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Low-Grade Central Osteosarcoma Arising from Bone Infarct: A Case Report

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Abstract:-Bone infarct is a aseptic or avascular necrosis of bone. It is relatively common orthopaedic finding and is often idiopathic but can be seen secondary to an underlying condition such as alcoholism, corticosteroid use, or Caisson's disease. It can occur anywhere in the skeleton but are predominately seen around the hip and knee. Secondary sarcoma arising in association with a preexisting bone infarct is extremely rare, accounting for less than one percent of all bone sarcomas. It's histology usually reflects a high-grade sarcoma, such as malignant fibrous histiocytoma of bone, fibrosarcoma or conventional osteosarcoma. Low-grade sarcoma arising from bone infarct has not been described well in the literature.We present a 65year old female patient with bone infarct in her right distal femur and proximal tibia, from which a low-grade central osteosarcoma developed. A histologic diagnosis of low-grade central osteosarcoma was confirmed by immunohistochemical expression of MDM2 and CDK4. She had also pathological fracture in right distal femur.

Keywords:-Bone Infarct, Malignant Fibrous Histiocytoma, Fibrosarcoma, Osteosarcoma, MDM2, CDK4

I. INTRODUCTION

Bone infarct is a aseptic or avascular necrosis of bone in metaphysial and diaphysial segment. It is relatively common orthopaedic finding and in most instant cause is unknown [1] but can be seen secondary to an underlying condition such as alcoholism, corticosteroid use, or Caisson's disease. It can occur anywhere in the skeleton but are predominately seen around the hip and knee. Although bone infarcts can initially be painful, most are discovered incidentally on imaging studies performed for the evaluation of other abnormalities. Secondary sarcoma arising in association with a preexisting bone infarct is extremely rare [2,3] , accounting for less than one percent of all bone sarcomas. It occurs in sixth decades of life and 2/3 rd patients are male [4]. Its histology usually reflects a high-grade sarcoma, such as malignant fibrous histiocytoma of bone in 60% cases[4], fibrosarcoma or conventional osteosarcoma. Low-grade sarcoma arising from bone infarct has not been described well in the literature.

Only few Case studies and a few compilations of the reported data have been published [4], so we present clinical and pathologic findings of our case in detail for this extremely rare condition.

II. CASE REPORT

A 65year old female patient , working as housewife, admitted to New medical teaching hospital, Kota , Rajasthan because of pain and swelling in right lower thigh and pain in proximal leg with restriction of knee flexon for last three month on October 2017. Digital skiagram was advised, it's lytic lesion of right distal femur and proximal tibia with sclerosis proximal to lytic lesion probability of bone island or bone infarct. There was pathological fracture of right distal third femur. On thorough clinical evaluation, no evidence of any other osseous or non osseous tumor.

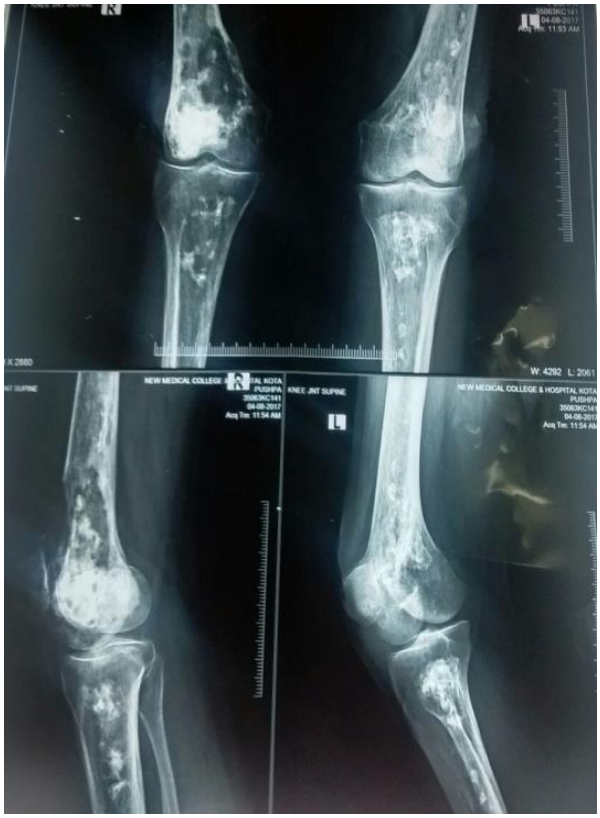

Fig. 1:Anteroposterior and Lateral Radiograph of Knee Joint Shows Multifocal Bone Infarct in Proximal Tibia and Distal Femur with Well Defined, Dense Sclerotic Margins and Adjacent Osteolysis in Distal Femur.

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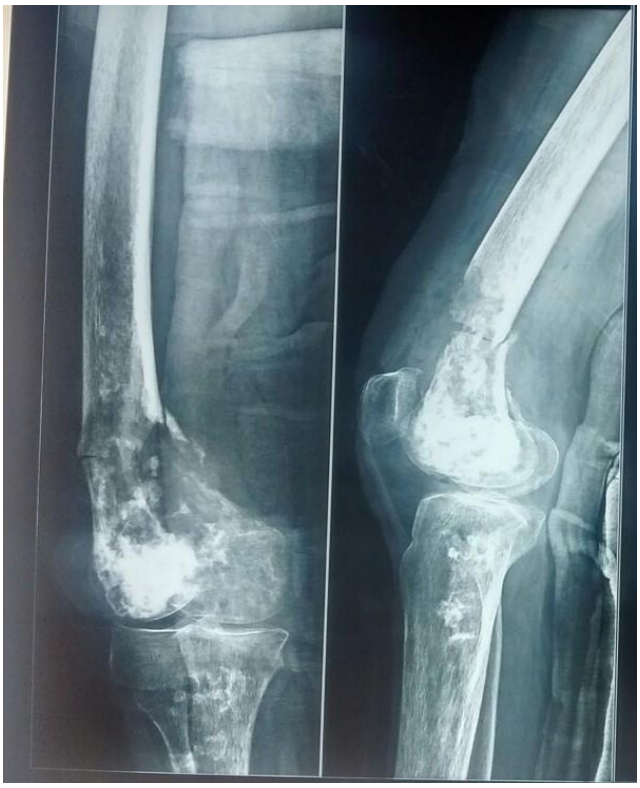

Fig. 2 Anteroposterior and Lateral Radiograph of Knee Joint Shows Pathological Fracture of Distal Femur

On physical examination 10x12cm soft tissue mass with varrigated consistency was present in the right lower thigh which was associated with pain, margin were irregular, overlying skin was shiny with venous prominence. No any scar, sinus or discharge present. Temperature slightly raised as compare to normal limb. There was 1cm wasting of thigh and 0.5cm wasting of calf muscle. No regional lymphadenopathy found. knee flexon was limited to 90^{o} with normal ankle movement.

- Chest examination and chest x ray found normal.
- Abdominal examination reveals soft abdomin with no other abnormality.
- Cardiovascular system examination reveals systolic murmur. ECG s/o left axis deviation as patient is on treatment of hypertension.
- Routine laboratory investigation s/o mild aneamia. s.ca^{+2} , phosphate, alkaline phosphatase level are within normal limit.
- NCCT bilateral knee joint was done.

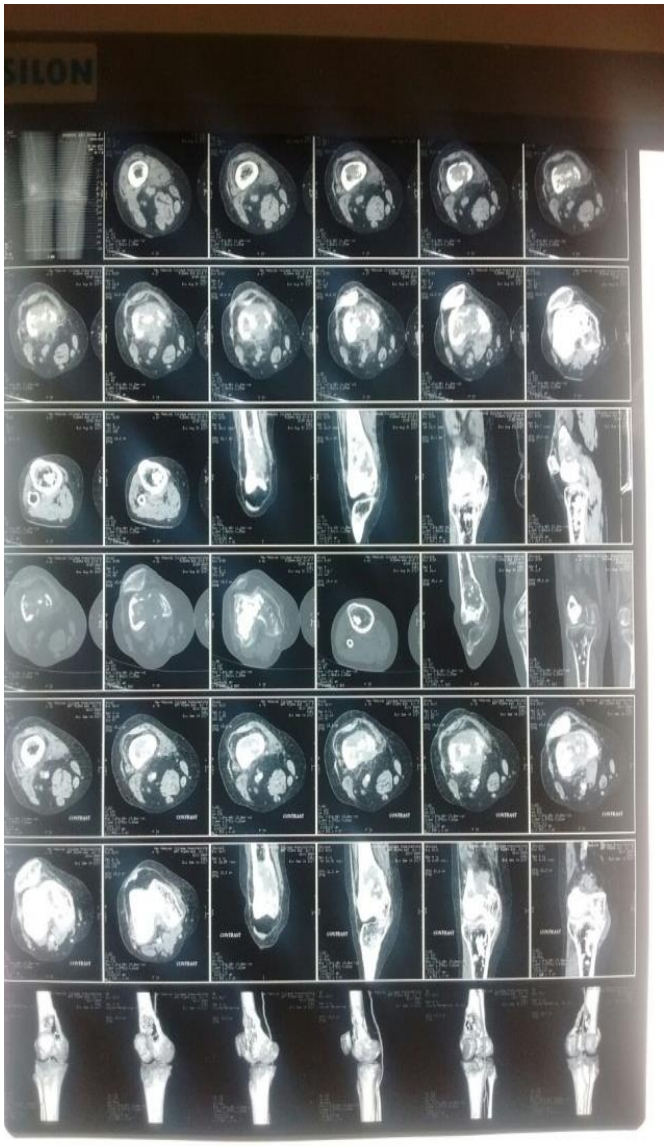

Fig. 3 NCCT b/l knee joint

Interpretation ; multiple irregular shape area of central hypodensity and peripheral serpiginous sclerosis is seen in left distal femoral and bilateral proximal tibial metaphyseal region s/o osteonecrosis [bone infarct]. Right distal femur showing significant sclerosis extending from metaphysis to epipyseal region with soft tissue attenuation mass lesion arising from central hypodensity region and causing cortical destruction of anterior and posterior surface of distal femur , however no significant enhacement is seen s/o ; Infarct Associated Sarcoma. [Left side bone infarct of distal femur and proximal tibia was incidental findinding]

To confirm the diagnosis we did incisional biopsy of bone and soft tissue and sent for histopathological examination.

Biopsy Findings were ; ill marginated , pleomorphic spindle cell tumor. Cells are large , oval to spindle shows moderate pleomoephism. Mitosis are 4-6/10HPF. Tumor cells arranged in storiform pattern. Dens areas of collagen also seen. Osteoid formation is not seen. Its s/o low grade

osteosarcoma. The diagnosis was confirmed by immunohistochemical expression of MDM2 and CDK4.

III. DISCUSSION

Infarct associated sarcoma is extremely rare accounting for less than 1%. They appear to arise in long-standing mature infarcts, particularly those that occur in the lower extremity around the knee. Many patients who develop infarct-associated sarcoma do not have a defined risk factor for the development of osteonecrosis, but a large proportion of patients have multifocal bone infarction. Pain is the most common presenting symptom and should alert the radiologist and clinician to carefully examine the bone surrounding the infarct for signs of malignancy; these signs can be extremely subtle and initially overlooked amid the distracting appearance of the sclerotic infarction.

In 1960 Furey *et al* [5] reported the first case of fibrosarcoma,. Since then, several other types of infarct-associated sarcomas have been reported, including malignant fibrous histiocytoma [6], osteosarcoma [7], angiosarcoma [8], and epithelioid hemangioendothelioma [9].

In 1992, Torres and Kyriakos [4] reported a case of osteosarcoma arising in a medullary infarct of the humerus

In 2009 Domson *et al*[10] described their experience with the treatment of 15 patients with infarct-associated sarcoma and also reviewed additional cases that had been documented after the article by Torres and Kyriakos was published.

In 2015 Gregory Scott Stacy,Ryan Lo ,Anthony Montag published results of Infarct-Associated Bone Sarcomas: Multimodality Imaging Findings. The imaging studies of 258 patients with sarcomas were reviewed to determine whether underlying osteonecrosis was present. Nine infarct-associated bone sarcomas were found in eight patients: seven malignant fibrous histiocytomas (MFHs) and two osteosarcomas. All occurred in the femur or tibia; multifocal infarction was documented in all patients except one. All lesions, including those treated at their institution and those found in the literature, were metaphyseal or diaphyseal, and although epiphyseal extension of sarcoma from a metadiaphyseal infarct was common, no purely epiphyseal lesions were encountered.

IV. CONCLUSION

Sarcomata's degeneration of a bone infarct is extremely rare and has been reported only sporadically, often as isolated case reports. It is challenging clinical entity for treating orthopaedician and patients.

The orthopaedic an and radiologist must be vigilant for this rare occurrence and should be suspicious when poorly defined areas of osteolysis (or mineralization) with cortical thinning and bone destruction arise next to a bone infarct. When infarct-associated sarcoma is suspected, biopsy and immunohistochemical study is recommended to confirm the development of a sarcoma and MRI for extent of bone and soft-tissue invasion. Infarct-associated bone sarcomas carry a poor prognosis; therefore, it is important to identify these malignancies early and to determine whether metastasis is present to properly guide therapy.

BIBLIOGRAPHY

[1]. Bullough PG, Kambolis CP, Marcove RC, Jaffe HL. Bone infarctions not associated with caisson disease. J Bone Joint **Surg** [Am] 1965; 47:477-91.
[2]. Healey JH, Buss D. Radiation and pagetic osteogenic sarcomas. Clin Orthop Relat Res. 1991;270:128–134.
[3]. Shaheen M, Deheshi BM, Riad S, Werier J, Holt GE, Ferguson PC, Wunder JS. Prognosis of radiation-induced bone sarcoma is similar to primary osteosarcoma. Clin Orthop Relat Res. 2006;450: 76–81.
[4]. Torres FX, Kyriakos M. Bone infarct-associated osteosarcoma. Cancer. 1992;70:2418–2430.
[5]. Furey JG, Ferrer-Torrer-Torells M, Reagan JW. Fibrosarcoma arising at the site of bone infarcts: a report of 2 cases. J Bone Joint Surg Am. 1960;42:802–810.
[6]. Michael RH, Dorfman HD. Malignant fibrous histiocytoma associated with bone infarcts: report of a case. Clin Orthop Relat Res. 1976;118:180–183.
[7]. Resnik CS, Aisner SC, Young JW, Levine A. Case Report 767: osteosarcoma arising in bone infarction. Skeletal Radiol. 1993;22:58–61.
[8]. Abdelwahab IF, Kenan S, Klein MJ, Lewis MM. Case report: angiosarcoma occurring in a bone infarct. Clin Radiol. 1992;45:412–414.
[9]. Pins MR, Mankin HJ, Xavier RJ, Rosenthal DI, Dickersin GR, Rosenberg AE. Malignant epithelioid hemangioendothelioma of the tibia associated with a bone infarct in a patient who had Gaucher disease: a case report. J Bone Joint Surg Am. 1995;77: 777–781.
[10]. Domson GF, Shahlaee A, Reith JD, Bush CH, Gibbs CP. Infarct-associated bone sarcomas. Clin Orthop Relat Res 2009; 467:1820–1825.