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A RARE CASE OF ANCIENT SCHWANNOMA MIMICKING MALIGNANCY

General Surgery

Dr. Sachin Sehrawat	Junior Resident, Department Of General Surgery, Sgt Hospital, Gurugram
Dr. Pawan Tiwari	Professor, Department Of General Surgery, Sgt Hospital, Gurugram
Dr. Rajdip Solanki	Junior Resident, Department Of General Surgery, Sgt Hospital, Gurugram
Dr. Sanni Lathiya*	Junior Resident, Department Of General Surgery, Sgt Hospital, Gurugram, *Corresponding Author
Dr. Vipin Shrama	Junior Resident, Department Of General Surgery, Sgt Hospital, Gurugram
Dr. Jairaj Kothari	Junior Resident, Department Of General Surgery, Sgt Hospital, Gurugram
Dr. Bhavya Garg	Intern, Department Of General Surgery, Sgt Hospital, Gurugram

KEYWORDS

INTRODUCTION

Schwannoma is a relatively uncommon, lobulated, encapsulated, slow-growing, benign tumor that is derived apparently from the Schwann cells lining the nerve sheath. Its most common locations are the flexor surfaces of the extremities, neck, mediastinum, retroperitoneum, posterior spinal roots, and cerebellopontine angle. The occurrence on the lower limb accounts for 1% of all cases. Ancient schwannoma is considered as a variant of schwannoma, comprising about 10% of all schwannomas. The long-standing schwannoma attributes to degenerative changes and is termed as "ancient" schwannoma.[1] Schwannomas constitute 5% of benign soft tissue neoplasms and occurs most frequently in persons between the ages of 20 and 50 years. Men and women are equally affected. Schwannoma typically presents as a solitary, well capsulated, ovoid or fusiform configuration and slow growing mass. Patients with a schwannoma often have no symptom except for a palpable mass. [2] These tumours most commonly arise in the soft tissues of the head and neck and on the flexor surfaces of the upper and lower extremities. Schwannomas can present with no symptoms, mild symptoms or severe symptoms mostly affecting the nerves. Most lesions are solitary and present as a slowly growing painless soft.tissue mass. Symptoms are unusual, unless the mass has become large enough to compress the adjacent nerve[3].

Case Report

A 31-year-old female patient presented to surgery outpatient department with complaint of swelling over anterolateral aspect of left thigh which was of twelve months duration, insidious in onset, gradually progressive in size over the past twelve years. On local examination the swelling was 14*10 cm, non-tender, smooth surface, regular margins, non-mobile, no local increase of temperature, peripherally hard and centrally soft in consistency. FNAC from thigh swelling done, suggestive of negative for malignancy. Core needle biopsy was suggestive of possibility of low-grade spindle cell neoplasm. CEMRI left thigh was suggestive of a well-defined encapsulated heterogeneous mass lesion measuring 7*9*13 cm is seen in anterolateral aspect in the intermuscular plane in left upper thigh. Peripherally the mass had a solid component appearing iso to hypointense on T1 Images, hyperintense on T2/STIR images. Centrally the mass is showing a well-defined area appearing heterogeneously hyperintense on all sequences suggestive of blood degradation products. Multiple patchy areas of cystic changes, necrosis and haemorrhage were seen within the mass- likely neoplastic aetiology. D/D considered- Fibroma, Rhabdomyoma, Haemangioma. All routine blood investigation were done like CBC, LFT, KFT, PT/INR, HIV, HCV, HBsAg, ECG and chest Xray. Pre anaesthesia check-up was done plan for excision and biopsy was made. Vertical incision was given over the swelling. Skin, subcutaneous tissue cut and separated. Tensor fascia lata opened along with incision given. We found that 14*10 cm, firm swelling in anterolateral aspect of thigh in intermuscular plane. It was easily shelled out from surrounding adhesions. Wound closed without any drain. Post operatively patient was kept in post operative care room and ward where hospital stay was uneventful and patient was discharged on post operative day five. Patient came for follow up on Post op day ten, patient was hemodynamically stable with no fresh complaints, removal of sutures done. The excised specimen on histopathological examination had impression as- Neurilemmoma with extensive retrogressive changes (Ancient Schwannoma). On follow up visit after three weeks, patient was symptomatically and hemodynamically stable and she recovered without any significant complications.

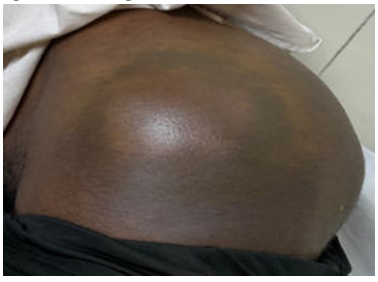


Figure 1 swelling over anterolateral aspect of thigh

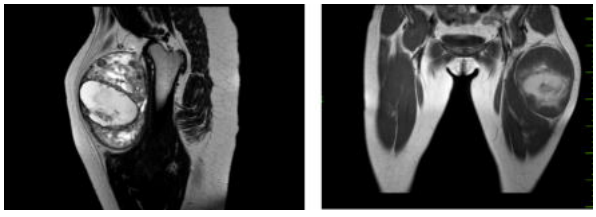
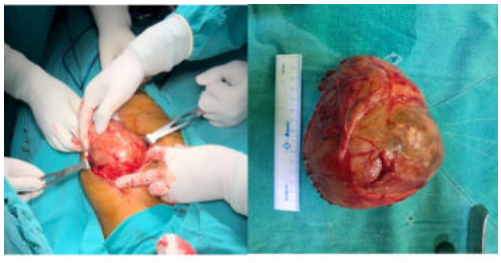


Figure 2 Well defined encapsulated heterogeneously enhancing mass lesion seen in Isointense to hypointense on T1 and Hyperintense on T2.



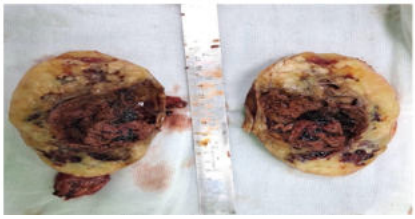


Figure 3,4,5 - Intra operative images showing the operative procedure of excisional biopsy.

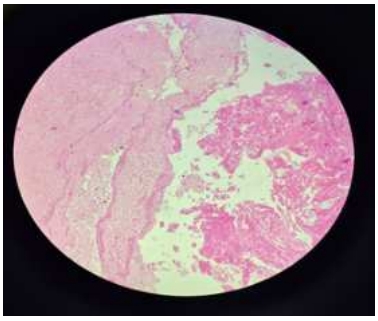


Figure 6 Low power view of H&E-stained section showing hypocellular and hypercellular areas in a myxoid background.

DISCUSSION

The first case of a solitary schwannoma was discussed by Liebau, who stated that schwannomas should be looked for in all cases where patients present with pain, paraesthesia of leg and foot, especially if all other injury has been excluded. The most common benign tumours of peripheral nerves are schwannomas and neurofibromas. Important features of schwannoma include the presence of a capsule, the eccentric position of the nerve relative to the mass, and a cystic lesion within it [4]. However, the spectrum of these neoplasms has been broadened in recent years. However, it is more common between the second and third decades of life. Schwannomas react strongly with S100 protein and immuno-histochemistry can be used to support diagnosis and to distinguish them from malignant peripheral nerve sheath tumours. This is particularly useful as the occasional appearance of hyperchromatic cells and cytological atypia may cause difficulty in establishing diagnosis especially when fine needle aspiration cytology (FNAC) is used to make a diagnosis as this technique rarely produces an adequate amount of sample. Schwannomas often have a capsule and they are located in relation to the nerve of origin[5]. They have homogeneous or heterogeneous appearance with areas of cystic degeneration (61-66% of cases) and calcifications (23% of cases) or haemorrhage on CT. When CT showed a heterogeneous lesion, the tumour can mimic pheochromocytoma or other malignant tumours. Magnetic resonance imaging (MRI) using Gadolinium contrast is superior to CT to define margins and identify the nerve origin. Schwannomas are hypointense on T1 and heterogeneous hyperintense on T2. Malignant schwannomas have irregular contour and tend to invade other structures. The accuracy of core needle biopsy is poor and diagnosis is usually made after the resection. Core needle biopsy to diagnose the schwannoma with an accuracy rate of 98% to distinguish benign from malignant tumours and an accuracy rate of 81% to identify benign tumour subtypes. Furthermore, biopsy carries a risk of tumour seeding in case of the malignant processes[8].

CONCLUSION-

This case highlights the diagnostic challenges and treatment considerations associated with schwannomas. Schwannomas are rare tumours that may be difficult and incidentally to diagnose. This patient presented with swelling over anterolateral aspect of thigh, after core needle biopsy report a diagnosis of low-grade spindle cell neoplasm was made. Surgical excision was undertaken successfully. Our experience underscores the importance of CEMRI, Core needle biopsy in diagnosing and managing schwannomas. This case contributes to the understanding of treatment efficacy or diagnostic accuracy and emphasizes the need for further considerations, e.g., follow-up, management. Malignant transformation of the tumour should always be kept in mind, and careful histopathological examination should be done to rule out malignancy.

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