CASE REPORT

Benign Solitary Schwannoma of Right Ulnar Nerve – A Case Report

Ajay Pant[1], Julfiqar[2], Najmul Huda[3], M. Aslam[4]

[1]M.S. Ortho., Professor and Head, Department of Orthopedics, Teerthanker Mahaveer Medical College and Research Centre (TMMC and RC), TMU, Moradabad, U.P., [2]M.S. Ortho, Assistant Professor, Department of Orthopedics, Teerthanker Mahaveer Medical College and Research Centre (TMMC and RC), TMU, Moradabad, U.P., [3]M.S.Ortho, Professor, Department of Orthopedics, Teerthanker Mahaveer Medical College and Research Centre (TMMC and RC), TMU, Moradabad, U.P., [4]M.S. Ortho, Assistant professor, Department of Orthopedics, Teerthanker Mahaveer Medical College and Research Centre (TMMC&RC), TMU, Moradabad, U.P.

ABSTRACT

Introduction: Benign peripheral nerve schwannomas are uncommon tumours. Extra cranial schwannomas have also been reported from uncommon and unusual sites including breast, pancreas, and gastrointestinal system. Peripheral nerve schwannomas may pose a problem in clinical diagnosis, however an appropriate diagnostic work-up including thorough history and clinical examination, Ultrasonography, magnetic resonance imaging, fine needle aspiration cytology, nerve conduction velocity and electromyography study may all help reaching the correct preoperative diagnosis. The important clinical differential diagnoses include traumatic neuroma, neurfibromas, lipoma, cold abscess and muscle hernia. The definitive treatment of benign peripheral nerve schwannoma is complete enucleation of the tumour mass without damaging the intact nerve fascicles followed by confirmatory histopathological examination. When there is a doubt on histopathology, a positive Immunohistochemical staining with S100 is helpful in confirming a diagnosis of schwannoma. **Case presentation:** We had 40 years female who had a slow growing swelling over the inner aspect of her right elbow for the last one year; this was followed by pain, tingling and numbness over inner one and half fingers of her right hand for six months. Tinnels sign was positive over the swelling. Her subsequent clinical examination and investigations including a magnetic resonance imaging was suggestive of a benign growth in her right ulnar nerve in the elbow region. Complete enucleation of the swelling was done from the right ulnar nerve in the elbow region and subsequent histopathological examination confirmed it to be a benign cellular schwannoma. Patient recovered successfully after the surgery and paresthesia in the distribution of her right ulnar nerve also improved six weeks after surgery. At her last follow-up six months after surgery, the patient was completely asymptomatic and highly satisfied with the results of surgery. **Conclusion:** A correct preoperative diagnosis of peripheral nerve schwannomas is possible, and it can be successfully managed with complete enucleation of tumour mass with satisfactory patient outcomes.

Keywords: Benign, Peripheral nerve, ulnar nerve, Schwannoma, Enucleation, and Magnetic resonance imaging

INTRODUCTION

Schwannomas are benign tumours of schwan cell origin, and can involve cranial nerve, spinal nerve roots, intercostal nerve, posterior mediastinum, reteroperitonium. Solitary peripheral nerve tumours are uncommon,[1-3] and an accurate preoperative clinical diagnosis may not be possible in a fair proportion of patients.[4] The Schwannomas can involve a peripheral nerve partially or completely. They usually present as an innocuous solitary mass in the extremities in 3rd to 5th decade of life, with or without associated neurological symptoms. There is no sex predilection.

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Pathologically Schwannomas are benign encapsulated neoplasms of schwan cell. Microscopically they are composed of two cell patterns Antoni type A and Antoni type B. Schwannomas usually arise from a single fascicle and grow circumferentially displacing the intact nerve fascilces and, are located eccentrically on the nerve root. Multiple Schwannomas are rare and have usually been reported in association with neurofibromatosis 2(NF2), or schwannomatosis. Diagnosis at times may be challenging and a careful clinical assessment and even technical investigations like Magnetic resonance imaging, CT and angiography may fail to indicate the correct preoperative diagnosis.[5,6] The important clinical differentials that are often confused with a benign solitary schwannoma of an extremity include traumatic neuroma, neurofibrma, lipoma, cold abscess muscle hernia, haemangioma, and synovial cyst etc.[6] Clinically it is extremely difficult to differentiate schwannoma from a neurofibroma, however intraoperative appearance of the tumour mass,

Corresponding Author:
Dr. Julfiqar, M.S. Ortho, Assistant professor, Department of Orthopedics, Teerthanker Mahaveer Medical College and Research Centre (TMMC and RC), TMU, NH-24, Near Pakwara, Dist. Moradabad, U.P. E_mail: drzulfiqar1983@rediffmail.com, Fax: 05912360077, Mob. No: +91-8979949300

its gross morphological characteristics and the subsequent histo-pathological and immuno -histochemical examination can settle the diagnosis in almost all the cases. Few authors have reported worsening of the nerve function following surgical enucleation of these tumours[4] but, fortunately majority of the benign peripheral nerve schwannomas can be safely and easily separated from the peripheral nerve trunk without damaging the peripheral nerve trunk.[7] Some authors have demonstrated an acceptable postsurgical nerve dysfunction in these patients.[8] Satisfactory surgical outcomes can be achieved without the risk of recurrence provided, complete tumour enucleation has been done. The objective of present case report is to show that although difficult but, a strong clinical suspicion along with relevant investigations can be helpful to have a correct preoperative diagnosis of these tumours. Some of the peculiar findings of peripheral nerve schwannomas has also been discussed in this case report.

CASE PRESENTATION

A 40 years female presented to us with a gradually increasing swelling over the back and inner aspect of her right elbow, for the last one year [Fig. 1], and paresthesia over inner one and half fingers of her right hand. To begin with the swelling was small and it gradually increased to the present status. She also developed paresthesia in the form of tingling and numbness in the distribution of right ulnar nerve for the last six months. On examination we found a single soft swelling measuring about 5X4 cm over the poster medial aspect of right elbow. The transillumination test was negative, and the swelling was freely mobile along the axis perpendicular to the right upper limb, and it was immobile along the longitudinal axis of the limb. The elbow range of motion was normal [Fig. 1]. The Tinnel's sign was positive over the swelling, sensations were diminished over right little and ulnar half of ring finger, no motor loss was found.

An x-ray of her right elbow antero-posterior and lateral views excluded any primary or secondary bony/articular involvement however; a globoid radio-opaque soft tissue swelling was seen over posteromedial aspect of right distal humerus. A careful scrutiny of this soft tissue shadow, further demonstrated an eccentrically located longitudinal soft tissue shadow (?ulnar nerve) [Fig. 2a and b]. A contrast magnetic resonance imaging (MRI) showed a well defined peripherally enhancing lesion measuring 3.4X2.8X3.8 cm in superficial postero-medial Supracondylar region, with an eccentrically located right ulnar nerve in the lesion. Characteristic "target sign" was also present on MRI [Fig. 3a and b]. The fat plane with underlying triceps muscle was found intact. There was no infiltration into the surrounding tissues, and there was no perilesional edema on MRI. An aspiration cytology revealed sheets of spindle cells with abundant thick cytoplasm and round to oval nuclei (?peripheral nerve sheath tumour) based on this a provisional diagnosis of a solitary benign peripheral nerve sheath tumour was made.

A complete surgical enucleation of the tumour mass followed by histopathological examination was planned. The tumour mass was exposed through a postero-medial skin incision over the right elbow under tourniquet control with the patient in supine position. Intraoperatively the tumour mass was easily separated with the underlying triceps muscle, as well as the intervening fat pad. The exposed tumour mass showed enlarged tortuous epinural blood vessels, characteristics of schwannoma [Fig. 4a], and the nerve fascicles stretched over the tumour mass. A surgical plane was developed between the intact nerve fascicles and tumour mass, to expose the actual tumour mass [Fig. 4b]. The complete enucleation of the tumour mass was possible without any obvious structural damage to the intact nerve fascicles. The gross morphology of the tumour was of a brownish, soft globular mass measuring 3x3X1.5 cm, with central haemorrrahge probably secondary to needle trauma at the time of aspiration cytology [Fig. 4c].

The final diagnosis of benign schwannoma was confirmed on histopathological examination, that showed an encapsulated hyper cellular tumour of pleomorphic spindle

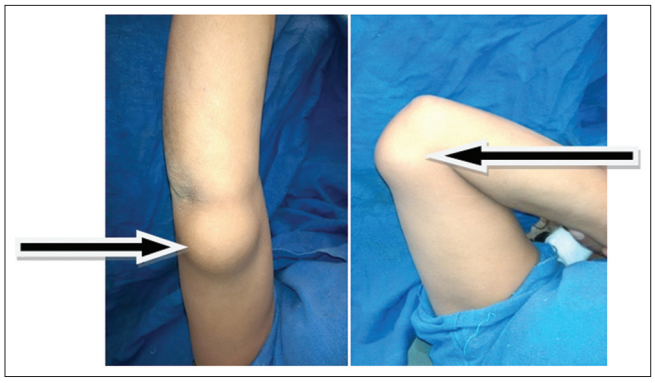

Figure 1: Clinical photograph of the patient showing a swelling over the posteromedial aspesct of her right elbow.

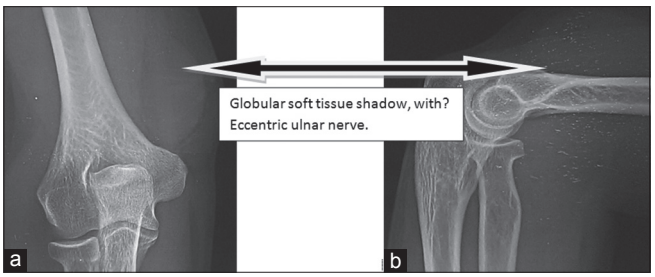

Figure 2: (a and b) Radiograph of the same patient showing soft tissue swelling embded in the posteromedial aspect of right supracondylar region, and ecccenrically located ulnar nerve shadow.

cells with tapering nuclear ends, growing in the uniform fascicular pattern with prominent palisading at most places i.e. Antoni A pattern, with few less cellular areas i.e. Antony B pattern with prominent myxoid back ground. Few palisades of spindle cells with intervening cytoplasmic process i.e. Verocay body [Fig. 5], were also seen. Further investigations like Immunohistochemical staining with S100 protein was not required to establish it as a case of benign solitary schwannoma of right ulnar nerve, in the light of above mentioned evidences.

DISCUSSION

Schwannomas represent 5% to 8% of all soft tissue neoplasms, and because of this rarity they usually pose a challenge to have correct preoperative diagnosis. Commonly schwannomas present as sporadic solitary peripheral nerve mass and, when multiple are usually seen in association with NF2 or schwannomatosis. They present as isolated mass in the head neck region or over the volar and extensor aspects of extremities. Failure to timely detect these tumours may lead to loss of significant neurological functions of the patient. The upper and lower extremity ratio of schwannoma is 2:1 as seen in major series.[4,7] The pathological variants of schwannomas include the most common conventional variety followed by, cellular, plexiform and the melanotic schwannoma.[9] Apart from the gross morphological differential diagnoses as discussed above, the important histological conditions that need differentiation from schwannomas include, palisaded leiyomyoma, palisaded myofibroblastoma of inguinal lymph nodes, gastro-intestinal stromal tumours, plexiform neurofibroma,palisaded encapsulated neuroma,neurotropic melanoma, clear cell sarcoma of soft parts etc.[9] The histological differentiation of these conditions from schwannoma certainly requires an expert and experienced histopathologist. In case of doubt on histology, Immunohistochemical examination using S100 is very helpful to differentiate schwannomas that stain positive for S100, from rest of the conditions. An appropriate clinical history and examination followed by investigations like Ultrasonography, MRI and electromyographic studies may sometimes help in a correct preoperative diagnosis, but this is not the truth in a significant proportion of cases. The role of fine needle aspiration cytology is doubtful, rather it is condemned by some, for two reasons, first being fear of trauma to the normal nerve tissue, and second due to poor predictability of cytological examination for schwannomas. Majority of solitary benign peripheral nerve schwannoma can be enucleated safely without any structural damage to the nerve trunk, because the tumour mass in schwannomas

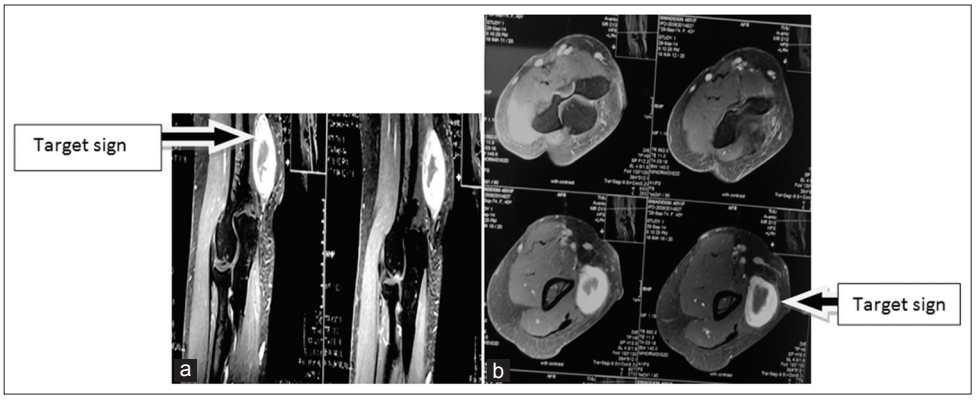

Figure 3: (a,b) MRI of the right elbow showing the characteristic 'target sign"

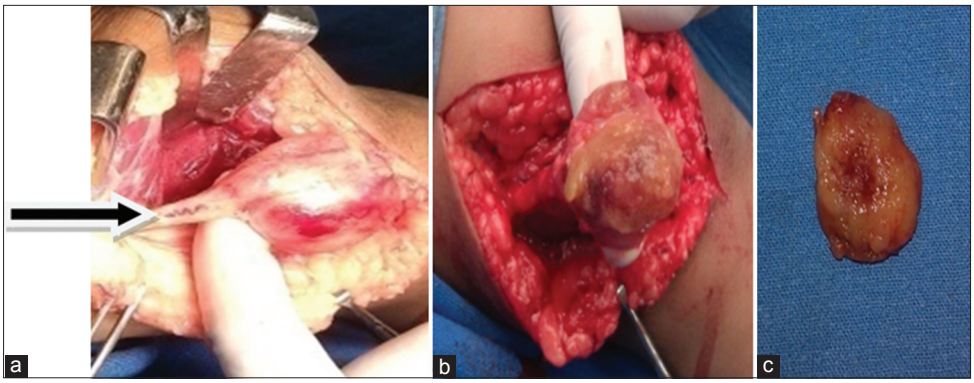

Figure 4: (a-c) Intraoperative clinical photographs of the same patient.

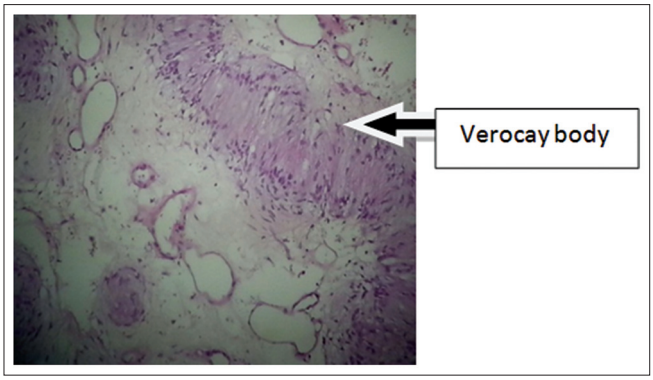

Figure 5: Microscopic picture showing the charecteristic "verocay body".

is less intermingled with the normal tissue. Benign solitary schwannomas have excellent prognosis and, unless incompletely excised there is no risk of recurrence. Magnetic resonance imaging is the imaging modality of choice that can identify the nerve of origin, encapsulation, position of the nerve trunk in the tumour mass, tumour dimensions, and perilesional edema, and the characteristic target sign.[10] Except in the conventional variant there is no risk of malignant conversion of schwannomas. Our patient had slow growing mass in the region of her right ulnar nerve with characteristics paresthesia in her right inner one and half fingers that lead to the first suspicion of a peripheral nerve tumour in this case. The plain radiograph of the elbow is also suggestive of an eccentrically growing mass in the right ulnar nerve in the region of elbow, a finding that has been described very rarely by other authors reporting peripheral nerve Schwannomas.[11] Intra-operatively we found dilated and tortuous epinural blood vessels that are characteristics of schwannomas and help to differentiate it from neurfibromas. Identification of intact nerve fascicles stretched over the tumour mass may at times require microsurgical instruments but it helps a lot to find the correct surgical plane, while operating these tumours. A correct surgical plane is critical to prevent any undesirable damage to the intact nerve fascicles and prevent postoperative neurological deficit. In our patient we did complete enucleation of the right ulnar nerve schwannoma, which was turned out to be benign cellular schwannoma on histopathological examination. Six weeks after surgery patient had fully regained normal sensations in her right inner one and half fingers, and she was highly satisfied with her surgical outcomes. At her last follow-up 6 months after surgery, she was doing well all her routine activities.

CONCLUSION

Although difficult but, a preoperative diagnosis of Solitary benign schwannomas of peripheral nerve is possible. These tumours can be completely enucleated successfully without damaging the nerve with excellent surgical outcomes. Identification of intact nerve fascicles is crucial to prevent damage to intact nerve fascicles.

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