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Case Report

Benign peripheral nerve sheath tumor-varied clinical presentation

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ABSTRACT

Peripheral nerve sheath tumors (PNST) are a group of heterogeneous, often benign and a rare condition that originates from the neuroectodermal or neural crest and display features that mirror the elements of the nerve. Schwannomas are one such peripheral nerve sheath tumors which entirely are made up of benign neoplastic Schwann cells. The objective of this case report is to highlight the diverse clinical presentations of these swellings. In this presentation, reporting three cases of PNST in which two presented with neurological symptoms of paraesthesia and pain and one who was asymptomatic swelling over his neck. All of whom were diagnosed with an alternate soft tissue swelling post clinical examination and taken up for excision as there were no significant clinical evidence for imaging. Intra-operatively we noted that all were closely related to the peripheral nerve of that anatomical region. Histopathological study revealed it to be PNST. PNST and schwannoma in particular although an entity that is not so common to come across in the surgical clinic we need to have and high indices of suspicion when associated close to peripheral nerves and symptomatic of a nerve involvement as we discuss here below.

Keywords: Schwannoma, Nerve sheath tumors, Neurilemmoma

INTRODUCTION

Peripheral nerve sheath tumors represent 5% of soft tissue tumors.[1] Schwannoma being the most common type of benign peripheral nerve sheath tumor arising from the schwann cells which support the peripheral nerve fibers. They mostly occur as a solitary lesion or in association with neurofibromatosis as schwannomatosis. Most commonly present in the head and neck. Lower limbs are affected less. They present as a soft tissue mass, pain, and/or focal neurological deficit which completely or partially recovered after complete excision.

CASE REPORT

Case 1

A fifty year married female with history of swelling over her dominant right forearm for one year with numbness and paraesthesia. On physical examination, swelling over the volar aspect of the right forearm 7×5 cm, borders are ill defined, cystic in consistency, tenderness present. Swelling was transversely mobile but longitudinally immobile; the skin over the swelling was pinchable. Clinical diagnosis made as lipoma with cystic changes.

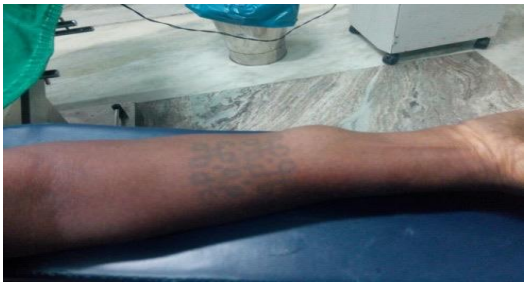

Figure 1: Right forearm swelling.

Case 2

A sixty year female with the history of swelling in the left ankle joint. On physical examination swelling 3×3 cm in the medial side of the left ankle joint, tenderness present, firm in consistency, not mobile but not attached to the skin. Clinical diagnosis made as ganglion left ankle.

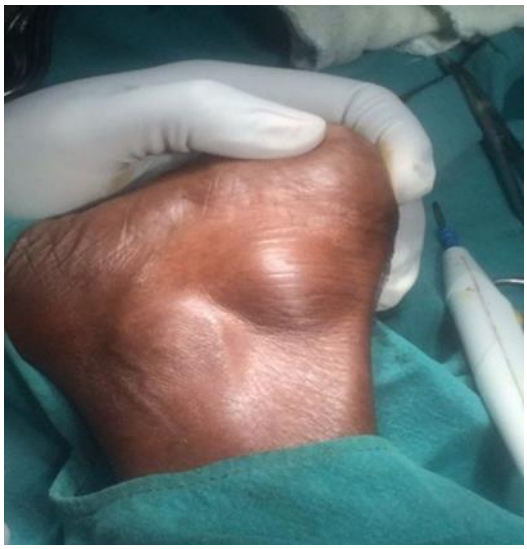

Figure 2: Swelling over left ankle, behind lateral malleolus.

Case 3

A fifty three year male with history of swelling in the right side of the neck. On physical examination, swelling of 5×4 cm, hard in consistency, mobile. Clinical diagnosis made as branchial cyst right side of the neck.

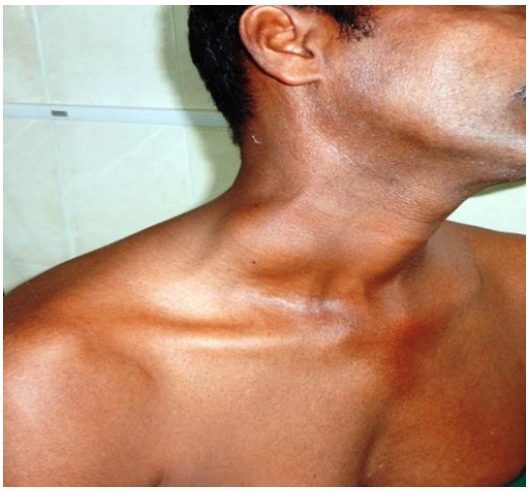

Figure 3: Swelling over the right side of the neck.

As diagnosis was made clinically surgical excision of the tumour was performed and cautious separation of the nerve and tumour was done for all three cases, In case 1 the tumour was related to ulnar nerve (Figure 4), case 2 related to posterior tibial nerve (Figure 5) and case 3 related to the brachial plexus (Figure 6).

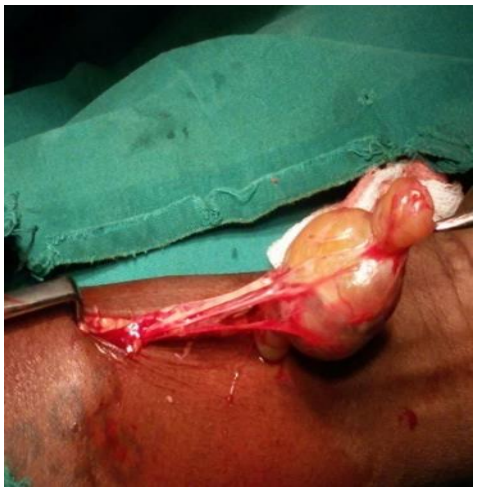

Figure 1 : Tumour related to ulnar nerve.

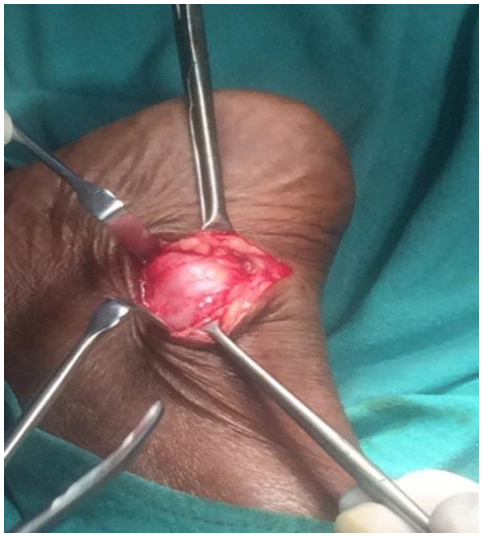

Figure 2: Tumour related to posterior tibial nerve.

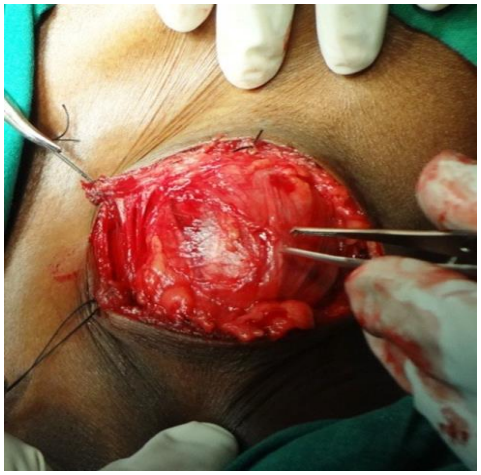

Figure 6: tumour related to brachial plexus.

Histopathology

Results of case 1: gross-multiple nodular grey white bits measuring 7×3×2.5 cm. External surface is unremarkable. Cut section solid to cystic. Cystic area appears haemorrhagic. Microscopy; encapsulated tumour with antoni A, antoni B areas of myxoid degeneration and presence of macrophages, cystic changes with hyalinisation of vessels is also seen. No mitosis or pleomorphism of cells noted (Figure 7). In case 2, single globular tissue piece measuring 2.5×1.5×1.5 cm outer surface smooth covered with white capsule cut section shows fatty yellow areas consistent with schwannoma (Figure 8), and case 3, multiple grey white bits largest measuring 4×3×2.5 cm resembling a cyst and multiple fragmented bits, consistent with schwannoma with degenerative changes (Figure 9).

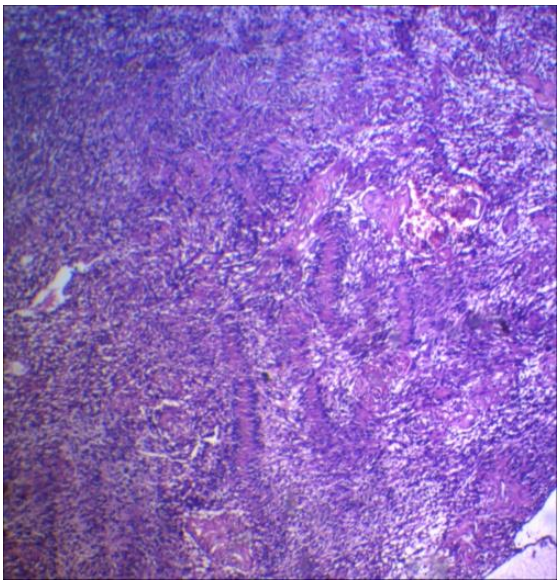

Figure 7: Verocay bodies.

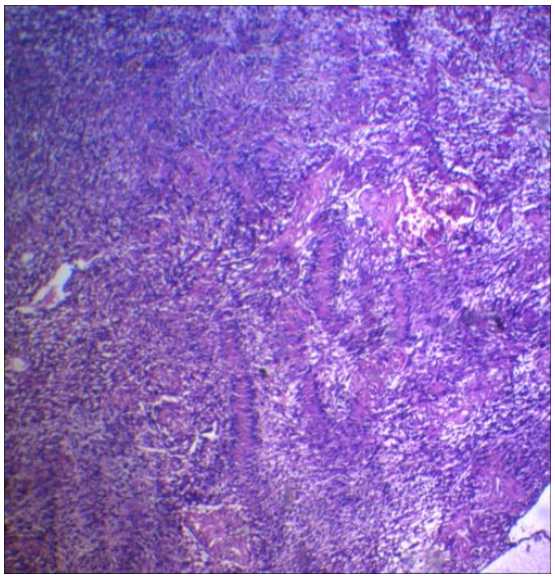

Figure 8: Nuclear palisading (high power).

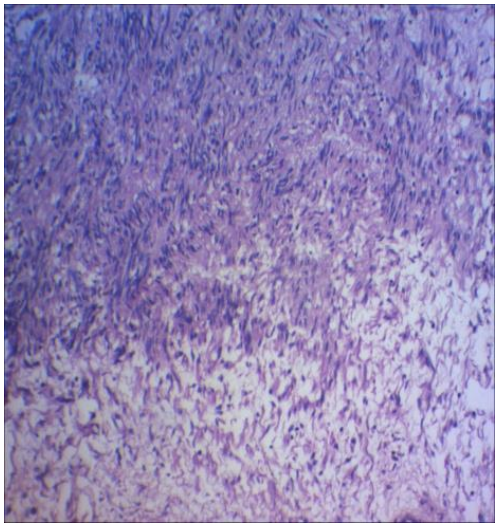

Figure 9: Antoni A and antoni B areas- H&Estain (low power).

Postoperative neurological deficit was not noted in our patients but it is not uncommon.

DISCUSSION

Nerve sheath tumor arises from the neuro ectodermal or neural crest origin. It is surrounded by epineurium and perineurium hence have a true capsule which facilitates enucleation. Benign nerve sheath tumors are divided into two i.e. schwannoma and neurofibromas.

Schwannoma are mostly solitary sporadic lesion but can also be associated with neurofibromatosis. They occur at all age groups and have no sex predilection. They can be present any part of the body. Mostly present in the head and neck and the extremities and less frequently in the chest, abdomen and pelvis. They are slow growing tumors with present with swelling which is soft, freely mobile but in restricted along the nerve, pain and paraesthesia. Tinel's sign, paraesthesia along the nerve elicited while percussion over the swelling might be present.[2] Neurological symptoms are rare but occur when the mass has compressed the neurovasular bundle.

Malignant transformation of such tumors is rare and are suggested by indistinct margins and infiltration of tumor.[3]

Dasgupta et al, studied 303 patients with benign solitary schwannoma where 45% of the tumors were seen in extremities and 9% in trunk and 14% in unusual sites. They had one documented case of malignant degeneration.[3]

Ultrasound defines the fascicular structure of nerves and allows to define the site, size of the Schwannoma as well as its relationship to surrounding structures and helps to plan surgery.[4] Ultrasound is cheaper, does not expose the patient to radiation and is widely available compared to

computed tomography and magnetic resonance imaging (MRI).

MRI is the best method for visualising nerve sheath tumors, it shows the extent and involvement of the tumor which typically have an intermediate signal intensity on T1-weighted images, very high signal intensity on T2-weighted images.

Management is surgical excision of the tumor. Surgical enucleation is possible, produces little damage to the underlying fascicles.

Post operatively, neurological complications is not uncommon.

Hirai et al studied the predictive factors for complications after surgery. They studied total of 139 patients, of which 49 developed postoperative complications. 42 with sensory disturbance and 8 with motor weakness. It was found that older age, tumors from upper extremities and major motor nerve involvement are associated with high complication rate.[5]

Histopathological hallmark features are the presence of alternating areas of compact spindle cells arranged in compact fascicles called antoni A areas, and less cellular and more disorganised areas called antoni B.

Schwannoma with degenerative changes include cyst formation, calcification, hemorrhage and hyalinisation. It is also known as ancient schwannoma. Since they behave like schwannoma the enucleation atypical is dismissed as degenerative changes.[6]

To conclude, schwannoma is often misdiagnosed due to their indirect signs and symptoms. Asymptomatic tumors are not followed by imaging unless considered high risk for neurological compromise. Since recurrence rate is very low, complete excision of the tumor with preservation of the nerve the standard therapeutic manoeuvre. Postoperative neurological deficit is common. The diagnosis of schwannoma is confirmed with histopathological findings.

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