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IP Journal of Surgery and Allied Sciences

Journal homepage: https://www.jsas.co.in/

Case Report

Cutaneous plexiform schwannoma of the digit- A case report

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ARTICLE INFO

Article history:
Received Case Report
Accepted 29-12-2021
Available online 16-03-2022

Keywords:
Digit
Plexiform Schwannoma
DIP & PIP joint
Surgical excision

ABSTRACT

Schwannoma is a most common type of benign peripheral nerve sheath tumor in adults. Plexiform schwannoma [PS] are relatively rare with reported incidence of only 5%. Skin and soft tissues are primarily involved structure. It is rare to find them on the digits. We report a case of expansile swelling on the digit. The definitive diagnosis was revealed only after exision biopsy. Schwannoma should be included in the differential diagnosis of any swelling arising from the digits.

1. Introduction

Soft tissue tumors are frequently encountered in the hand surgeon's practice. Ganglion is the most common hand tumor followed by giant cell tumor (GCT) and soft tissue tumors[1] Soft tissue tumors include a variety of tumors like lipoma and its variants, fibroma and peripheral nerve sheath tumor[2] Peripheral nerve sheath tumor includes neurofibroma and schwannoma, schwannoma being the most common among these. Schwannoma is a benign proliferation of Schwann cells. It grows slowly eccentrically over the peripheral nerve and is usually solitary. manifests about 5% of benign soft-tissue neoplasm. Schwannomas usually involve in head, neck, flexor surfaces of extremities and nerves. Plexiform schwannoma is a rare distinctive variant, accounting for 2-5% of all schwannomas. It frequently affects patients aged 30–40 years with no sex predilection, and occurs commonly in the dermis and subcutis with a predilection for head and neck.

Histologically, schwannoma has the consistent appearance of differentiated Schwann cell and has 2 components: highly ordered dense arrays of spindle cells (Antoni A areas) and a hypo cellular region of connective tissue with less organized spindle cells (Antoni B areas).[3] Immunohistochemistry helps in differentiating schwannomas from neurofibromas.

The management consists of excision biopsy and regular follow up. The recurrence of this tumor type is extremely rare. The malignant transformation is also very unusual.[4]

2. Case Report

A 17year old male presented with a painless, slow growing swelling over right ring finger. He did not complain of any neurological deficit in the finger and no other similar swelling over other body parts.

There was no prior history of trauma. Examination revealed a single, firm, non tender, non- fluctuant swelling of 3x2 cm size with smooth surface and well defined margin present over dorsum of ring finger of right hand extending from DIP joint to PIP joint (Figure 1). It was not fixed to the bone and the patient showed no signs of sensory loss or movement impairment. Mobility restricted on flexion of DIP joint. X ray right hand showed normal bony architecture and radiolucency noted over mid phalanx 4^{th} digit of right hand (Figure 2). USG showing

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https://doi.org/10.18231/j.jsas.2022.006

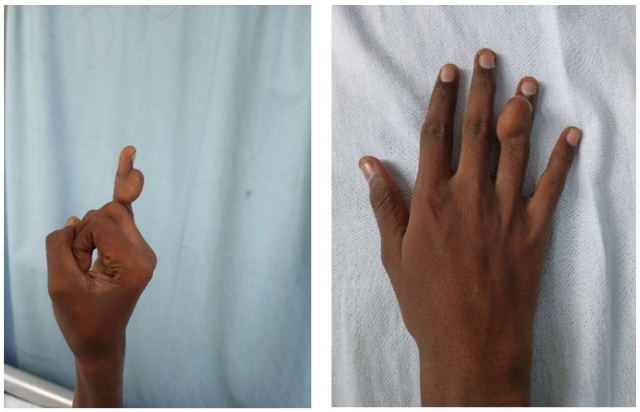

Fig. 1: Clinical photograph of patient.

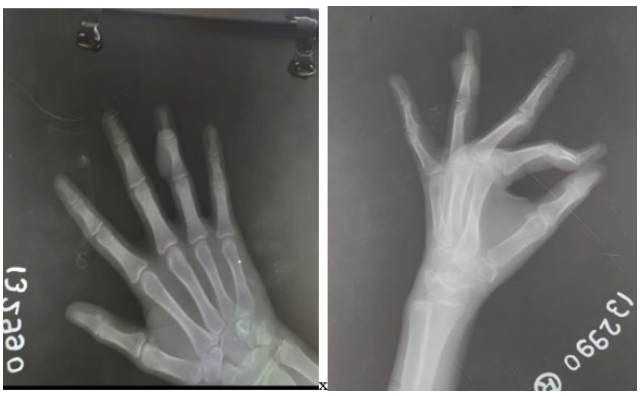

Fig. 2: Xray right hand showing radiolucency over 4^{th} digit.

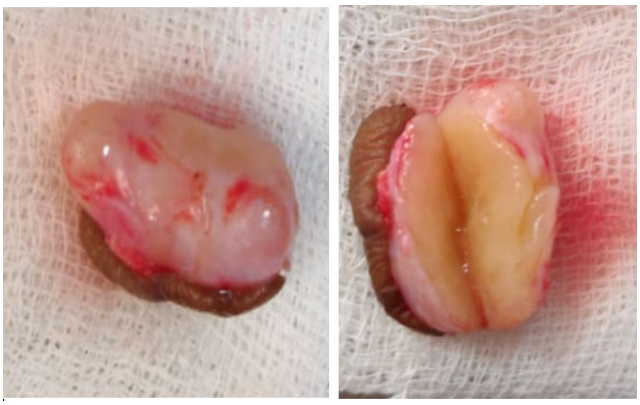

Fig. 3: Specimen photograph and cut section

heterogeneously hypoechoic lesion suggestive of benign etiology. FNAC was inconclusive. Surgical excision was planned under digital nerve block. Intra operatively the lesion was seen to be well encapsulated and arising from the radial digital nerve. It was easily dissected off the digital nerve. The skin was noted to be thinned out over the swelling and not adherent to the tumor. The excision included a small island of skin since there was stretching of skin over the swelling (Figure 3). The excised tumor measured 3x2cm in dimension. After excision the skin was closed primarily. The histopathological analysis showed benign type of plexiform schwannoma. In postoperative period there was no neurological deficit of the involved finger.

3. Discussion

Accounting for 5% of all schwannomas, PS was first described by Harkin and Reed in 1978, a rare benign tumour composed exclusively of Schwann cells exhibiting a plexiform growth pattern.[5] Though a common mimicker of Plexiform neurofibroma {PN}, PS can occur sporadically and in weak association with NF2.[6] It is important to differentiate between the two, as recurrence and malignant potential are common with PN. PS can be seen both in superficial and deep locations. In the superficial location, 90% are cutaneous in skin and subcutaneous tissues. Among the deep seated locations, PS can occur in the deep somatic soft tissues of extremities,retroperitoneum, trunk, parotid, thoracic space and vulva.[7] Usually solitary, PS can be multiple in cases of Schwannomatosis.[8] Woodruff JM et al., reviewed six cases of congenital and childhood plexiform schwannomas. Of the six, one was congenital and none had features of NF1. Two thirds presented in infancy, with F:M ratio of 4:3. Tumour size ranged from 2 to 9 cm, two were well circumscribed while four had both circumscribed and infiltrative margins. All tumours had high cellularity, hyperchromasia and brisk proliferative activity making them prone to be misdiagnosed as Malignant Peripheral Nerve Sheath Tumours (MPNST), but their nuclei were 3 times the size of NF nuclei and showed minimal variation. Mitoses ranged from 4-31/10 hpf. All the six cases showed diffuse S-100 positivity but were p53 negative. Surgical excision was done in all cases after a course of chemotherapy which failed to decrease the size. Recurrences were commonly seen in <1 year after initial excision which was termed as incomplete resection rather than recurrence. Argenyi ZB et al., described a childhood plexiform tumour diffusely showing S-100 positivity and ultrastructural features of schwann cells and called it-"congenital neural hamartoma-fascicular schwannoma". PS differs from conventional cellular Schwannoma by its multinodularity, greater cellularity, increased proliferative activity and potential for infiltrative growth into surrounding tissues due to lack of encapsulation. Absence of well-

defined capsule, degenerative changes and lymphoid collections were common features in PS.[8] Cellular schwannomas were more commonly seen in adults while PS was commonly observed in children. The proliferative activity of PS in childhood is attributed to factors such as rapid tissue growth in children. So called recurrence in many is because of incomplete resection, as clinicians avoid wide excision in young individuals, as seen in present case.[9] Malignant transformation is more common in PN than PS.[10] However a misdiagnosis is possible, especially in biopsy material from a cellular PS with hyperchromasia and increased mitosis which may be labelled as PN/MPNST.

4. Conclusion

The plexiform type of schwannoma is a rare, benign neural tumour prognostically distinct from plexiform neurofibroma. Awareness of this subtype, especially in biopsy material is crucial in dictating the treatment. Judicious use of cytochemical and immunohistochemical stains can aid in the accurate diagnosis of plexiform schwannoma, which needs adequate surgical removal without extensive follow-up, unlike plexiform neurofibroma. Even though peripheral nerve sheath tumors are rare, they should be kept in mind as differential diagnosis of a slow-growing painless soft tissue tumor over the digit.

5. Source of Funding

None.

6. Conflict of Interest

The author declares that there is no conflict of interest.

References

1. The J, Whiteley G. MRI of soft tissue masses of the hand and wrist. *Br J Radiol*. 2007;80(949):47–63. doi:10.1259/bjr/53596176.
2. Cheng JW, Tang S, Yang YT. Sonographic features of soft tissue tumors in the hand and forearm. *Chang Gung Med J*. 2007;30(6):547–54.
3. Kurtkaya YÖ, Scheithauer B, Woodruff JM. The pathobiologic spectrum of schwannomas. *Histol Histopathol*. 2003;18(3):925–34. doi:10.14670/HH-18.925.
4. Sunita B, Harsh K, Pinky S, Charusheela G. Malignant epitheloid schwannoma: A rare tumor causing a diagnostic dilemma. *Int J Pharm Biomed Sci*. 2012;3(4):238–41.
5. Woodruff JM, Marshall ML, Godwin TA, Funkhouser JW, Thompson NJ, Erlandson RA, et al. Plexiform (multinodular) schwannoma: a tumour simulating the plexiform neurofibroma. *Am J Surg Pathol*. 1983;7(7):691–7. doi:10.1097/00000478-198310000-00009.
6. Seong H, Lim JJ, Chung KY. Neurofibromatosis type 2 with multiple plexiform schwannomas. *Int J Dermatol*. 2004;43(5):336–40. doi:10.1111/j.1365-4632.2004.01864.x.
7. Narsimha PA, Prakash S, Cristina RA. Deep seated plexiform schwannoma- a pathological study of 16 cases and comparative analysis with superficial variety. *Am J Surg Pathol*. 2005;29(8):1042–8.
8. Woodruff JM, Scheithauer BW, Kurtkaya-Yapicier O, Raffel C, Amr SS, Laquaglia MP. Congenital and Childhood plexiform (multinodular) cellular schwannoma. *Am J Surg Pathol*. 2003;27(10):1321–29. doi:10.1097/00000478-200310000-00004.
9. Argenyi ZB, Goodenberger ME, Struass JS. Congenital neural hamartoma (fascicular Schwannoma): a light microscopic, immunohistochemical and ultrastructural study. *Am J Dermatopathol*. 1990;12(3):283–93.
10. Enzinger FM, Weiss SW. Benign tumours of peripheral nerves In: Enzinger and Weiss's Soft tissue tumours. S W, JR G, editors. St Louis: Mosby; 2008. p. 867.

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Cite this article: Ahmad Z, Audichya A, Shah NJ. Cutaneous plexiform schwannoma of the digit- A case report. *IP J Surg Allied Sci* 2022;4(1):29-31.