

A Rare Case of Ancient Schwannoma Mimicking Malignancy

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Abstract: *The schwannoma is a rare benign neural tumor, arising from the neural sheath Schwann cells of the peripheral, cranial, or autonomic nerves. Ancient schwannomas are long standing tumors with degenerative changes, found in the head, neck and flexor surfaces of extremities. Thus an 'ancient' variety of a rare tumor makes diagnosis furthermore challenging. We report a case of a 60 year old female with a large swelling over the thigh mimicking a malignant tumor. The diagnosis was established on clinical, radiological and histopathological findings.*

Keywords: Ancient schwannomas, neural tumor

1. Introduction

Schwannomas are rare, encapsulated tumours arising from the neural sheath Schwann cells of the peripheral, cranial, or autonomic nerves and are benign in nature. Schwannoma with pronounced degenerative changes is known as 'Ancient' schwannoma, which is a rare variant of schwannoma and is usually a deeply situated large mass of long duration, representing 0.8% of all soft-tissue tumours. They usually arise in females between the ages of 20-50 years, with a predilection for the head, neck and flexor surfaces of extremities.

2. Case Report

A 60 year old female reported to the Department of Surgery, SGRM IM & HS, Dehradun with solitary painless ovoid swelling over the right upper thigh for a few years, with no significant history of trauma. The swelling had significantly increased in size causing patient discomfort for the past 6 months which led her to seek surgical consultation. Patient was a known case of coronary artery disease, chronic kidney disease and psychosis; on medications. The swelling was firm approximately 10 x 8 cm with a smooth surface with well defined margins and had restricted mobility but showed no sign of fixity to the underlying bone or any signs of inflammation. Skin over swelling was normal with no sinuses or discoloration or loss of skin appendages but the patient complained of reduced sensations over the medial side of the thigh extending up to the knee with intact motor functions.

3. Management

The patient underwent a MRI scan which suggested, evidence of large well defined heterogeneous signal area over the medial aspect of right upper thigh involving the adductor compartment muscles, extending to subcutaneous tissue measuring approx 11.4 x 9.7 x 8.2 cm. Periphery of the lesion appeared hypointense on T2 and STIR sequences and isointense on T1 sequences. The lesion shows multiple thin internal septations with a central cystic part appearing hyperintense on T2 and STIR sequence and mildly

hyperintense signal on T1 sequence. There was no evidence of any obvious bony erosion [image 1]. On USG correlation, no obvious vascularity was noted in the lesion. The differential diagnosis of intramuscular haematoma or a low grade neoplasm were suggested. We went forward with an invasive procedure to confirm the diagnosis, FNAC revealed granular necrotic debris with a single focus of cluster of plump spindle cells mixed with a few lymphocytes and neutrophils, which could not confirm the diagnosis. An excisional biopsy was planned as the lesion was not suggestive of any malignant etiology and showed no invasion to the surrounding areas. Intra-operatively an intermuscular encapsulated globular soft tissue mass was encountered with attachment to the periosteum of pubic bone which was surgically excised in toto under spinal anaesthesia [image 2]. The microscopic examination showed an encapsulated spindle cell neoplasm exhibiting hypercellular and hypocellular areas. The hypercellular areas composed of wavy bundles of spindle cells showing mild cellular atypia, however no mitosis was seen. The hypocellular areas show loosely woven spindle cells. Large areas of collagenization, cystic changes and haemorrhages with scattered cyst macrophages and siderocytes were seen with a few thick walled blood vessels noted in the parenchyma. Nerve bundles were seen attached to the periphery of the capsule along with fibrofatty tissue confirming the diagnosis of Ancient Schwannoma. [image 3] Post operative period of the patient was uneventful. The suction drain was removed on the 3rd post operative day and the patient was discharged on the 5th post operative day in satisfactory condition. Regular follow up showed no further decrement of sensations over the thigh, stitch site is healthy with no signs of recurrence.

4. Discussion

It was found in 1951 by Ackerman and Taylor that schwannomas presented with clear areas of hypocellular tissues and accredited this to the long standing degenerative changes. Hence, they coined the term "ancient" schwannoma for such type of benign neurogenic tumor. These degenerative features are due to the growth and "aging" of the tumor, therefore the term "Ancient

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schwannoma". Regardless of these degenerative changes, ancient schwannomas behave similar to schwannoma. They are benign, slow-growing tumors with rare malignant transformation.

They exhibit spindle cells with focal nuclear palisading patterns^[1] arranged in distinctive dense (Antoni A) and loose (Antoni B) areas^[1, 2]. Usually, there is presence of connective tissue fragments (Verocay bodies) and intranuclear vacuoles (lochkern)^[3-5].

Antoni A areas are more organised, hypercellular, and made up of spindle cells arranged in short bundles or interlacing fascicles. On the other hand, Antoni B regions are hypocellular, less organised and contain more myxoid, loosely arranged tissue, with a high water content. There is intermixing of these components within schwannomas in varying amounts^[6]. Schwannoma is a slow-growing, benign tumour or a large tumour with degeneration, especially when it is situated in the deep seated regions like the mediastinum and retroperitoneum^[7]

Also, as the tumours are frequently infiltrated by large numbers of siderophages and histiocytes, and exhibit cellular degenerative changes, including nuclear atypia and pleomorphism, along with a tendency to nuclear palisading^[4, 6], malignancy may be incorrectly diagnosed^[8].

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^[9-11]. This is particularly useful as the occasional appearance of hyperchromatic cells and cytological atypia may cause difficulty in establishing diagnosis^[12,13] especially when fine needle aspiration cytology (FNAC) is used to make a diagnosis as this technique rarely produces an adequate amount of sample.^[14,15] False positive diagnosis of malignancy may lead to over treatment with radical resections being carried out without need.

Since the tumour contains cystic areas, ancient schwannoma has been radiologically misdiagnosed as other tumour types, like malignant fibrous histiocytoma, malignant peripheral nerve sheath tumour, liposarcoma, synovial sarcoma or haemangiopericytoma. Though, only a handful of reports have dealt with the radiological features of ancient schwannoma in the thigh as the tumour is so rarely encountered^[16]. On MRI, the peripheries of ordinary schwannomas yield low signal intensities on T1 weighted images and high-signal intensities on T2 weighted images, corresponding to the Antoni B region, and low-to-intermediate signal intensities in other areas on T1 and T2 weighted images with strong enhancement by gadolinium contrast media, characteristic of the Antoni A regions^[16].

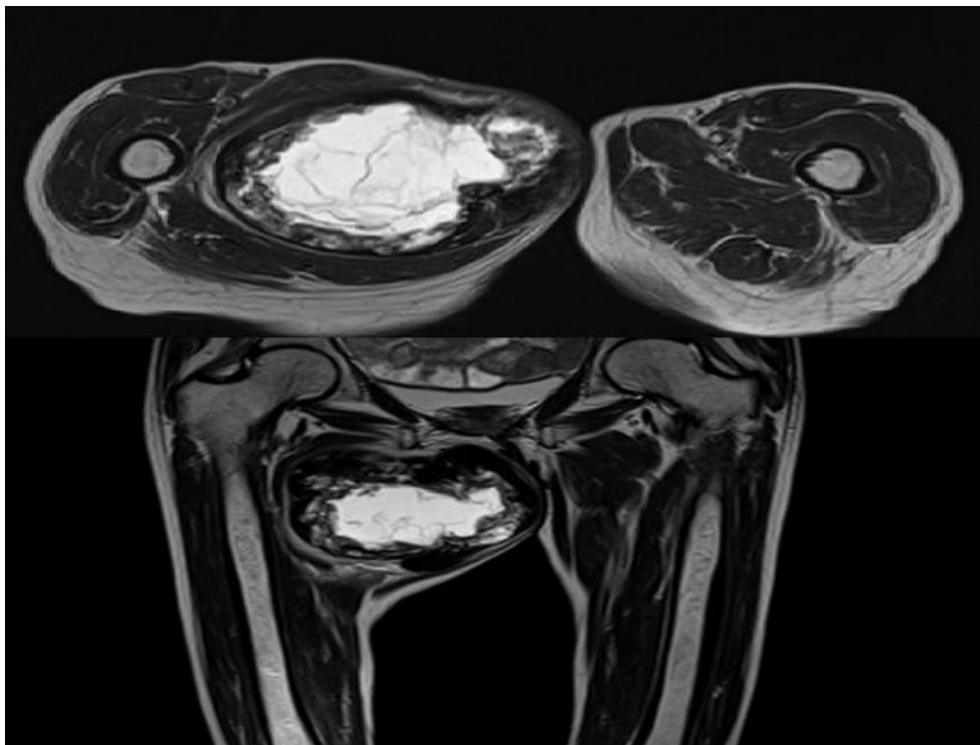


Image 1: MRI scan suggesting evidence of a large well defined heterogenous signal area over the medial aspect of right upper thigh. Periphery of the lesion appeared hypointense on T2 and STIR sequences and isointense on T1 sequences. The lesion shows multiple thin internal septations with a central cystic part appearing hyperintense on T2 and STIR sequence and mildly hyperintense signal on T1 sequence.



Image 2: Images showing the operative procedure while performing the excisional biopsy.

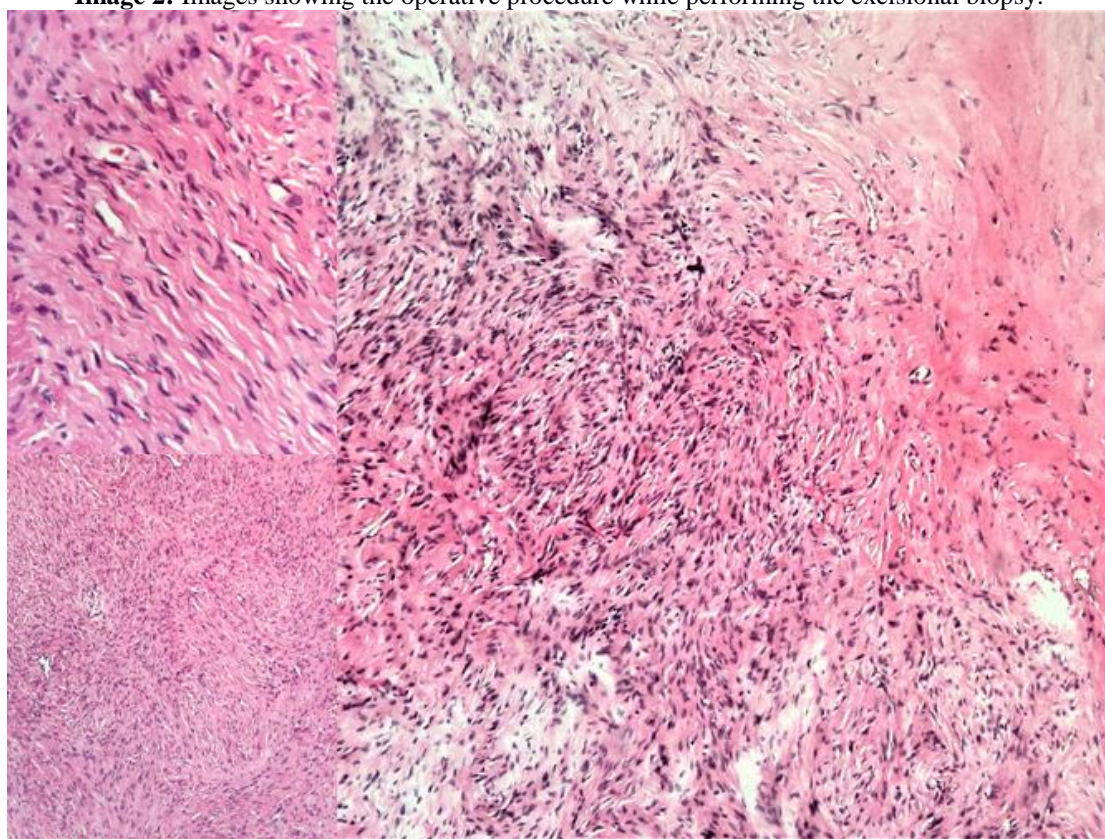


Image 3: Microscopic examination showing an encapsulated spindle cell neoplasm exhibiting hypercellular areas composed of wavy bundles of spindle cells showing mild cellular atypia, and hypocellular areas showing loosely woven spindle cells

5. Declarations

- Funding: Self
- Conflict of interest: None
- Ethical approval: Approved from Institutional Ethical Committee

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