Case Report

Spindle cell hemangioma of femur

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ABSTRACT

Spindle cell hemangioma (SCH) an uncommon vascular tumor typically appears as solitary or multiple cutaneous and subcutaneous nodules in middle-aged adults. Its occurrence in bone is extremely rare. We report a case of SCH arising at an uncommon site, the distal femur with radiological local invasion, but without histological pleomorphism and mitosis. It is important to avoid misdiagnosis as these lesions are considered to be benign, non-neoplastic reactive vascular proliferations, with high incidence of recurrence.

Key words: Bone tumors, femur, spindle cell hemangioma, vascular tumors

INTRODUCTION

Spindle cell hemangioma (SCH) or hemangioendothelioma was described as a low-grade angiosarcoma resembling cavernous hemangioma and Kaposi sarcoma.[1-4] Later, it has been reported as non-neoplastic reactive vascular proliferations.[1,2,5,6]

Spindle cell hemangioma almost exclusively affects the dermis and subcutis of extremities.[2-6] Occurrence of SCH in bones is extremely rare, and only three reports are found in English literature. In one case the radius and ulna and in another, the medial malleolus was involved, and both cases were reported to be local spread from the overlying skin and subcutaneous tissue.[2,4] The third case involved the frontal bone.[7] We report a 53-year-old male with SCH involving the distal femur and adjoining vastus intermedius and lateralis muscles with normal overlying skin and subcutaneous tissues.

CASE REPORT

A 53-year-old male presented with dull aching pain over right distal thigh since 2 months. Pain was insidious in onset, progressive in severity and associated with difficulty in weight bearing. There was no preceding trauma or constitutional symptoms. General and systemic examinations were normal. Local examination revealed tenderness over the lateral aspect of right distal thigh without swelling, deformity or irregularity of the underlying bone. The skin and subcutaneous tissue over the thigh and range of movements of the hip and knee were normal with intact distal neurovascular status. The blood counts, serum calcium, phosphorous, alkaline phosphatase and protein electrophoresis were normal. Radiographs showed an ill-defined lytic area in the distal metadiaphysis of right femur without obvious soft tissue swelling or intralesional calcification. The knee joint was normal [Figure 1]. Bone scan showed mixed osteoblastic and osteolytic changes over the distal third of right femur. Magnetic resonance imaging of the right thigh showed an ill-defined altered signal intensity lesion involving the lateral and anterior cortex and adjacent marrow in the distal third of right femur with minimal linear periosteal reaction and focal cortical discontinuity along the lateral aspect. Hyperintensity involving the vastus lateralis and intermedius muscles which enhanced in post-contrast sequences were noted in T2 weighted images. There was no involvement of the neurovascular bundle [Figure 2].

Incisional biopsy was taken from the lesion that demonstrated a neoplasm composed of proliferating vascular channels arranged in nodules with an intervening fibrocollagenous stroma. Vascular channels were closely packed, and thin-walled with a lining of flattened epithelioid endothelial cells. Spindle cells with elongated slender nuclei and eosinophilic cytoplasm were seen. However, no significant mitotic activity or necrosis was seen. Sections

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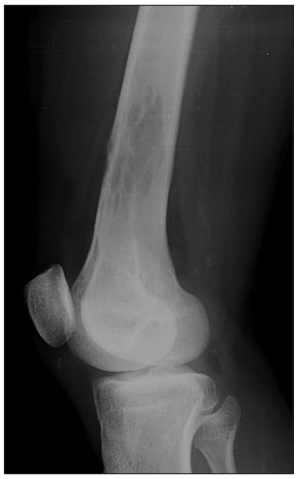

Figure 1: Lateral radiograph of the right distal femur showing the ill-defined lytic lesions in the metadiaphysis with erosion of the anterior cortex without periosteal reaction and normal knee joint

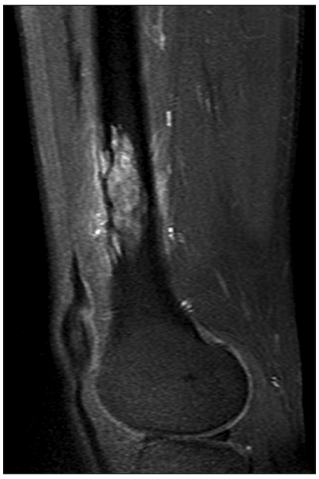

Figure 2: Sagittal magnetic resonance image of the right distal femur showing lesion involving the anterior cortex and adjacent marrow, minimal periosteal reaction, cortical discontinuity and heterogenous changes in the surrounding muscles

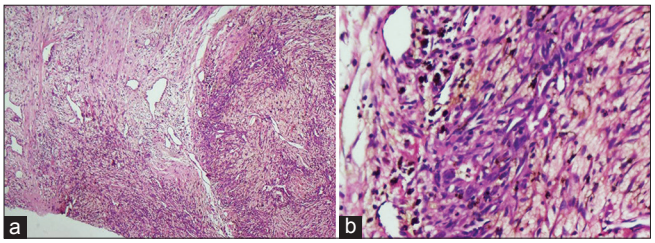

Figure 3: (a) Low power (H and E, ×10) and (b) high power (H and E, ×40) slides showing proliferating vascular channels lined by flattened endothelial cells, intermingled with spindle cells with elongated slender nuclei and eosinophilic cytoplasm

from marrow showed lamellar bone [Figure 3]. On immunohistochemistry (IHC) the endothelial cells stained positive for CD34 while the spindle cells did not. On this basis, a diagnosis of SCH was confirmed. Wide excision of the tumor and prophylactic intramedullary nailing of the right femur was done [Figure 4]. He is asymptomatic without any evidence of recurrence 3 years following surgery [Figure 5].

DISCUSSION

Spindle cell hemangioma is commonly reported in middle aged adults. Solitary lesions tend to be more common in males and multifocal in females.[5] Most cases of SCH are sporadic in nature, but multifocal lesions are associated with Maffuci syndrome, Olliers disease, chronic lymphedema, Klippel-Trenaunay syndrome, von Willibrand disease, acute myelomonocytic leukemia and early onset varicose veins.[1,5,6]

Spindle cell hemangioma lesions present as slow growing solitary or multiple nodules almost exclusively in the dermis and subcutis of the distal extremities and rarely affects the proximal extremities, head, neck, axilla, trunk, penis, spleen and pancreas.[2,5,6] The occurrence of this lesion in the bone is extremely rare. SCH involving the skin and subcutaneous tissue with extension onto the bones (medial malleolus and distal radius and ulna respectively) were reported.[2,4] In our patient, the superficial skin and subcutaneous tissues were normal, and intramuscular extension from the primary bony lesion was noted.

Majority of the lesions occurs entirely or partially intravascular, and the intravascular growth contributes to multiplicity and high rate of local recurrence.[1,6]

Histologically the lesion is characterized by cavernous vascular proliferations lined by flattened endothelial cells alternating with cellular areas consisting of spindled cells.[1,6] The endothelial lining appears epithelioid or histocytoid with round or polygonal cells and frequently have intracytoplasmic vascular lumens. The thin walled cavernous spaces are either dilated, partially collapsed or filled with blood, organizing thrombi or phleboliths.[3] SCH generally lacks significant nuclear atypia and show no or low level of mitotic activity and the distribution and percentage of the main histologic components may be highly variable.[1,6]

Immunohistochemically, the endothelial lining cells and the histocytoid cells with vacuolated cytoplasm stain positive for CD31, CD34, factor VIII related antigen, vimentin, *Ulex europaeus* lectin and HAM-56. In contrast, the spindle cells are negative for endothelial markers and react for vimentin, actin, desmin, factor VIII related antigen, MAC 387 and HAM-56.[5,6]

The differential diagnosis of SCH includes Kaposi sarcoma, cavernous hemangioma, epithelioid hemangioma, intravascular papillary endothelial hyperplasia, Kaposiform hemangioendothelioma, angiosarcoma and epithelioid and spindle cell hemangioma.[1,3,5,8,9] [Table 1].

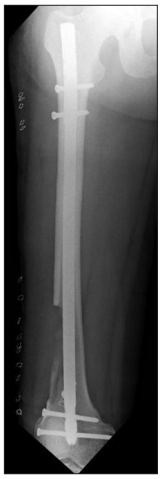

Figure 4: Immediate postoperative anteroposterior radiograph of the right femur showing the excised anterior and lateral segment of the distal femur with interlocking nail *in situ*

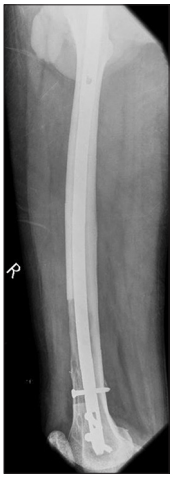

Figure 5: Lateral radiograph of the right femur at 3 years follow-up showing reformation of the anterior cortex of the distal femur without evidence of local recurrence or satellite lesions

Table 1: Differential diagnosis of SCH

Disease	Salient differentiating features from SCH
Kaposi sarcoma	More infiltrative growth, nuclear atypia, hyaline globules, higher mitotic rate Lack epithelioid endothelial cells IHC positive for HHV 8 latent nuclear antigen Spindle cells are CD34+
Cavernous hemangioma	Spindle cells are absent
Epithelioid hemangioma	Solid appearance, infiltrative pattern, myxohyaline matrix Lacks cavernous spaces and spindle cells
Intravascular papillary endothelial hyperplasia	More acellular, has many papillae Spindle and epithelioid endothelial cells are absent
Kaposiform hemangioendothelioma	Form distinct glomeruloid nests
Angiosarcoma	Infiltrative growth pattern, nuclear atypia, higher mitotic rate Shows atypical mitotic figures
Epithelioid and spindle cell hemangioma	Epithelioid cells scattered within spindle area Epithelioid cells protrude into vascular lumen in tombstone fashion Spindle and epithelioid cells stain positive with antibodies to vimentin, factor VIII and *Ulex europaeus* Spindle and epithelioid cells do not stain with desmin, keratin and S 100

IHC: Immunohistochemistry, HHV: Human herpes virus, SCH: Spindle cell hemangioma

Though spontaneous remission of SCH is rarely reported, wide excision of the lesion showed excellent prognosis for single or small multiple lesions.[5,6,10] However, recurrence may develop even many years after the initial excision.[1,3-5] Treatment of extensive multiple lesions is mainly conservative as there is no evidence for metastatic potential. Low-dose interferon alpha 2b and intralesional and intra-arterial administration of recombinant interleukin 2 have been successful in treating and preventing recurrence of inaccessible or multiple SCH.[5,6] Radiotherapy should be considered cautiously due to possible malignant transformation.[3,10] Patients having SCH in association with Maffuci's syndrome are at increased risk for development of chondrosarcoma and other neoplasms due to dyschondroplasia.[6]

CONCLUSION

Emphasis needs to be given on proper histopathological and immunohistochemical examination of vascular bony lesions to avoid misdiagnosis of benign tumors like SCH as a more aggressive vascular tumor. SCH of bones can also occur in elderly patients and can primarily involve the bone without affection of the skin and subcutaneous tissues. Wide excision of the lesion can give excellent results.

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REFERENCES

1. Sheehan M, Roumpf SO, Summerlin DJ, Billings SD. Spindle cell hemangioma: Report of a case presenting in the oral cavity. J Cutan Pathol 2007;34:797-800.
2. Chung DH, Keum JS, Lee GK, Kim CJ, Park SH. Spindle cell hemangioendothelioma – A case report. J Korean Med Sci 1995;10:211-5.
3. Ganapathy B, Ramesha N, Bhat VA. A case of spindle cell haemangioendothelioma. Indian J Cancer 1994;31:274-7.
4. Lai FM, Allen PW, Yuen PM, Leung PC. Locally metastasizing vascular tumor. Spindle cell, epithelioid, or unclassified hemangioendothelioma? Am J Clin Pathol 1991;96:660-3.

5. Tosios KI, Gouveris I, Sklavounou A, Koutlas IG. Spindle cell hemangioma (hemangioendothelioma) of the head and neck: Case report of an unusual (or underdiagnosed) tumor. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008;105:216-21.
6. Coras B, Hohenleutner U, Landthaler M, Hohenleutner S. Spindle cell hemangioma. Dermatol Surg 2003;29:875-8.
7. Zeng TY, Fan J, Liu B, Wang Q. Spindle cell hemangioma in frontal bone: Report of a case. Zhonghua Bing Li Xue Za Zhi 2010;39:120-1.
8. Keel SB, Rosenberg AE. Hemorrhagic epithelioid and spindle cell hemangioma: A newly recognized, unique vascular tumor of bone. Cancer 1999;85:1966-72.
9. Maclean FM, Schatz J, McCarthy SW, Scolyer RA, Stalley P, Bonar SF. Epithelioid and spindle cell haemangioma of bone. Skeletal Radiol 2007;36 Suppl 1:S50-7.
10. Ono CM, Mitsunaga MM, Lockett LJ. Intragluteal spindle cell hemangioendothelioma. An unusual presentation of a recently described vascular neoplasm. Clin Orthop Relat Res 1992;281:224-8.

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