Case Report

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Plexiform Schwannoma Masquerading as Giant Cell Tumor of Tendon Sheath- A Case Report

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ABSTRACT

Plexiform schwannoma is a rare benign variant of Schwann cell tumor characterised by multinodular plexiform growth pattern. Usual location of this tumor includes trunk, head, neck and upper extremities but it may rarely be found in lower extremities. It generally affects young adults with no sex predilection. We report a case of 37 years old male who presented with gradually progressive swelling of right little finger of 28years duration that was diagnosed as giant cell tumor of tendon sheath clinically, radiologically and intraoperatively.Authors wish to discuss the case due to its clinical rarity and diagnostic difficulties due to many common histological mimickers at that site.

Keywords: *Plexiform, Schwannoma, Giant Cell Tumor, Neurofibroma, Spindle Cells.*

Introduction

Schwannomas also known as neurilemmoma are benign truly encapsulated neoplasms of schwann cell origin. They comprise 5% of all benign soft tissue tumors. Most of the cases are solitary and sporadic but multifocality has been associated with type 2 neurofibromatosis. Most common locations include flexor surface of extremities, neck, mediastinum, retroperitoneum, posterior spinal roots, and cerebellopontine angle. Plexiform schwannoma is a unique variant and comprise about 4.3% of all schwannomas. They generally present as painless slow growing asymptomatic nodules with a diameter generally less than 2cm.[1] Histologically they mimic plexiform neurofibroma which has a strong association with von recklinhausen's disease and has a malignant potential.[2]

Case Report

We report a case of 37year old malewith no known co morbiditieswho presented with swelling over tip of right little finger of 28years duration. Swelling was painless and gradually progressive and not associated with any functional disability(Fig1(a&b)). There was a history of surgery done twice in 1997 and 2009 in a village for which no records were available with the patient. There was no history of trauma. There were no similar lesions on others parts of body. There was no history of similar swelling in the family members.

Examination revealed 04x03 cm soft tissue swelling over tip of right little finger. It was not associated with tenderness, overlying skin was normal and there was no distal neurological deficit. Range of motion of little finger was normal.

Plain radiograph of lesion showed increased radio-opacity in soft tissues around distal phalanx extending upto level of proximal interphalangeal joint. Cortical irregularity was noted in underlying distal phalanx (Fig 2a).

USG of lesion showed well defined hypoechoic lesion with multiple septae with few cystic areas noted within. Multiple vessels were seen coursing through the lesion. Based upon the location, clinical features and radiological findings a diagnosis of giant cell tumor of tendon sheath was made and FNA was not done. He was planned for excision of tumor with disarticulation of terminal phalanx. Intraoperatively surgeons tried to preserve distal phalnx and hence piecemeal dissection was done, however the distal phalanx could not be salvaged and entire distal phalanx was removed in bits and pieces.

Grossly the excised specimen measured 3.5x3.0x2.0 cm with a whitish tail of multiple nodules (Fig 2b). No areas of haemorrhage or necrosis were seen on cut surface

On microscopy, a tumor composed of spindle cells arranged in nodules in a plexiform architecture was seen in the dermis (Fig 3a). These spindle cells had wavy serpentine nuclei arranged in a palisading manner at places forming verocay bodies (Fig 3b). Tumor showed more hypercellular (Antoni A) and less hypocellular (Antoni B) areas. There was no nuclear atypia or mitosis. On immunohistochemical staining, S100 protein was strongly expressed in tumor cells (Fig 4a). Mib 1 labelling index was low (< 1%) (Fig 4b). Sections from bone could not be taken as it was received in form of sub-centimetric fragments.

Based on the characteristic microscopic findings confirmed by IHC, a diagnosis of plexiform schwannoma was given in the instant case.

Six months after surgery patient is doing well without any recurrence.

Discussion

Schwannomas are common, readily encountered and easily diagnosed soft tissue tumors due to characteristic histopathological features. They are generally classified as peripheral, visceral, intraspinal and intracranial based on their location. Peripheral schwannomas present as a papule or nodule and may undergo cystic or haemorrhagic degeneration within. Apart from this classification, schwannomas also contain several histological variants such as cellular schwannomas, pseudo-glandular schwannomas, epitheloid schwannomas, ancient schwannomas, neuroblastoma like schwannomas and plexiform schwannomas which might lead to difficulty in diagnosis sometimes.[3] Plexiform schwannoma is a rare variant and accounts for 2-5% of total cases. In 1978 Harkin et al reported the first 6 cases of plexiform schwannoma and distinguished them from plexiform neurofibroma.[4] It is known that most reported cases are small tumors and occur in individuals without any known predisposing factors but multiple lesions are more common in individuals with neurofibromatosis type 2, Gorlinkoutlas syndrome, schwannomatosis, family history and history of trauma. Our patient had no predisposing factors. It generally presents in childhood but congenital forms are also known to occur. They are usually located on head and neck region (23%), the upper trunk (22%), lower extremities (22%) and trunk (18%).[5] Patients generally show normal karyotype but few case reports involving numerical changes in chromosomes 22, 7 and sex chromosomes have been described. Cases with trisomy 17 have also been described.[6] Differential diagnosis includes plexiform neurofibroma and malignant peripheral nerve sheath tumors (MPNST). Plexiform neurofibroma is composed of tortuous mass of nerve branches formed by proliferation of all elements of peripheral nerves in a background of loose myxoid stroma and it lacks characteristic Antoni A, Antoni B and verocay body. S 100 protein also stains purely Schwann cell population in plexiform schwannoma strongly whereas it has a variable/focal expression in plexiform neurofibroma. MPNST on other hand shows nuclear atypia, nuclear overcrowding, necrosis with marked mitotic activity.[7] Differentiating plexiform schwannoma from plexiform neurofibroma is of utmost importance because the latter has a propensity for malignant transformation. Local excision is the treatment of choice for most of cases of plexiform schwannoma. Prevalence of recurring tumors after their complete removal is rare (2%).[8] Incomplete removal during earlier surgeries may explain the recurrence in our case.

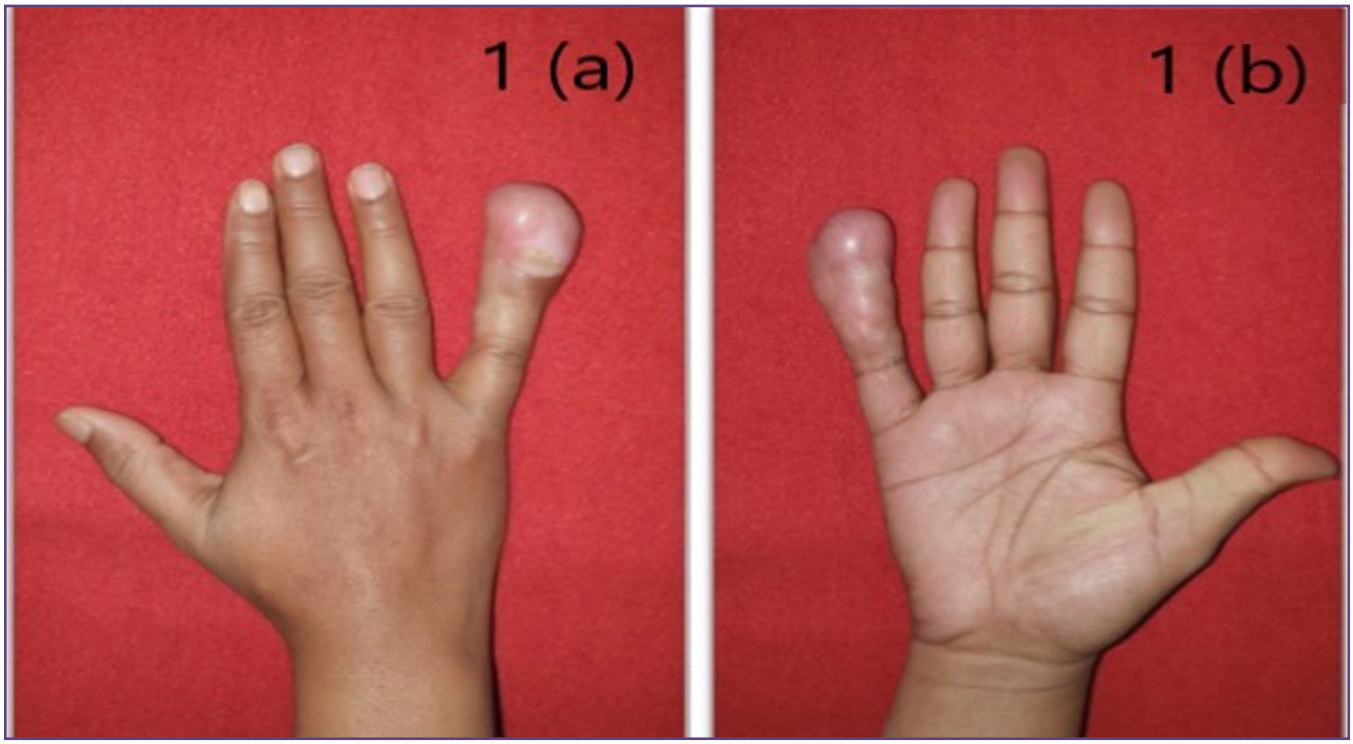

Fig. 1(a&b): Globular Swelling at tip of right little finger.

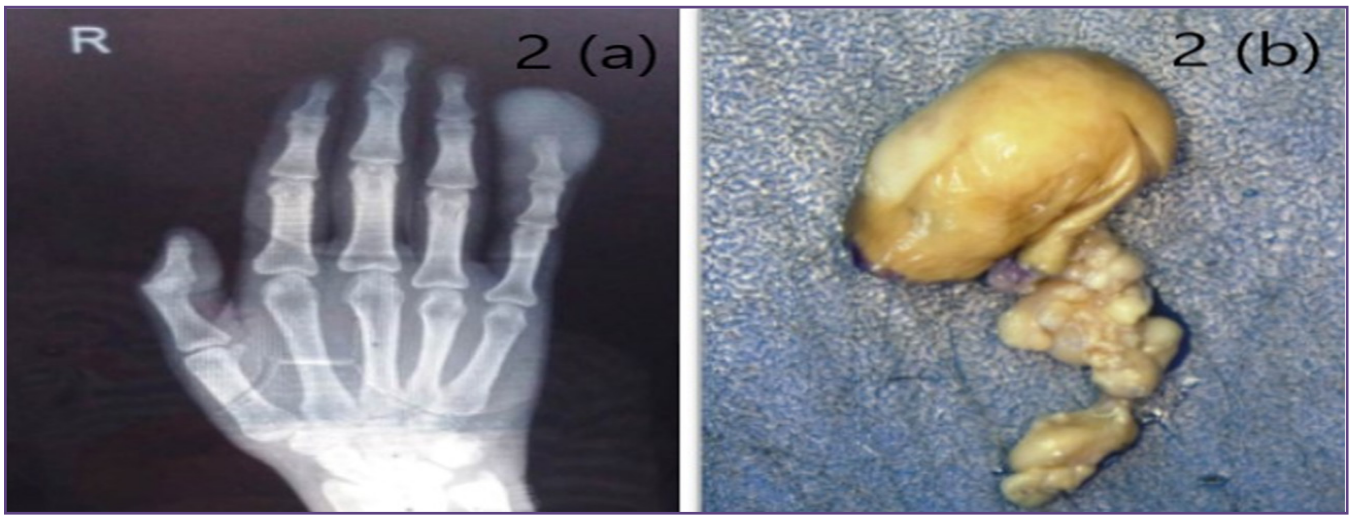

Fig. 2(a): Radio-opacity in the soft tissues around distal phalanx of right little finger with mild cortical irregularity in the underlying bone.

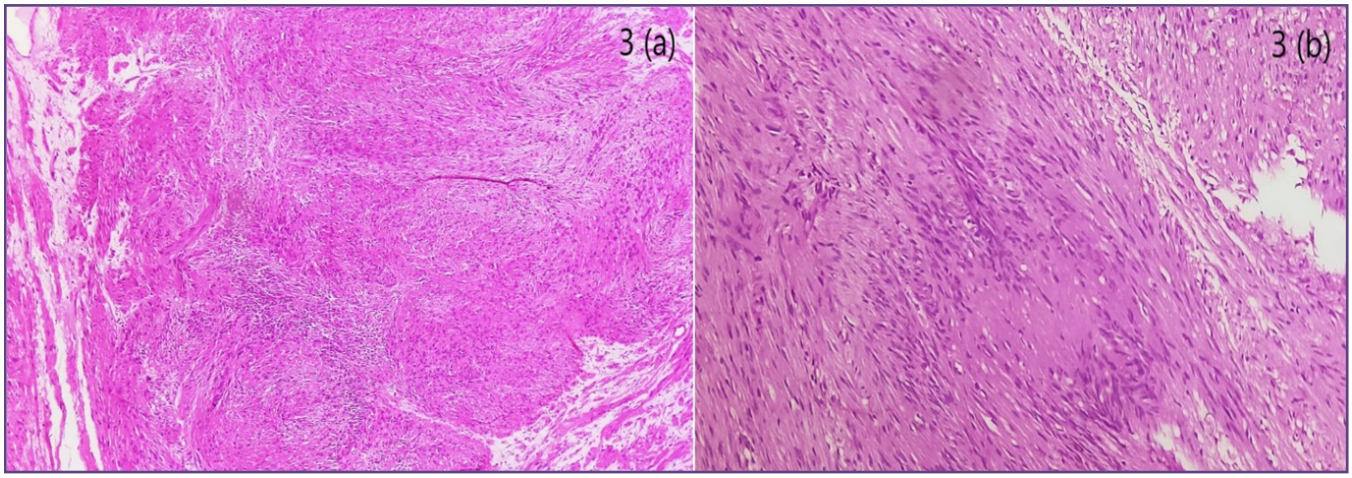

Fig. 3(a): Multiple nodules of oval to spindly cells with a plexiform architecture (100x, black arrow); Fig 3(b): Antoni A area showing nuclear palisading and Verocay body (400x, black arrow).

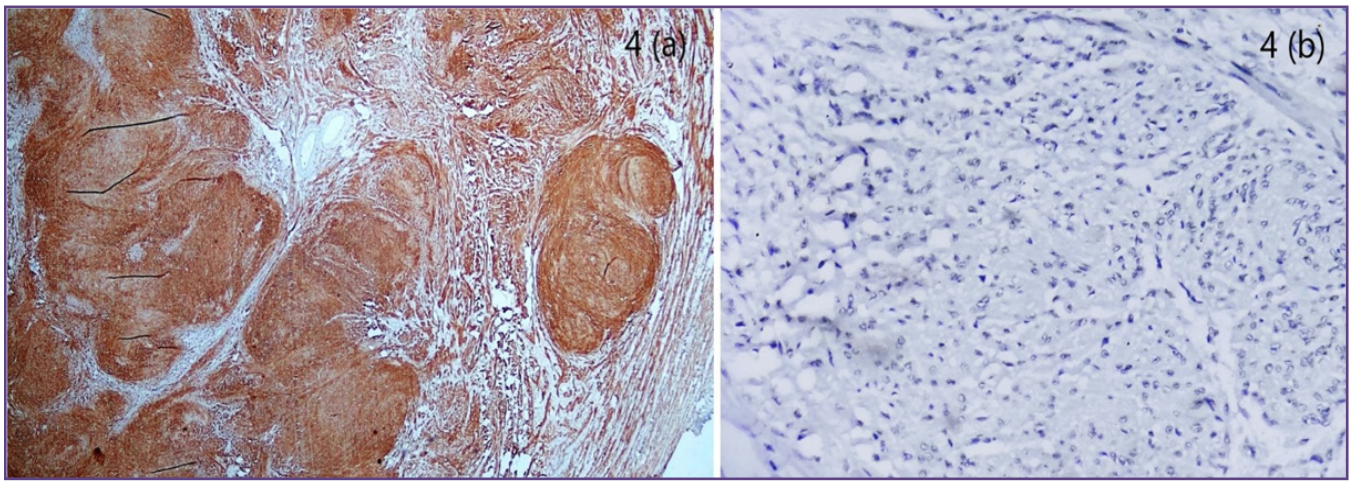

Fig. 4(a): S100 is strongly positive in tumor cells (100x); Fig 4(b): Mib1 labelling index < 1% (400x).

A variety of unusual presentation of schwannomas in hand have been described including intraosseous schwannomas[9] but we could not find any reports of unusual presentation of plexiform schwannoma with clinical and radiological characteristics analogous to giant cell tumor of tendon sheath. Giant cell tumor of tendon sheath is the second most commonly encountered lesionof hand after ganglion cyst and generally presents in 3rd to 5th decade of life as a slow growing soft tissue growth over palmar surface of radial three digits near distal interphalangeal joint.

Conclusion

We present a rare case of plexiform schwannoma arising over tip of right little finger which was diagnosed as giant cell tumor of tendon sheath clinically and radiologically. Careful gross examination of the tissue and histopathological examination helped in making a correct diagnosis in this case. Our case also had recurrence of the lesion twice though prevalence of recurrence is very low in peripheral schwannomas.

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Competing Interests

Nil

Refrences

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