Primary Pleomorphic Liposarcoma of Bone: A Rare Tumor in Unusual Location with Review of Literature

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Summary

Primary liposarcoma of bone is an extremely rare tumor and accounts for only 0.03% of all primary bone tumors. Herein we report two rare cases of primary pleomorphic liposarcoma of long bone. Both cases presented with localised swelling and pain. Imaging study revealed that origin of tumor was primarily of bone. J needle biopsy in both cases showed highly pleomorphic cells with no malignant osteoid. Diagnosis of high-grade sarcoma was given. Histomorphology of post-chemotherapy resected specimens showed many pleomorphic lipoblasts with large areas of necrosis and no malignant osteoid. After ruling out the various differentials, final diagnosis of primary pleomorphic liposarcoma of bone were given in both cases. Hence, biopsy material alone is very challenging for the diagnosis. Careful histopathological examination with keeping in mind of a rare diagnosis along with a clinicoradiological correlation as well as immunohistochemistry play a useful role in confirmation of the diagnosis.

Keywords: Malignant bone tumor, Pleomorphic liposarcoma, Liposarcoma of bone, Rare primary bone tumor

Introduction

Primary liposarcoma of bone is an extremely rare tumor, with prevalence of only 0.03% among all bone tumors.[1] In 1955, Dawson reported the first convincing case of primary liposarcoma of bone. Subsequently over the years other authors also reported this rare entity. But in 1982, Addison and Payne reviewed all and found only 6 convincing cases.[2] Among all malignant adipocytic tumors, Pleomorphic Lipo Sarcoma (PLS) was least common.[1] We report two cases of primary PLS of bone with the aim to enrich the existing literature and a focus on potential pitfalls in diagnosis.

Case Details

Twenty-six and 16-year-old young males presented with pain and swelling in their left knee joint for 2 past months. Magnetic resonance imaging (MRI) revealed ill-defined tumor involving epi-meta-diaphysis of distal femur and proximal tibia, respectively. Both the cases showed altered marrow signal intensity which appeared hypointense on T1W and heterogeneously hyperintense on STIR with associated periosteal reaction, periosteal thickening and soft tissue component. Possibility of primary malignant bone tumor? Osteosarcoma was suggested in both the cases.

J needle biopsy of both the cases showed clusters of atypical cells having highly pleomorphic, hyperchromatic nuclei, irregular nuclear membrane and vacuolated cytoplasm. Malignant osteoid were not identified in both the cases (Figure 1a). Correlation of histopathology with clinicoradiological findings, diagnosis of high-grade sarcoma with possibilities of osteosarcoma with clear cell component, pleomorphic liposarcoma and metastatic carcinoma were given.

Both the cases received chemotherapy followed by surgery. Gross feature of case 1 showed, a 17x17x6 cm tumor involving the epi-meta-diaphysis of left distal femur. Cut surface was mainly necrotic (≥ 90%) and bony area was solid, greyish white to tan. (Figure 1b). Case 2 showed, a 17x8x6 cm tumor involving epi-metaphysis of left proximal tibia. Cut section of tumor was solid, greyish yellow to white, soft to firm, also areas of necrosis were evident. Both showed soft tissue extension and case 2 also showed overlying skin ulceration. Microscopic examination of both the cases showed many uni- and multi-vacuolated pleomorphic lipoblasts with extreme pleomorphism, brisk mitotic activity and areas of necrosis. Malignant osteoid were not identified. (Figure 1c, 2a, b).

Differential diagnosis include metastatic pleomorphic liposarcoma, primary pleomorphic liposarcoma, high grade osteosarcoma, dedifferentiated liposarcoma and undifferentiated pleomorphic sarcoma. Both patients were screened by positron emission tomography and computed tomography PET-CT scan which suggested primary bone tumor with no distant metastasis at time of diagnosis. Absence of malignant osteoid and no well differentiated liposarcomatous area ruled out osteosarcoma and dedifferentiated liposarcoma respectively. Presence of typical pleomorphic lipoblasts ruled out the undifferentiated pleomorphic sarcoma.

IHC was done to rule out the various differentials. Tumour cells were immunoreactive for vimentin, focally for S100 protein and negative for SATB2. (Figure 1d, e, f, Figure 2c, d).

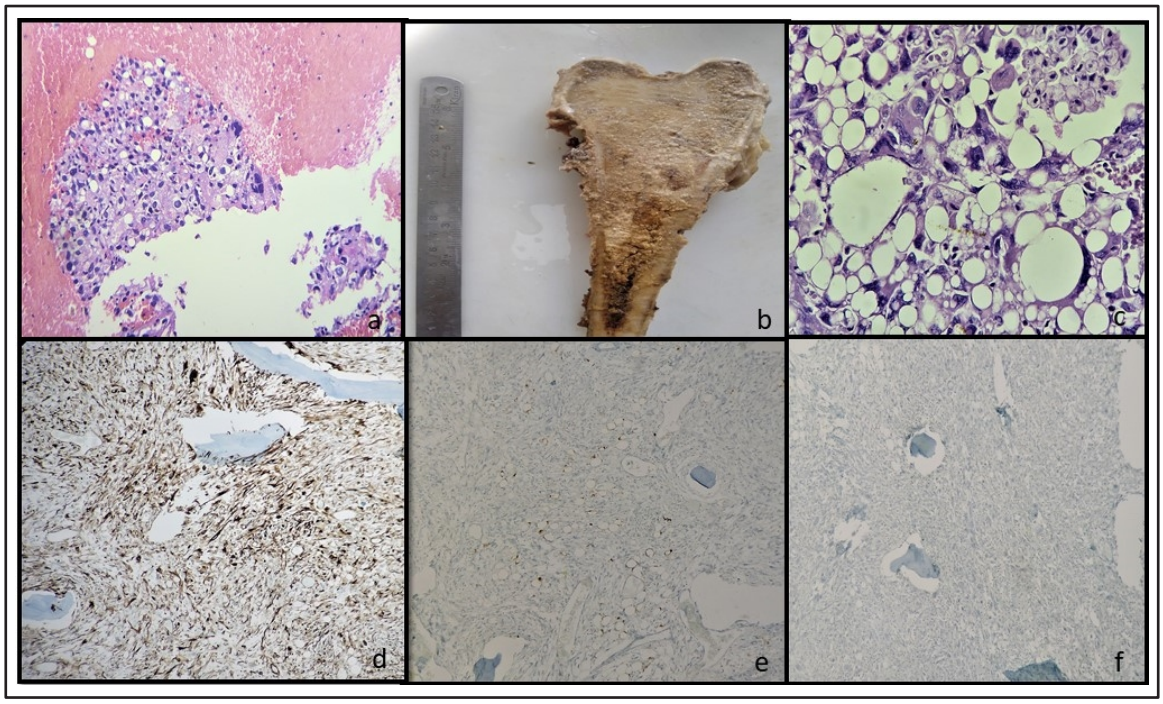

Figure 1: Case 1/ (a) H and E staining from J needle biopsy showed many atypical cells with hyperchromatic nuclei and vacuolated cytoplasm 20x (b) Gross photograph of post chemotherapy specimen (c) Many pleomorphic lipoblasts in resection specimen 40x (d to f) Immunohistochemical stains 20x (d) vimentin(cytoplasmic) positive (e) S100 protein (nuclear and cytoplasmic) focal positive (f) Negative SATB2.

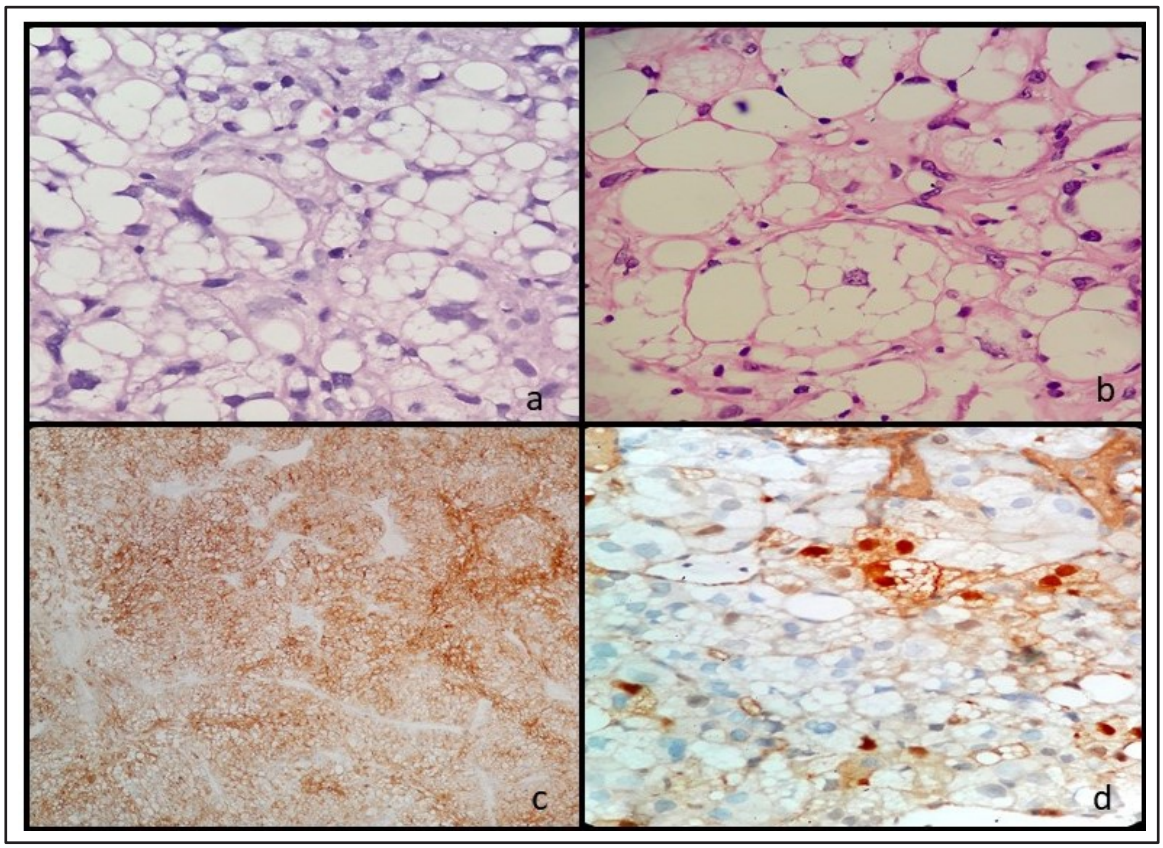

Figure 2: Case 2(a, b) Histomorphology of resected specimen shows many pleomorphic lipoblasts 40x (c,d) Immunohistochemical stain (c) vimentin (cytoplasmic) positive (d) S100 protein (nuclear and cytoplasm) focal positive.

Table 1: Previously reported cases of pleomorphic liposarcoma of bone

Study	Age/sex	Site	Metastasis	Treatment received	Follow up
Torok et al[2]	34/M	Femur	Lung	Wide resection, radiation, chemotherapy	Died, 16 months
Hamlat et al[3]	45/F	Thoracic spine	Lung and rib	Laminectomy T7-T8 and radiotherapy	Alive,19 months
Torigoe et al[4]	38/F	Humerus	Liver	Wide resection, Initial high-dose ifosfamide followed by cisplatin and doxorubicin	Died, 8 months
Lmejjati et al[5]	35/M	Lumber spine	Locally invasive	Emergency decompression at L4/L5 and radiotherapy	Died, 3 months
Rasalkar[6]	13/M	Femur	Lung	Neoadjuvant chemotherapy (methotrexate, cisplatin), Surgery, Adjuvant chemotherapy (ifosfamide/etoposide and adriamycin/cisplatin)	Alive, 13 months
Tiemeier GL et al[7]	18/M	Tibia	Femur and lung	Chemotherapy (methotrexate, doxorubicin and cisplatin), wide resection	Alive,12 months
Fajolu O et al[8]	19/M	Femur	Lung, liver,vertebra, scapula	Chemotherapy (etoposide,ifosfamide) and wide resection	Died, 8 months
Our study	16/M	Tibia	Contralateral Femur	Chemotherapy and wide resection	Lost follow up after 7 months
	26/M	Femur	-	Chemotherapy (Doxorubicin, cisplatin), wide resection	Died, 6 months

After excluding all differentials final diagnosis of primary pleomorphic liposarcoma of bone were given in both the cases. Case 2 also showed metastasis to the contralateral side of distal femur after 2 months of initial diagnosis.

Both cases were followed up. Case 1 died after 6 months of initial symptoms, while we lost follow up of case 2 after 7 months of initial symptoms.

Discussion

Malignant adipocytic tumor accounts for 10-35% of all soft-tissue sarcomas,[1] and PLS is the least common subtype, with prevalence of only 5- 15% of all liposarcomatous lesions. Liposarcoma arising from the bone is a rare entity and very few confirmed cases of primary PLS of bone have been reported in literature.[1] Total of 1008 primary malignant bone tumors were reported between 2018 to 2020 in our hospital. Most common primary malignant bone tumor was osteosarcoma (57.83%) followed by Ewing sarcoma (26.58%) and chondrosarcoma (15.37%), and only two cases of primary liposarcoma of bone were reported. Till date only 7 cases of primary PLS of bone have been reported in literature (Table 1).[2-8] Diagnosis of primary liposarcoma of bone can be considered only if it arises from the bone and have characteristic gross and histological features.[1]

Primary PLS of bone affect a wide age range from 15-53 years and equally affects both genders.[1] Long bones of lower extremities and upper extremity most frequently affected while rarely it can affect scapula, maxilla and mandible.[1]

Radiology of primary liposarcoma of bone shows non-specific features such as large osteolytic lesion with cortical destruction. PLS contain less adipocytic component as compared to other subtypes and this makes imaging diagnosis difficult.[1] MRI study shows intermediate signal intensity with T1 weighting and intermediate to high signal intensity with T2W.[1] These findings were concordant with our cases.

Histomorphology of PLS includes a lipogenic area with variable number of pleomorphic lipoblasts and non-lipogenic area with high grade sarcoma features. Both our cases had similar histomorphology with lipogenic areas showing many pleomorphic lipoblasts and non-lipogenic areas having high grade sarcomatous features.

Immunohistochemistry and molecular analysis have very limited diagnostic role. Murine Double Minute 2 (MDM2) amplification is negative in PLS while osteosarcoma, low grade and parosteal and those that dedifferentiate into high grade osteosarcoma show positive staining.[9] Gebhard et al,[10] observed that S-100 protein immunoreactivity was seen in up to 48% of lipogenic areas which were concordant with our cases.

Primary osseous liposarcoma is an aggressive malignancy with mean survival rate of two years only.[1] Wide surgical resection is the mainstay of

treatment of primary osseous liposarcoma, with radiation therapy used for palliation or following excision to prevent local recurrence.[1] Both our cases received chemotherapy prior to surgery. One patient succumbed to death post operatively while another showed osseous metastasis in contralateral long bone. However, we lost follow up of the patient after this.

Conclusion

Primary pleomorphic liposarcoma of bone is an extremely rare tumor. Diagnosis alone on biopsy material is very challenging. Due to rarity of tumor, despite of the presence of lipoblasts, one may still miss the diagnosis if not aware of the entity.

Conflict of interest

No conflict of interest.

Acknowledgements

None

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