Case Report

An uncommon case of big schwannoma of hand in a child

***N.N.Samanta[1], Hareesha[2], V.Narsimha Rao[3] , L.Jayanth[2], M.Praveen[4]**

[1]Professor & HOD, Department of Pediatric Surgery, [2]Post Graduate, Department of General Surgery, [3]Assistant Professor, Department of Plastic Surgery, Kamineni Institute of Medical Sciences, Narketpally, [4]Civil Assistant Surgeon, District Hospital, Nalgonda.

ABSTRACT

Schwannomas are benign encapsulated neoplasms of the peripheral nerve sheath. They arise from proliferation of differentiated Schwann cells of single nerve funiculus. These tumors may affect any nerve within the body and arise from supporting cell within a nerve sheath that contains a basement membrane and resembles a schwann cell. They arise in the sensory portion of the nerves but can arise not only in association with any peripheral nerve but also along the flexor surfaces of extremities.

Here, we submit an interesting case of Schwannoma over the volar aspect of hand in a 11 year old female.

Keywords: Schwannoma, neurofibroma, hand, soft tissue tumor

Introduction

Benign tumours involving peripheral nerves of the upper extremity are uncommon.[1] Schwannomas also known as neurolemmas usually originate from Schwann cells located in the peripheral nerve sheaths. They are the most common tumours of the hand (0.8–2%).[2] They usually grow slowly and appear as painless swellings for several years before being diagnosed.[3] They usually present with solitary swelling along the course of the nerve[1], however multiple lesions may be present in cases of Neurofibromatosis type 1, familial neurofibromatosis, and sporadic schwannomatosis. Incidence is similar between both genders and commonly seen in 3rd and 6th decades.

Though generally asymptomatic, pain, numbness, and fatigue may accompany increasing size of the tumor. It is unusual for a schwannoma to exceed three centimeters in diameter.[1] EMG, MRI, and USG are helpful in the diagnosis. Surgical removal is usually curative.

***Corresponding Author :**
Dr. N.N.Samanta,
Professor & HOD, Dept. Of Pediatric Surgery,
Kamineni Institute of Medical Sciences,
Narketpally, Nalgonda district,
Telangana state-508254
Email: nripendrasamanta4@gmail.com

CASE REPORT

A 11 year old female presented with complaint of

Swelling in palm of right hand since 5 years.

Mild pain over the swelling

No history of trauma or foreign body implantation

History of excision of a smaller swelling 5 years back, in local hospital, details of previous surgery or biopsy report are not available.

On local examination of Right hand :

Inspection : Single swelling of size 8cm x 6cm, bosselated surface with vessels over the surface of the swelling. No evidence of visible pulsations over the swelling (Fig 1).

Palpation : No local rise of temperature over the swelling. No tenderness. All the inspectory findings

are confirmed on palpation. Nodular, non pulsatile, non compressible swelling, firm in consistency, with mobility restricted in both the directions. No evidence of any contracture. No palpable regional lymph node.

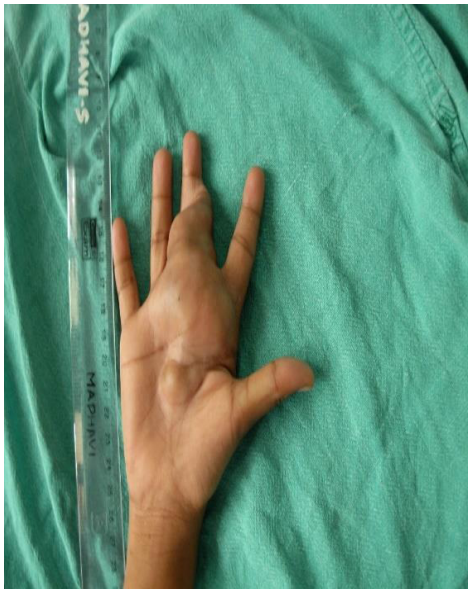

Fig 1 – Pre Operative picture of the swelling of Right hand

Management :

Routine investigations revealed -

Hb - 11.2gm% ; TC - 7,100/dL; BT - 3'00''; CT - 4'00'';

CUE, Blood Urea, Serum Creatinine, Serum Electrolytes - All are within normal limits

HIV, HBsAg, HCV - Non Reactive

Chest Xray and ECG are normal

Specific investigations :

FNAC of the swelling : Features suggestive of Nerve sheath tumor – ? Schwannoma

X Ray Right hand AP and Oblique view - Soft tissue swelling adjacent to right 3rd metacarpophallangeal joint. Underlying bone normal.

USG of the swelling were done to establish the diagnosis - Mixed echoeic well defined lesion, confined to subcutaneous plane, without any calcifications. ? Neurofibroma.

MRI of Right hand – T2 weighted T1 weighted hypointense aggressive lesion with no erosion or infiltration of adjacent structure.

? Infantile aggressive fibromatosis

? Giant Cell Tumour of tendon sheath

Intra Operative findings and Procedure :

Pre Operative marking done on the volar aspect of the hand including the previous scar and under tourniquet control & using microsurgical technique (loupe). Nodular swelling encircling the digital nerves was found. Complete excision of the swelling with sparing of digital nerves done.

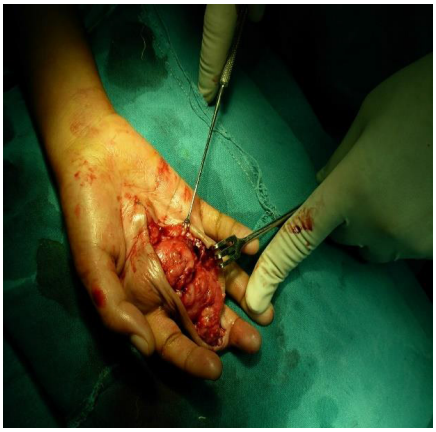

Fig 2 – Intra Operative picture showing well defined swelling with bossellated surface, surrounding the 3rd metacarpal and proximal phallynx

Figure 3 – Picture taken on 2nd Post Operative day

Post Operative events :

Post Operative period was uneventful. On 5th post operative day, all the functions of right hand along with flexion, extension of middle finger were found to be preserved and she was able to write, make a grip and hold objects.

On 9th post operative day, patient was discharged after suture removal. In follow up after 1 month, all the functions of the hand preserved and no sensory loss was found.

Histopathological Report :

Sections studied show capsulated lesion consisting of hypercellular and hypocellular areas. In hypercellular areas, nuclei showing palisading appearance with dense chromatin. Verocay bodies noted. Loose reticulated hypocellular areas noted. Features suggestive of Schwannoma (Fig 4).

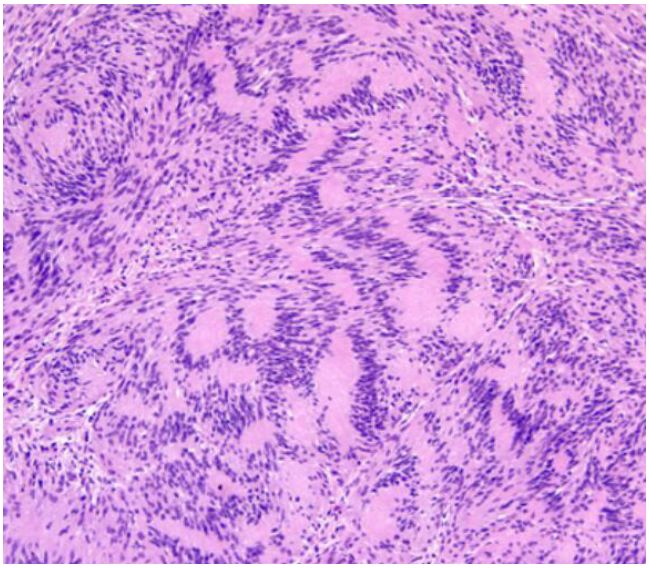

Fig 4 –Histopathology showing Verocay bodies

DISCUSSION:

Schwannomas are rare tumours. They are usually solitary and benign lesions; however they can be multiple suggesting an underlying tumour predisposition syndrome[3] and may be associated with neurofibromatosis type 1 and schwannomatosis. In the last 15 years, there have been several reports of multiple schwannomas with no evidence of a vestibular tumour leading schwannomatosis being considered a clinical entity different from other forms of neurofibromatosis.[2] The incidence of multiple schwannomas has been reported as 1% to 23%. Mostly affected nerves are ulnar and median nerves.[4] Several cases of multiple schwannomas of median nerve reported in the literature. Wrist and palm involvement similar to our case is reported only in one of these studies.[5] The reported interval between onset of symptoms and surgery has varied from a few months to years.[3]In our patient the onset of swelling was 2 months after the excision of the first swelling and pain started 15 days prior to presentation.

These tumours are slow growing, soft in consistency, mobile in nature, and sometimes painless so they may be misdiagnosed as lipoma, fibroma, ganglion, or xanthoma. Both USG & MRI were not successful in diagnosing schwannoma in our case. MRI can provide useful information about morphological data on the median nerve tumours; Dynamic information is provided by USG of the swelling.[6] Surgical excision is the most effective method of therapy. Use of torniquet and careful microsurgical dissection in a bloodless field, is advised to avoid damaging the nerve fibres during the epineural and endoneural dissection. Paresthesia is the most common postoperative complication. In this case, we have also used microsurgical technique to remove the tumour and tried to protect the nerves.

References

1. D. S. Louis and F. M. Hankin, “Benign nerve tumors of the upper extremity,” Bulletin of the New York Academy of Medicine (1985),61(7),611–620.

2. O. Ozdemir, C. Kurt, E. Coskunol, I. Calli, and M. H. Ozsoy, “Schwannomas of the hand and wrist: long-term results and review of the literature,” Journal of Orthopaedic Surgery(2005), 13(3),267–272.

3. A. Gonzalvo, A. Fowler, R. J. Cook et al., “Schwannomatosis, sporadic schwannomatosis, and familial schwannomatosis: a surgical series with long-term follow-up: clinical article,” Journal of Neurosurgery(2011), 114(3), 756–762.

4. M. R. Patel, K. Mody, and V. J. Moradia, “Multiple schwannomas of the ulnar nerve: a case report,” Journal of Hand Surgery(1996), 21(5), 875–876.

5. H. J. Kang, S. J. Shin, and E. S. Kang, “Schwannomas of the upper extremity,” Journal of Hand Surgery(2000), 25(6), 604–607.

6. A. K. Bhattacharyya, R. Perrin, and A. Guha, “Peripheral nerve tumors: management strategies and molecular insights,” Journal of Neuro-Oncology(2004), 69(1–3), 335–349.

7. S. H. Lee, H. G. Jung, Y. C. Park, and H.-S. Kim, “Results of neurilemmoma treatment: a review of 78 cases,” Orthopedics(2001), 24(10), 977–980.surgical .2004;47;2: 95-98.