

MRI Manifestation and Early Diagnosis of Bone Infarct: A Rare Complication of Steroid Therapy for Pemphigus

UC Parashari, MD, S Khanduri, MD, S Bhadury, MD, P Sarkar, DMRD (STD)

Department of Radio Diagnosis, Era's Lucknow Medical College, Lucknow, India

ABSTRACT

The objective of the study was to analyse the magnetic resonance imaging (MRI) manifestation of bone infarction and explore the value of such imaging in early diagnosis. In bone infarction, aseptic osteonecrosis occurs in the metaphysis and diaphysis of long bones. The "Double line" sign in MRI is most characteristic of this lesion and is seen as high signal intensity in the inner zone along with low-signal-intensity line in the outer zone in a T2 weighted image. In clinically suspected patients, MRI can be used for early diagnosis. Here we describe a case of pemphigus resulting from steroid therapy leading to development of bone infarction in the metadiaphyseal region of the femur and tibia.

Key Words:

MRI, Double Line Sign, Bone Infarct, Aseptic Osteonecrosis, Steroid Therapy, Pemphigus

INTRODUCTION

Bone infarct or osteonecrosis refers to ischaemic death of the cellular elements of the bone and marrow in the metadiaphyseal region. Osteonecrosis can be idiopathic or secondary to a number of conditions. Clinical manifestations of osteonecrosis are not typical especially in early infarction; even plain radiography examination is usually negative in early stages¹. MRI and bone scans therefore play a significant role in early diagnosis thus reducing the severity and number of complications and morbidity associated with the disease.

CASE STUDY

A 58-year-old male with a known case of pemphigus diagnosed three years previous to presentation was treated with 40-60 mg per day oral prednisone for the last year. He complained of recent onset of left knee joint pain that worsened progressively in the few days prior to presentation. There was no history of recent infection or trauma. Examination of the affected knee revealed mild swelling with tenderness while general examination was unremarkable. Plain radiograph revealed an ill defined sheet

like lucency with sclerosis in the metadiaphyseal region of the left lower femur and upper tibia (Figure 1). These findings were nonspecific, however with the strong suspicion of bone infarct, MRI was performed.

MRI showed serpiginous lesions in the metadiaphyseal region of the lower femur and upper tibia (Figure 2) in T2 weighted images. The "double line sign" was seen as high signal intensity in the inner zone and low-signal-intensity line in the outer zone on a T2 weighted image, representing the borders of osseous resorption and healing. The lesions themselves were seen as geographic low signal areas surrounded by a serpiginous high signal rim in the lower femur and upper tibia in short T1 inversion recovery images (Figure 3). In T1W images the lesions were seen as geographical areas with hypointense periphery and central area iso intense with marrow (Figure 4).

DISCUSSION

Bone infarction refers to ischaemic death (aseptic necrosis) of the cellular elements of the bone and marrow. These infarctions occur mainly in the metadiaphyseal region of long bones; 77% of cases involve lower limbs. Often these



Fig. 1: Plain AP radiograph of left knee revealing an ill defined sheet like lucency with sclerosis in the metadiaphyseal region of left lower femur and upper tibia.

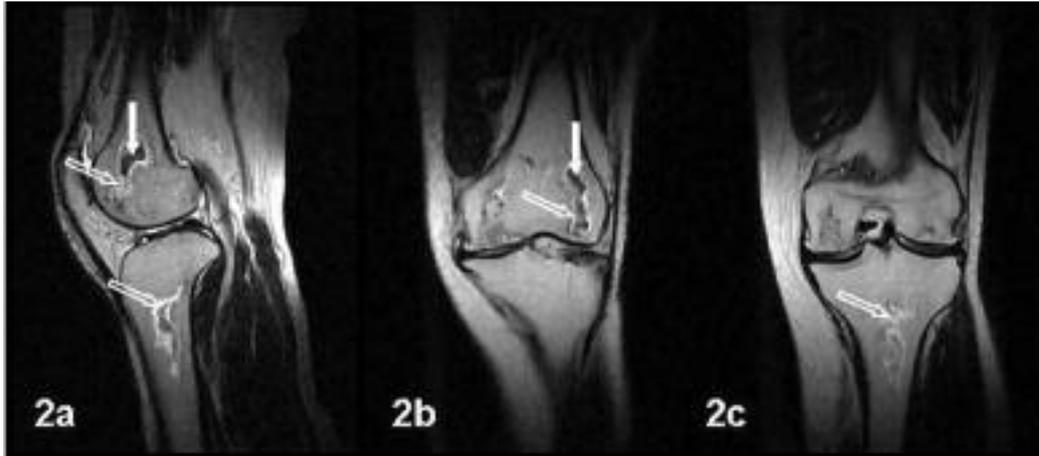


Fig. 2: Sagittal (2a) and coronal (2b & 2c) T2W MR Images of left knee showing serpiginous lesions in metadiaphyseal region of lower femur and upper tibia displaying hyper intense outer ring and central hypo intense area. The "double line sign" is seen as curvilinear adjacent low and high signal intensity present at the edge of infarct.

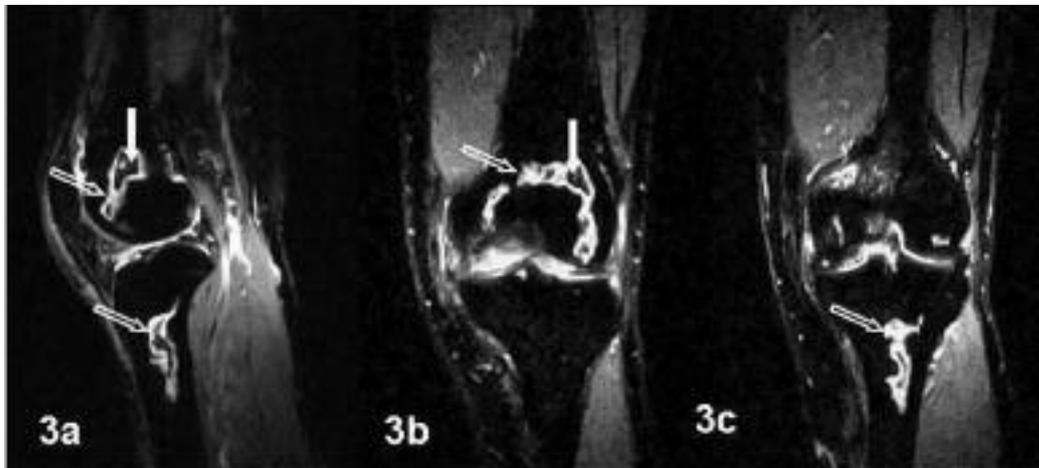


Fig. 3: Sagittal (3a) and coronal (3b & 3c) STIR MR images of left knee showing geographic low signal areas surrounded by a serpiginous high signal rim in the lower femur and upper tibia.



Fig. 4: Sagittal T1W MR shows geographical lesions with peripheral hypo intensity with central area iso intense with marrow.

lesions occur in multiples and most frequently are symmetrical; most cases occur in male patients around middle age. Most prevalent are presentation in the lower region of the femur, tibia, and proximal humerus.

Bone infarctions are usually asymptomatic or present with mild symptoms. Underlying aetiology may include use of steroids, alcoholism, dyslipidaemia, cytosteatonecrosis, lupus erythematosus, renal transplant or presentation may be idiopathic. Rarely symptom onset mimics a clinical picture of acute inflammation especially in Gaucher's disease, cytosteatonecrosis, drepanocytosis (sickle cell anaemia), trauma or malignancy^{2,3}. Pathogenesis of bone infarction is complex. The mechanism of bone infarction is thought to be localized bone pathology caused by reduced blood circulation. Long limb bone marrow is rich in fatty tissue, with small marrow nutrient vessels that are easily embolised leading to bone infarcts. The basic pathological change in osteonecrosis is divided into stages of cell necrosis and phases of bone repair, the bases of which are seen in plain radiography, CT and MR imaging.

Although corticosteroids have been commonly utilised in treatment of pemphigus, bone infarctions around knee joint are rarely reported as sequelae. Osteonecrosis of the femoral head usually occurs in patients under corticosteroid therapy. The patient described here is an exceptional case of bone infarction occurring around the knee joint in pemphigus secondary to high dose steroid administration.

Clinical manifestations of bone infarction tend to be nonspecific. Plain radiographs are usually negative, and can lead to missed and misdiagnosis thereby delaying treatment¹. Osteonecrosis is seen as an arc-like, subchondral, translucent lesion and may be associated with patchy areas of decreased bone density mixed with sclerotic areas and bone collapse. CT imaging results in improved visualisation of bone infarction; it is not the modality of choice for diagnosing early bone infarction. CT is however the optimal modality for follow-up of bone infarction. Bone scans, although quite sensitive are not highly specific with the lesion appearing as a 'donut' sign.

MRI is the gold standard for early diagnosis of bone infarction; accordingly, it can more accurately define the bone infarction process reflecting its various stages in imaging results. Du Yu Qing *et al*⁴, divided bone infarction in three stages: acute, sub-acute and chronic; alternatively, Mitchel⁵ divided infarct in four types: fat signal type, blood signal type, water signal type, and fibre signal type. Irregular shape map plate-like lesions are seen with clear margins in typical MRI findings of bone infarction. Joint effusion may occur in bone infarction due to decreased venous return.

Radiological differentiation of bone infarction includes distinction from tumours and osteomyelitis, especially in

unique cases such as the present patient. Both acute osteomyelitis and infarction present similarly. In acute osteomyelitis, soft tissue swelling with marrow oedema is present; however in bone infarction typical map plate-like appearance is seen. In contrast-enhanced MRI, thick irregular peripheral enhancement is seen in the bone centre for cases of acute osteomyelitis; however in bone infarction, edge enhancement is seen. Chronic osteomyelitis is yet another differential diagnosis - on plain radiography and CT, chronic osteomyelitis appears as sinus, fistula, and sequestrum none of which are seen in bone infarction. Malignant bone tumours can also mimic bone infarction. Though in early stages, it is difficult to differentiate the presence of soft tissue mass and destruction helps in the diagnosis of bone infarction. Simple bone oedema should also be ruled out with follow up.

There is no specific treatment for bone infarcts; clinicians should continue to manage the primary pain generator and predisposing conditions, yet remain astute to the possibility of developing bone pain from malignant change within the bone infarct. Bisphosphonates have also been used in some cases. Disease progression can be slowed by transplanting nucleated cells from bone marrow after core decompression, although further research is needed to establish this technique as a standard treatment. The prognosis of bone infarction is often excellent, although in rare cases malignant transformation may occur. Secondary infection is a less common complication. Radiological follow up seems to be necessary however there is no consensus for this. We conclude that when clinical signs and symptoms are consistent with bone infarction, MRI examination should be carried out as soon as possible to facilitate early diagnosis and treatment.

REFERENCES

1. Chengtao W, Jiang XG, Tang XF, et al. Long tubular bone infarction imaging analysis. *Journal of Medical Imaging*, 2005; 15(6): 497-499
2. Mehsen N, Barnetche T, Redonnet-Vernhet I, Guérin V, Bentaberry F, Gonnet-Gracia C, et al. Coagulopathies frequency in aseptic osteonecrosis patients. *Joint Bone Spine* 2009; 76(2): 166-9.
3. Assouline-Dayana Y, Chang C, Greenspan A, Shoenfeld Y, Gershwin ME. Pathogenesis and natural history of osteonecrosis. *Semin Arthritis Rheum* 2002; 32(2): 94-124.
4. DU Yu-qing, pore-like spring, LAU Yuk Hing. Bone infarction imaging manifestations and pathologic basis. *Clin Radiol* 2004; 23(2): 143-6.
5. Mitchell DG. Using MR imaging to probe the pathophysiology of osteonecrosis. *Radiology* 1989; 171(1): 15-26.