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DIAGNOSTIC DILEMMA: SCHWANNOMA OR GANGLION CYST

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Abstract

Schwannomas are rare, slow growing benign tumors of peripheral nerve sheath originating from Schwann cells surrounding the nerve associated with delayed presentation of pain and paresthesia. Their incidence being 5% of all upper extremity tumors. They may be a part of neurofibromatosis called schwannomatosis with multiple peripheral schwannomas. We present a case of solitary schwannomas from the peripheral nerves probably superficial branch of median nerve mistaken initially for ganglion cyst.

Keywords

Ganglion cyst, schwannoma, peripheral nerve sheath tumors, diagnosis

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INTRODUCTION:

Schwannoma is a benign nerve sheath tumour arising from differentiated Schwann cells. It is an encapsulated, well circumscribed tumour. The World Health Organization classifies schwannoma as a grade I benign tumour. All ages can be affected, but is more common in 30 - 60 years of age and has equal male to female ratio. 90% are sporadic, 3% with neurofibromatosis type 2, 2% with schwannomatosis, 5% with meningiomatosis with or without neurofibromatosis type 2(1). It occurs more frequently on the limbs with a predilection to the upper limbs, followed by the head and neck area, including the oral cavity, orbit and salivary glands(2). More commonly larger nerves of upper limb are involved(3). Malignant transformation is very rare. It is usually asymptomatic; measuring between 10-250mm, with larger variants causing neurological symptoms(4). Hence misdiagnosed at presentation with other gangliomas, tenosynovitis(5). Hems et al. reviewed a series of 104 peripheral-nerve benign tumours, and demonstrated that in only seven cases the preoperative diagnosis was accurate(6).

CASE REPORT:

We present a case of 33 yrs old female, presenting with swelling in the volar aspect of right wrist since 3 years, pain since 1 month. There is no history of constitutional symptoms, no history suggestive of malignancy and no history of trauma. On examination the swelling was 4*3 cms, smooth, non-tender, mobile and no adhesions to surrounding structures.

The patient was advised ultrasonography and x-ray of wrist which showed a hypoechoic lesion over flexor carpi ulnaris and palmaris tendons, non-communicating with wrist joint suggestive of ganglion cyst and no bony abnormalities respectively. The patient was reassured and treated conservatively. The patient however came again with same symptoms, not relived on treatment. The FNAC of the lesion was done, which showed Spindle cells. Then MRI left wrist was done, which showed solid hypodense lesion suggestive of malignancy. The blood investigations were within normal limits. Then under due precautions and consent, the tumour was excised in toto. The specimen was sent to histopathology which gave definitive result as schwannoma. The patient was negative for neurofibromatosis on retrospective assessment. On follow-up after 1 month, patient was asymptomatic and clinically better.

DISCUSSION:

Schwannomas peaking its incidence in 3rd and 4th decade, arising from peripheral nerve Schwann cells; with up to 19% incidence in upper extremity, should be considered in differential diagnosis of swellings in upper extremity. Being a nerve sheath tumour, they have predilection to volar surface of upper limb due to concentration of nerves. Whenever, encountering volar wrist soft tissue mass always consider neurofibroma, schwannoma, fibroma of tendon sheath, ganglion cyst, tenosynovitis and sarcomas. Schwannoma is usually present as slow growing painful or painless swelling; associated with paraesthesia and positive Tinel sign(7). Neurofibroma usually present as bosselated/ plexiform, invasive, intraneural tumour with pain and paraesthesia with more incidences of malignant transformation(8). Ganglions are the most common soft tissue masses of the hand and wrist. They may be symptomatic or asymptomatic. Most ganglion cysts are located at the dorsal

aspect of the wrist (60–70%), whereas volar wrist cysts account for approximately 18–20%(9). Preoperative diagnosis is most important as soft tissue sarcomas account for 1% of all malignancies with 13% incidence in upper extremity(10). Hence a diagnostic protocol of clinical examination, ultrasonography, MR Imaging, biopsy, En-block excision and histopathology is required. Reports have shown that 5–80% of patients with soft tissue sarcoma undergo unplanned resection, leading to higher local recurrence and increased morbidity and mortality(11). Ultrasound is another cost-effective imaging modality with a high negative predictive value for soft tissue masses. US can help to determine the consistency (solid or cystic), relationship to adjacent structures, vascularity, size, and shape of the mass(12). MRI is the most sensitive and specific imaging modality available in the evaluation of soft-tissue masses. MRI is the study of choice for the localization and staging of soft-tissue tumours as it offers the best delineation of the soft-tissue structures and the relationship of the mass with respect to neurovascular structures(6). Though it is a seemingly simple procedure, biopsy of soft-tissue masses is technically challenging and potentially fraught with complications. Careful planning is required to determine the exact location and approach to the mass since they is a high chance of local extension and/or neurovascular damage(13). At US examination schwannoma appears like a capsulized mass with a “basket” internal architecture. Ultrasonography cannot always reveal the connection of the tumour with the nerve. Sometimes a cystic area is revealed in large-sized schwannomas; this area can cause large schwannomas to be mistaken for a ganglion thus causing an incorrect pre-operative diagnosis(14). The MR aspect of schwannoma is that of a homogeneous mass, with a middle intensity signal at SE T1-weight sequences, whereas at T2-weight sequences it shows a high signal(6). Surgical management of schwannoma is marginal excision. Due to their encapsulated nature, schwannomas are more amenable to intraneural dissection than neurofibromas, allowing preservation of the associated nerve. If the affected nerve is not of

functional importance, then tumour excision with primary nerve repair versus nerve grafting is the recommended treatment. Cutaneous nerves can be easily excised with sacrifice of the nerve(8).

CONCLUSION:

In approaching a palpable mass in the upper limbs, the possibility of a peripheral nerve neoplasm should always be considered. The clinical examination must be very meticulous. We have to look for typical signs of schwannomas, such as the positive Tinel sign and peripheral paraesthesias. Step wise assessment with radiology, ultrasound, MRI, biopsy and last the surgery. Microsurgical techniques are recommended for better clinical outcomes.

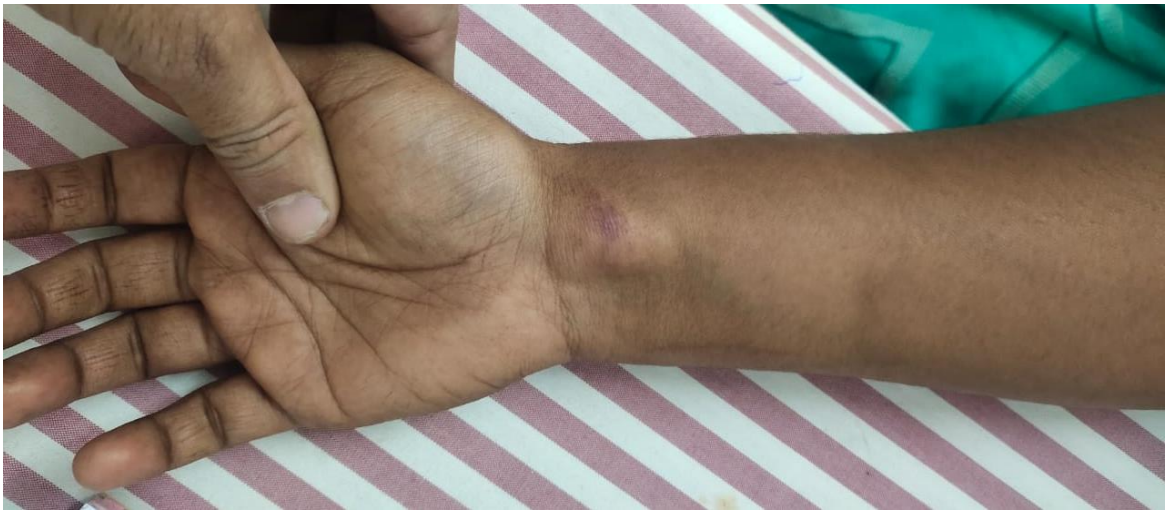

Figure 1 clinical picture showing volar swelling in right wrist

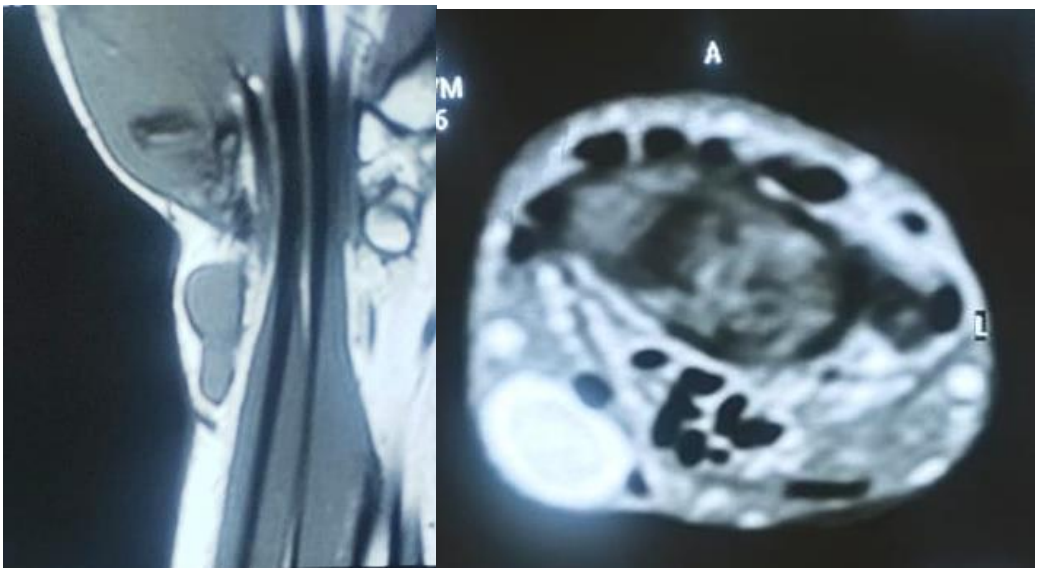

Figure 2 MRI showing hypointense lesion on T1(left) and hyperintense lesion on T2(right) in volar subcutaneous plane of right wrist

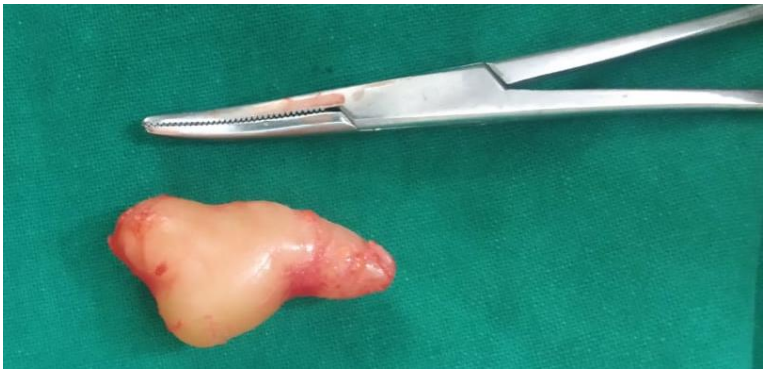

Figure 3 showing the completely excised encapsulated tumour

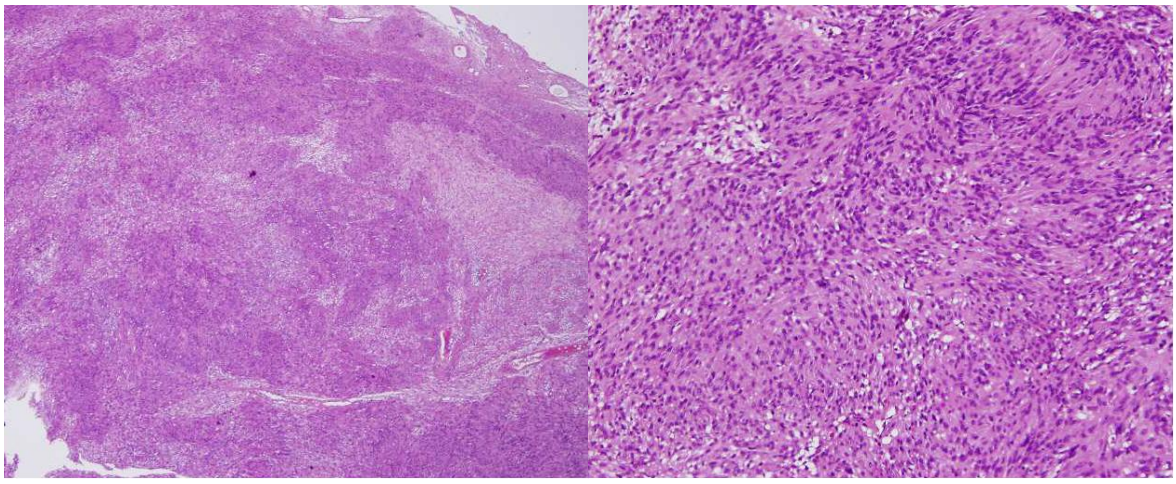

Figure 4 Histopathologic examination shows hypocellular Antoni (A) and hypercellular Antoni (B) Verocay bodies, confirming the diagnosis of benign schwannoma

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