



Research paper

Schwannoma of the femur: A rare case report

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A B S T R A C T

Schwannoma is a benign tumor derived from Schwann cells that cover myelinated nerve fibers (1). Among primary bone tumors, intraosseous schwannoma accounts for less than 0.2% (2). We are reporting a rare case of an 18-year-old female presented with swelling in the posterior right thigh. The patient was asymptomatic and this was incidentally discovered by her massage therapist. However, the swelling was gradually growing since then.

The plain radiograph findings were suggestive of a benign bone tumor. MRI with gadolinium was performed for further evaluation but it demonstrated suspicious features as evidenced by the extra osseous enhancing soft tissue component. Three phase bone scan was done thereafter and it showed hyperemia with mild increased uptake of the lesion on the delayed phase. Osteosarcoma was suspected and chest CT was performed for staging purposes which turned out to be negative for metastasis.

The lesion was biopsied under ultrasound guidance. The histologic features and the immunohistochemically profile were consistent with a benign schwannoma.

The posterior thigh soft tissue mass was surgically resected with femur bone grafting. Recurrence of schwannoma should be considered but its malignant transformation is exceedingly rare and this can be discounted (3).

1. Introduction

Schwannoma is a benign tumor of neuroectodermal derivation that originates from Schwann cells which cover the peripheral nerves [3].

Approximately 25–40% of all schwannomas occur in the head and neck. These tumors most commonly arise in the soft tissues of the head and neck and on the flexor surfaces of the upper and lower extremities [3].

Intraosseous schwannoma is a slow growing tumor and the patient is usually asymptomatic. Swelling is the most common complaint at presentation. Intraosseous schwannoma is a rare entity and in this study we are reporting a case of a schwannoma in the femur [3].

2. Case Report

This is an 18-year-old female presented with swelling in the posterior aspect of the right thigh. There was no pain and this was incidentally noted by the patient's massage therapist. However, the swelling was gradually increasing in size since it has been depicted.

On physical examination, the mass was palpable at the posterolateral aspect of the mid to distal right thigh. The mass was fixed and firm but it was not painful to deep palpation. No lymph nodes were

detected in the popliteal fossa. There were no overlying skin changes. Distal neurovascular examination was normal.

Plain radiographs were done for initial assessment and it showed a bubbly appearing mildly expansile cortically based lucent lesion with sclerotic margins in the distal right femur diaphysis posteriorly but with no cortical breakthrough, aggressive looking periosteal reaction or obvious soft tissue component suggesting a benign non-ossifying fibroma.

The MRI revealed a cortically based tumor in the distal right femur diaphysis posteriorly associated with an exophytic soft tissue mass. The lesion showed low signal intensity on T1WI and predominantly high signal intensity on the fluid sensitive sequence with avid enhancement following gadolinium administration.

Three phase bone scan was performed demonstrating mild diffuse increased radiotracer accumulation at the distal right thigh on the angiographic and blood pool imaging indicative of mild hyperemia. The delayed phase images showed mild increased uptake of the lesion at the distal right femur diaphysis.

This mass was biopsied under ultrasound guidance and the histologic features as well as the immunohistochemically profile were consistent with a benign nerve sheath tumor, schwannoma.

Surgery was done and the posterior thigh soft tissue mass was

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Fig. 1. Lateral projection radiograph showing bubbly appearing mildly expansile cortically based lucent lesion with sclerotic margins in the distal right femur diaphysis posteriorly.

resected with femur bone grafting. No postoperative complications were encountered.

3. Discussion

Schwannoma is a slow-growing benign tumor derived from Schwann cells, the sheath cells that cover myelinated nerve fibers [1]. Patients with schwannoma are usually symptom-free and swelling is the most common complaint at presentation [3]. Intraosseous schwannomas are rare and they account for less than 0.2% among the primary bone tumors.

The preoperative diagnosis of intraosseous schwannoma is challenging because of its rarity [2].

Schwannomas can affect the bone either by an extra osseous soft tissue mass that causing bony erosion or it can arise from nerves that course through a canal in a bone causing erosion of the bone and creating a dumbbell-shaped configuration. Less commonly, schwannoma can originate centrally from the medullary cavity of the bone [4].

In our case, the radiographic features were not suspicious whereas the MRI findings were concerning for malignancy. Three phase bone scan was done which showed mild hyperemia with mild increased uptake of the lesion on the delayed phase. CT of the chest was also performed for staging purposes and it did not show metastases to the lungs. Subsequently, the lesion was biopsied under ultrasound guidance.

The histologic sections showed a mildly hyper cellular spindle cell lesion. The spindle cells were seen within dense fibrous or loose fibrous areas. There was prominent nuclear palisading (Verocay bodies).

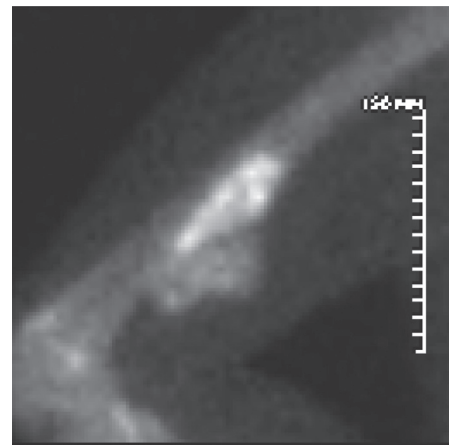


Fig. 3. Delayed phase bone scan spot image of the right femur showing mild increased radiotracer uptake by the bone tumor and the soft tissue component.

Occasional moderately pleomorphic nuclei were present due to degenerative (ancient) changes. No mitoses were identified. The immunohistochemically studies showed the lesion was positive for S-100 protein and focally positive for CD34. The lesion was negative for nuclear Beta-catenin, MUC-4, STAT-6, desmin and smooth muscle actin. All of these findings were consistent with schwannoma.

This was reviewed at multidisciplinary sarcoma rounds. This is quite an unusual case of a bone cortically based nerve sheath tumor with a large soft tissue component. The pathology was reviewed for both interpretation and to ensure there were no sampling or administrative errors. Once it was confirmed after multidisciplinary discussion, the patient was brought to the operating theater for resection of the posterior thigh soft tissue mass with bone grafting femur as required (Figs. 1–4).

The procedure was tolerated well without any post-operative complications. In general, schwannomas have a negligible malignant potential and a low tendency to recur, however, long-term follow-up is recommended to observe the biological behavior of this intraosseous schwannoma.

4. Conclusion

We are reporting a rare case of an intraosseous schwannoma in the right femur on the basis of imaging features, histopathological pattern and immunohistochemically profile. Pre-operative diagnosis was confirmed by tissue biopsy which is crucial for optimal management plan and to avoid unnecessary adjuvant therapy.

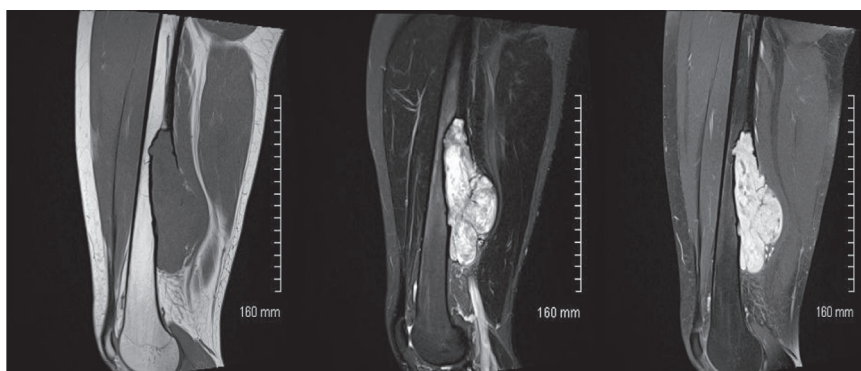


Fig. 2. MRI showing low signal intensity of the lesion on T1WI and predominantly high signal intensity on the fluid sensitive sequence with avid enhancement following gadolinium administration.

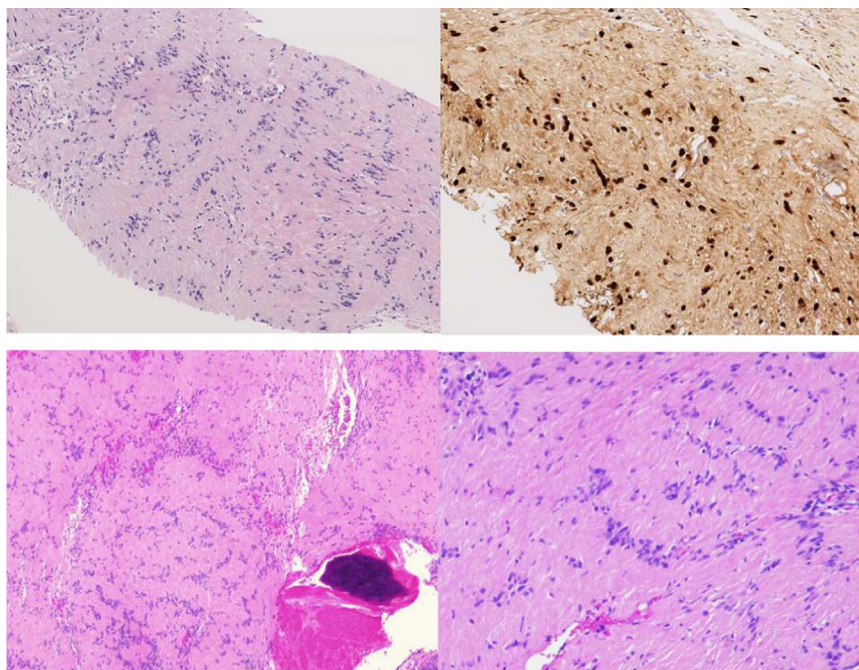


Fig. 4. The histologic sections showing a mildly hyper cellular spindle cell lesion. The spindle cells are within dense fibrous or loose fibrous areas. There are prominent nuclear palisading (Verocay bodies). Occasional moderately pleomorphic nuclei are present due to degenerative changes. There are no mitoses. The immunohistochemically studies show the lesion is positive for S-100 protein and focally positive for CD34.

Conflict of interest statement

I have no conflict of interest to declare. I warrant that the article is the Author's original work. I warrant that the article has not received prior publication and is not under consideration for publication elsewhere.

References

- [1] Hina Zainab, Alka D. Kale, Seema Hallikerimath, Intraosseous shwannoma of the

- mandible, JOMFP 16 (2) (2012) 294–296.
 [2] H. Igai, M. Kamiyoshihara, N. Kawatani, T. Ibe, K. Shimizu, Sternal intraosseous schwannoma mimicking breast cancer metastasis, J. Cardiothorac. Surg. 9 (2014) 116.
 [3] Goro Kawasaki, Souichi Yanamoto, Hajime Yoshida, Akio Mizuno, Shuichi Fujita, Tohru Ikeda, Intraosseous shwannoma of the mandibular symphysis: report of a case, Oral. Sci. Int. 7 (2) (2010).
 [4] Pelin BAĞCI1, Sergülen DERVİŞOĞLU1, Murat HIZ2, Kaya KANBEROĞLU, Intraosseous schwannoma of the radius, Turk. J. Pathol. 26 (2) (2010).