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Giant Ancient Solitary Schwannoma Masquerading as Juxtacortical Osteosarcoma of Femur - A Rare Case Report and Literature Review

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Abstract

Introduction: Ancient schwannomas are rare variant of peripheral nerve sheath tumors characterized by the degeneration and hypocellular areas due to long-standing growth. Clinicoradiologically, these tumors can masquerade other tumors arising from the adjacent tissues. Their resemblance to malignant bone tumor has been reported very rarely in the literature. We tend to report a case of benign peripheral nerve schwannoma that greatly mimicked a juxtacortical osteosarcoma of femur.

Case Report: A 23-year-old male presented with a slow-growing painless mass with paresthesias in his right thigh for the last 2½ years. Clinically, it was suspected to be soft tissue tumor with secondary involvement of adjacent neurovascular bundle; however, plain radiograph and magnetic resonance imaging of his right thigh were suggestive of juxtacortical osteosarcoma of the right femur. Surgical exploration of the mass revealed a well-defined encapsulated mass over the anterior aspect of the right thigh, under the quadriceps muscle without infiltration into the surrounding tissue. Histopathological examination confirmed it to be an ancient schwannoma.

Results: The patient was extremely satisfied with outcomes of surgery, and he was symptom-free and there was no clinical evidence of the recurrence on subsequent follow-up.

Conclusion: A correct pre-operative diagnosis of benign peripheral nerve sheath tumors can be difficult at times. However, a slow-growing mass with the absence of other features of a malignant growth and subsequent histopathological examination including immunostaining can settle the diagnosis in almost all the cases.

Keywords: Benign, Peripheral nerve, Ancient schwannoma, Juxtacortical osteosarcoma.

Introduction:

Ancient schwannoma or degenerative neurilemmoma refers to a rare subtype of schwannoma characterized by degeneration and hypocelluar areas due to long-standing growth [1]. Ancient schwannomas can be misdiagnosed as sarcomas and other soft tissue tumors due to its clinical and imaging features [2] as well as degenerative changes [3] seen on cytopathological examination. Ancient schwannomas have been reported to arise from sural nerve, plantar digital nerve, and tibial nerve, lateral cutaneous nerve of thigh, sciatic nerve [3], cervicothoracic spinal nerve roots [4], retroperitoneal area [5], and parotid gland [6]. Lee et al. [7] have reported two cases of ancient schwannoma of thigh mimicking malignancies. The other important clinical differential diagnoses in such cases include Ewing's sarcoma and surface osteochondroma. To the best of our knowledge, no case report has been reported in the literature of an ancient schwannoma of thigh mimicking a juxtacortical osteosarcoma of femur. Here, we report a case of 3-year-old male who presented with slow-growing tumor mass in his right thigh masquerading juxtacortical osteosarcoma of femur due to the calcification of the tumor mass adjacent to the femoral shaft.

Case Report

A 23-year-old male patient presented with a painless slow-growing mass over the front of his right thigh for the last 2½ years. The swelling was associated with gradually progressive paresthesias including tingling and numbness over right thigh. There was no history of trauma, fever, weight loss, appetite loss, similar previous, and/or recurrent swelling in the same or different anatomical locations. Clinical examination of the swelling revealed a single well-defined, non-tender soft to firm mass measuring 10 cm × 8 cm, located predominately over the anterior aspect of the right thigh, with some mediolateral extension. The mass was extending from the proximal third to middle third of the thigh; it was freely mobile along the transverse axis of thigh but had very limited mobility along the long axis of the thigh. Tinel's sign was present over the swelling. The overlying skin was healthy and mobile, no superficial dilated veins seen. The range of motions of

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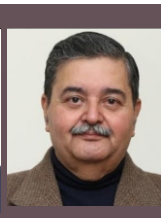
Dr. Ajay Pant

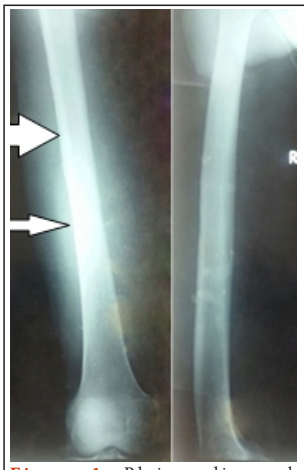

Figure 1: Plain radiograph anteroposterior and lateral views of the right thigh showing prominent unmineralized soft tissue mass with periosteal reaction over middle third of right femur.

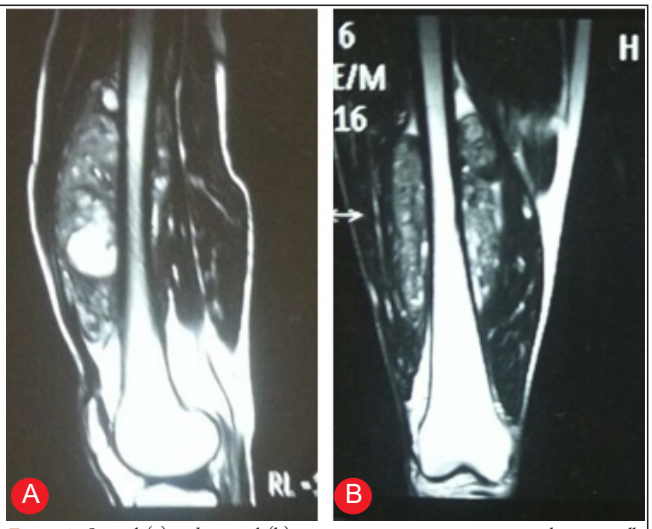

Figure 2: Sagittal (a) and coronal (b) magnetic resonance imaging cuts showing well-defined juxtacortical altered signal intensity mass along anterior cortex of midfemur, with cortical buttressing and periosteal reaction.

the ipsilateral hip and knee was within normal limits. There was no limb length discrepancy. There was no neurological deficit, and the right posterior tibial and dorsalis pedis artery were palpable and comparable to the normal extremity. A plain radiograph of the right thigh anteroposterior and lateral views showed a prominent soft tissue shadow present over the anterolateral aspect of the middle third of the thigh, along with a prominent periosteal reaction over the medial aspect of middle third of the femur (Fig. 1). The magnetic resonance imaging (MRI) sagittal (Fig. 2a) and coronal (Fig. 2b) cuts showed a well-defined, lobulated, and midthigh juxtacortical heterogeneous altered intensity mass measuring 7 cm × 8 cm × 16 cm, along anterior cortex of midfemur, with cortical buttressing and periosteal elevation. There was no loss of cortical hypointensity. Medullary cavity was unremarkable. The mass was closely abutting the quadriceps muscle. The radiologist was of the opinion of juxtacortical osteosarcoma of the right femur; however, there was no infiltration into the surrounding soft tissues and no lymphadenopathy. A true cut core biopsy from multiple sites was suggestive of a benign peripheral nerve tumor. Intraoperatively, we found a well-defined gray-white ovoid, soft-to-firm tumor mass measuring 5.5 cm × 8.5 cm × 16 cm in the anterolateral aspect of the right thigh (Fig. 3a). The surface of the tumor mass abutting the shaft of the femur had an impression of the shaft of femur (Fig. 3b). Gross examination of the received specimen showed an ovoid, gray-white, soft-to-firm tumor mass measuring 16 cm × 8.5 cm × 5.5 cm. Cut sections showed white lobulated areas with cystic changes. Microscopic examination showed encapsulated neoplasm having two component predominantly hypocellular areas (Antony B) and occasional hypercellular areas (Antony A). Hypocellular areas showed elongated wavy nucleus arranged end-to-end suspended in clear myxoid matrix. Blood vessels were also seen. Hypercellular areas consisted of monomorphic spindle-shaped cells with poorly defined eosinophilic cytoplasm and pointed basophilic nuclei arranged in interlacing fascicles pattern. All these findings confirm it to be schwannoma. Postoperatively, the patient recovered well and at his last follow-up there were no symptoms related to the previous tumor mass, and the patient was extremely satisfied with the surgical results.

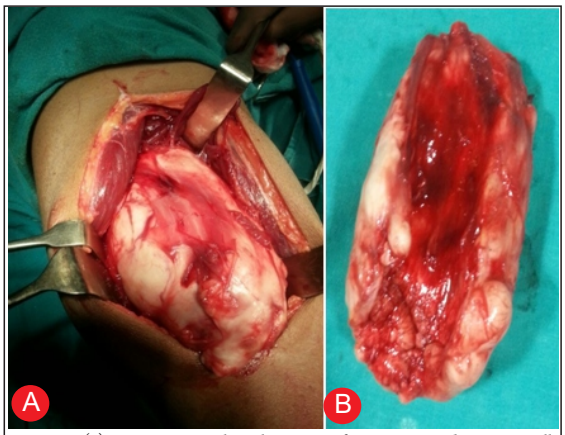

Figure 3: (a) Intraoperative clinical pictures of same patient showing a well-defined gray-white ovoid mass in the anterolateral aspect of the right thigh. (b) Clinical pictures of excised specimen showing soft to firm tumor mass, with an impression of the shaft of femur over tumor mass due to close proximity to femoral shaft.

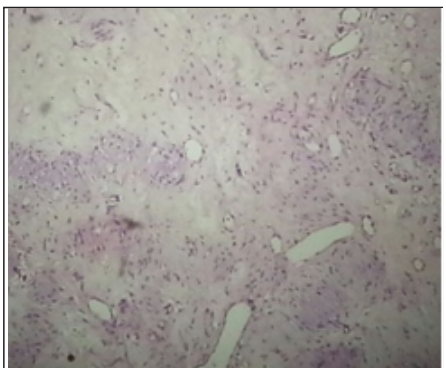

Figure 4: Photomicrograph showing hypocellular Antony Type A pattern with verocay bodies.

Discussion

The term "ancient neurilemmoma" was first used by Ackerman and Taylor [8]. Ancient schwannoma represents the rare subtype of long-standing schwannomas lying in deeper tissues, and characterized by prominent degenerative changes [9, 10], and represent 0.8% of all soft tissue tumors. Microscopically, ancient tumors are characterized by predominantly hypocellular Antony Type B areas, cystic changes, and microcalcifications. Due to prominent degenerative changes, these tumors have been radiologically misdiagnosed as other tumor types including malignant fibrous histiocytoma and liposarcoma. In our patient, plain radiographs of the femur show a clear radiodense shadow mimicking periosteal reaction at the middle third of the femur (thin arrow, Fig. 1), surrounded by a large fusiform soft tissue shadow (Thick arrow, Fig. 1). In our case, calcification of the degenerated tumor mass and its close proximity to the femoral shaft as evidenced by impression of the shaft of femur over the tumor mass could be responsible for the radiopaque shadow on the plain radiograph giving the impression of periosteal reaction over the middle of the femur. The same reason could be responsible for the misinterpretation of the tumor mass as juxtacortical osteosarcoma of femur on MRI. However, intraoperatively, the tumor mass was not infiltrating into the surrounding soft tissues and it was

easily separable as well as the gross morphological appearance of the tumor mass in the thigh was not like a juxtacortical osteosarcoma of femur. The classical form of peripheral nerve schwannomas is characterized by the presence of hypercellular Antony Type A and hypocellular Antony Type B areas with occasional verocay bodies with varying proportions [11]. The final diagnosis of ancient schwannoma was confirmed on histopathological examination showing predominant hypocelluar Antony Type B pattern with degenerative changes and occasional verocay bodies, with less prominent cellular Antony Type B areas (Fig. 4). There are three different forms of juxtacortical osteosarcoma, the periosteal osteosarcoma, parosteal osteosarcoma, and high-grade surface osteosarcoma that together constitute 4–10% of all osteosarcoma [12]. These tumors have male predominance, femur and tibia are common bone involved and the mean age of presentation is 20 years [13, 14]. Case described in the present manuscript has all the three demographic properties described for juxtacortical osteosarcoma. Moreover, within the femur periosteal osteosarcoma predominate in the anteromedial portion of the diaphysis as is seen in our case [15]. Therefore, based on the above discussion, juxtacortical osteosarcoma is a valid differential diagnosis in our case. Apart from juxtacortical osteosarcoma, the important clinical differential diagnoses of the present case include Ewing's sarcoma and juxtacortical chondrosarcoma. However, further, diagnostic workup including the imaging investigations, histopathological examination, and immunohistochemistry can help reaching the exact diagnosis in almost all the case.

Conclusions

Ancient schwannoma should be kept as a differential diagnosis for any tumor mass presenting in the thigh, to avoid missing this potentially curable benign soft tissue tumor.

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