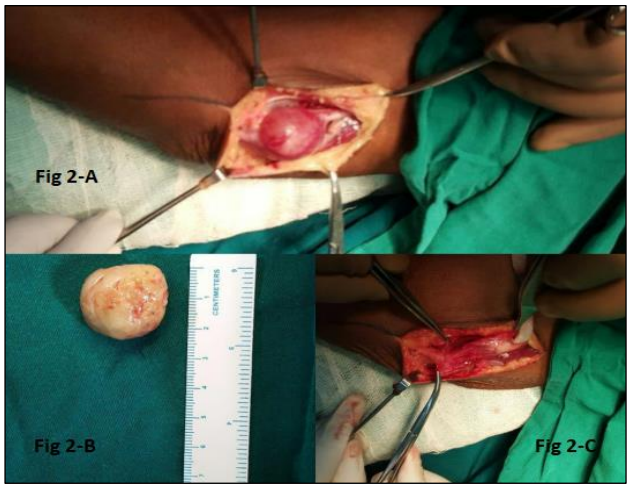

**Figure 2: (A): Schwannoma of ulnar nerve
(B): Excised specimen after enucleation
(C): Ulnar nerve in continuity after excision of the schwannoma.**

Complete excision with preservation of nerve was done in all cases except for one case in which nerve graft was used (Figure 3-A and B).

In the group of tumours arising from major peripheral nerves, preoperative evaluation revealed positive Hoffmann-Tinel sign and presence of paresthesias in 16 out of 18 patients (88.9%). Pain was present in 13 out of 18 patients (72.2%). In all 18 patients, pain was triggered by applying pressure to the tumour mass and appeared after exertion. Superficial sensory function impairment was detected in 12 out of 18 patients (66.7%) and motor deficit in 4 out of 18 patients (22.2%). In the group of tumours originating from small nerve branches, presence of tumour mass with moderate pain caused by applying pressure to it and after exertion was detected during

preoperative examination. Neither paresthesias nor sensory or motor deficits nor positive Hoffmann- Tinel sign was observed.

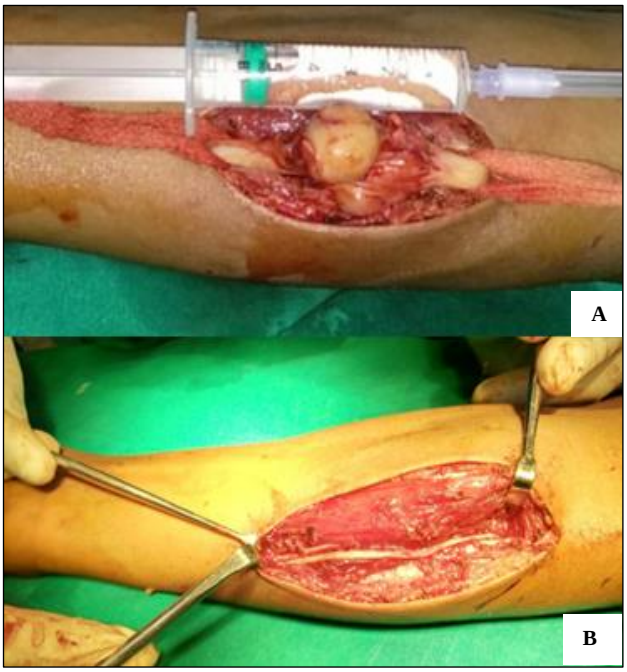

Figure 3: (A): Schwannoma of the ulnar nerve in forearm, (B): After excision of schwannoma, defect repaired with sural nerve graft cables.

In all of the examined tumour, histopathological pattern typical of schwannoma was observed. In the group of tumour arising from major peripheral nerves, 22 classical schwannomas were diagnosed. Tumour originating from small nerve branches had histopathological pattern of classical schwannoma (Antoni A) in 2 cases, plexiform schwannoma in 2 cases.

In none of the patients tumour recurrence occurred. In the group of tumour arising from major peripheral nerves, pain resolved in 18 patients (100%). No relationship between presence of pain and the size of tumour has been observed. Paresthesias resolved in 13 out of 16 patients who had suffered from them preoperatively (81.2%). Negative Hoffmann-Tinel sign was observed in 14 out of 16 patients, in whom it had been positive before operative treatment (91.6%) The tumour arising from small nerve branches were removed in one piece without identification of the site of origin. After removal of those tumours, definitive healing was achieved.

DISCUSSION

Schwannoma is a benign, well-defined, and solitary nerve sheath tumour and accounts for 8% of all soft tissue tumours.[15] Peripheral nerve sheath tumours comprise a spectrum of neoplastic potential ranging from benign neurofibroma and schwannoma to high-grade peripheral nerve sheath tumours. It is not only a very rare tumour, but it is often misdiagnosed due to lack of awareness and often confused with other common lesions like lipoma, fibroma or ganglion. Benign nerve sheath tumours can be either neurofibroma which is associated with Von Recklinghausen disease or could be a schwannoma which are mostly solitary. Neurofibromas are multiple, lack tumour capsule and mostly originate from terminal nerve ends. However, schwannomas are well circumscribed, encapsulated, eccentrically located and usually involving the proximal nerves.[4,16] Schwannoma tend to displace the nerve, however neurofibroma tend to grow within the nerve causing fusiform dilatation. Although malignant transformation of a schwannoma is exceedingly rare, in a review by Ziadi 10 out of 32 cases might have originated from a schwannoma. However, all these 10 cases were Malignant Nerve Sheath Tumours (MNST) of cranial nerves. He thus raised the hypothesis that intracranial MNSTs might be more associated to schwannoma than their peripheral counterpart.[17]

Schwannoma can practically occur in any part of the body and any peripheral nerve. Authors have however, demonstrated its presence in peripheral nerves of upper and lower limbs. According to Stull MA et al, schwannoma is usually found on the anterior aspect of upper limb and posterior aspect of the lower limb.[18] In our study also all lower limb schwannomas were located on the posterior aspect and in the upper limb six out of seven were located in the anterior compartment. Schwannoma usually present as slow growing painless tumour of variable size ranging from 2 cm to 20 cm. However, in our study the largest schwannoma was 8 cm. Schwannoma usually occurs between 20-50 years. In our study the findings were somewhat similar ranging from 28 to 66 years.

Most of these tumour are ignored by the patient until they become significantly large in size or when they start compressing the nerve trunks. As the size increases the tumour can compress over the primary nerve trunk and, hence causing variable symptoms. In case of extremities, painless swelling and paraesthesia were the most common symptom in this study (50%). Preoperative clinical diagnosis in these cases is difficult with differential diagnosis of fibroma, ganglioma, lipoma and xanthoma. Diagnosis purely on clinical assessment can be misleading and inaccurate. Collaborating with this fact, Phalen reported accurate diagnosis of schwannoma only in 31% of upper extremity schwannoma cases.[19] Similarly, White accurately reported schwannoma in only 5 of 32 cases.[6] Preoperative sonography and MRI can be supportive for a definitive diagnosis. It provides information about the size, anatomical location, relationship with other structures and malignant infiltration. In this study preoperative MRI was done in all cases with 100% accuracy. Although, preoperative imaging is not essential in every soft tissue swelling of extremity, it should be performed whenever in doubt. Depending on the nerve of origin either they can be very superficial and easily accessible or can be deep in the

intermuscular plane. Planning of the incision should be done in such a way that it is directly overlying the tumour and extending on to the uninvolved segment of the nerve. Tracing the nerve down from the virgin territory makes it possible to identify the summit between the mass and the nerve trunk which is the point of injury in most of the cases. It is a must to operate all these cases under tourniquet because intraoperative bleeding can completely obscure the field of vision and making it almost impossible to identify the plane of dissection. Authors advocate use of magnification, either with help of microscope or a surgical loupe.

Sometimes it may be very difficult to distinguish between the tumour and the nerve tissue. Once the right plane is identified it is possible to completely enucleate the tumour without damaging the nerve fascicles which can also be confirmed with intraoperative nerve stimulation. Schwannoma is a very well encapsulated tumour and rarely infiltrate the adjacent nerve.

Hence, there is very little chance to leave behind any tumour tissue which would cause recurrence. Extracapsular excision is an operative technique commonly used for removal of schwannomas.[4,14,20] To reduce the risk of damage to nerve fascicles during dissection of a tumour, different authors suggest modifications of the operative technique. Hussain et al, propose "tumour release by incising the capsule far lateral to the path of the nerve and dissecting the tumour circumferentially from its capsule. The epineural capsule is then left behind and acts as a protective covering of the nerve."[20]

The intracapsular technique was used by Date et al.[21] The above authors have compared results obtained after extracapsular and intracapsular enucleation of schwannoma and have found the latter technique to be superior due to lower risk of complications.[21] In case of presence of adhesions between epineurium and the capsule, Date and co-workers performed microenucleation of the tumour (tumour was resected piece-by-piece). The only case where we had to excise a nerve segment and reconstruct using a nerve graft was in a 30-year-old male having an ulnar nerve schwannoma (Figure 2). This patient had postoperative anaesthesia over the ulnar nerve territory which recovered in four months.

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