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Double line sign in systemic lupus erythematosus

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Abstract
Double line sign is a pathognomonic sign seen in T2W images of MRI of bone infarct at border between the viable and non-viable bone. In these images, two lines are seen at the border of infarct-the inner bright line due to granulation tissue and the outer darker line due to sclerosis. Osteonecrosis is reported in 3-30% of SLE. Osteonecrosis in SLE is possibly steroid induced. This has to be differentiated from 'rim sign' which is seen in avascular necrosis due to osteochondral fragment with fluid surrounding it.

Keywords: double-line sign, SLE, avascular necrosis

Introduction

Double Line Sign in Systemic Lupus Erythematosus

Double line sign is a pathognomonic sign seen in T2W images of MRI of bone infarct. In these images, two lines are seen at the border of infarct-the inner bright line due to granulation tissue and the outer darker line due to sclerosis. Bone infarct / osteonecrosis is cell death of bone and marrow due to ischaemia. Avascular necrosis (AVN) is infarct in the epiphysis, whereas osteonecrosis involves meta/diaphysis. Osteonecrosis / bone infarct may be idiopathic / due to reduced blood supply to bone Focal marrow ischaemia may be secondary to intrinsic or extrinsic compression of blood vessels or combination of both. In red marrow cell death occur 12hour following anoxia and in yellow marrow it occurs within 5 days. When the blood supply is interrupted there is central area of necrosis. This necrotic core is surrounded by hyperaemic ischaemic zone**.** Gradually collagen granulation tissue layered around necrotic core. The variable imaging appearance depends on the normal marrow, ischaemic zone and necrotic core. Sludging of cells in vessels causes bone infarct in Sickle cell disease, Haemoglobinopathy, polycythaemia vera, Lymphoproliferative disease. Fat cells causing medullary hypertension and intra osseous venous occlusion and bone infarct in steroid use. SLE, RA cause vasculitis leading to arterial occlusion. Plain radiograph is initial imaging modality to detect the lesion and MRI is diagnostic [Figures-1, 2]. There is significant delay in onset of infarcts and appearance on radiographs. Radionuclide scan is more sensitive.

Osteonecrosis (ON) in SLE is first reported in 1960 [1] and since then the incidence is frequently reported and is 3-30% [2]. Osteonecrosis in SLE is possibly steroid induced. 27-42% of SLE patients have APLA-AB syndrome which is characterised by arterial and veno-occlusive disease, thrombocytopenia and vascular thromboses and miscarriages [3]. Atherosclerosis in SLE may be accelerated by corticosteroid induced dyslipoproteinemia, secondary to renal hypertension, vasculitis. Vasculitis involves small vessels and involve any organ resulting in ischaemia.

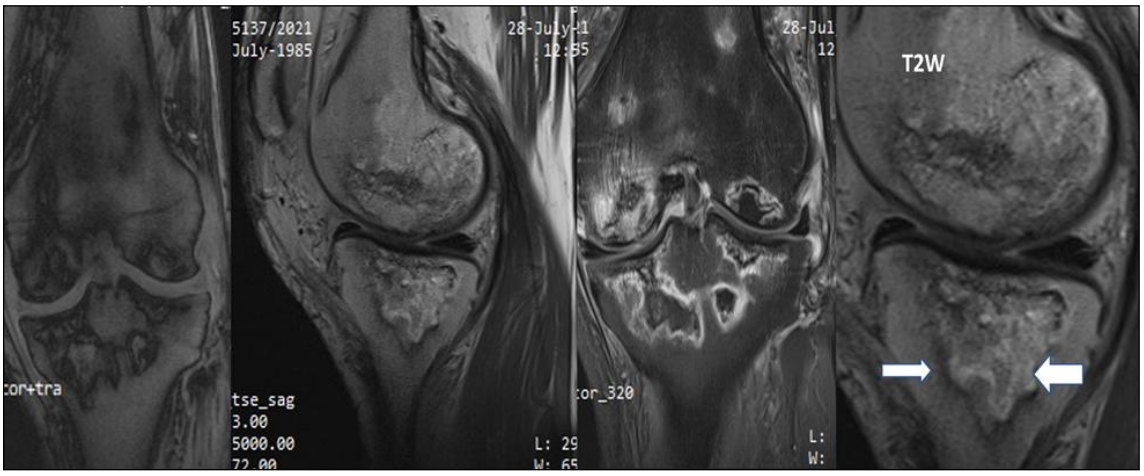

Fig 1

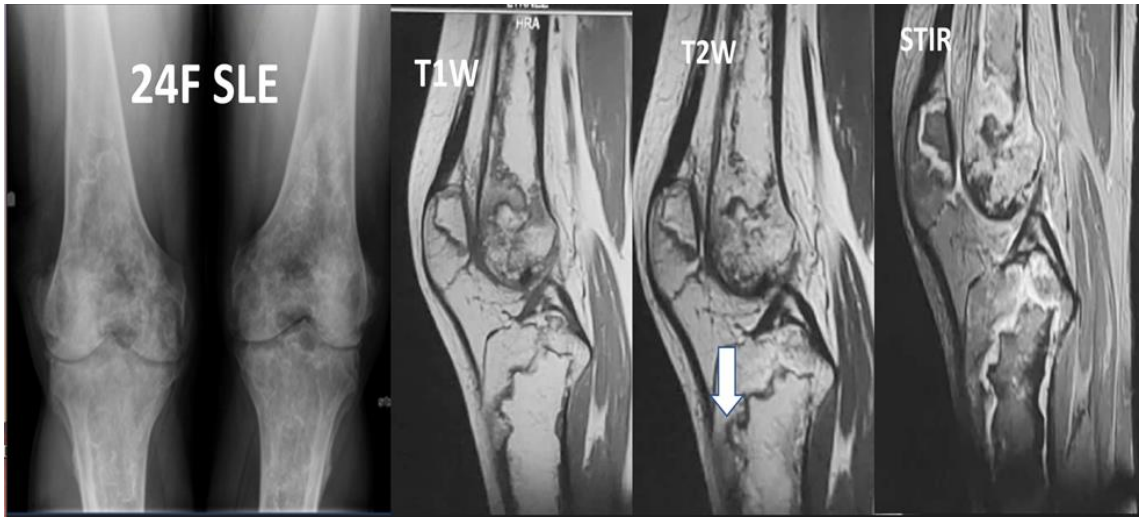

Fig 2

Fig 1 and 2: Two different cases demonstrate the double line sign in T2W image

Note: central area is normal bone having the signal characteristic of normal marrow that is hyper on T1 and T2W images, supressed on STIR

Radiograph in figure 2 shows central lucency bordered by serpigenous sclerosis

Prevalence of asymptomatic AVN is 29-45%; but the incidence has reduced over past 2 decades due to modification of treatment modality and reduction in use of corticosteroids [4]. The extent of fat conversion paralleled with prednisolone intake in patient with osteonecrosis in SLE. The increase in marrow fat content elevates the marrow pressure reducing arterial perfusion leading to osteo-necrosis [5]. Often lesions are multiple, bilateral and symmetrical occurring in medullary cavity and in metaphyseal region.

In early-stage radiograph show nonspecific findings and there may be areas of mottled lucencies and sometimes with mild sclerosis. An area of preserved bone marrow signal surrounded by a serpiginous line. This central lucency surrounded by serpiginous sclerotic border giving smoke up the chimney lesion. Intramedullary calcification seen within the infarct in conventional radiograph after months or years. At this stage the lesions are to be differentiated from Enchondroma. Central area of marrow signal surrounded by T1 hypointense rim representing sclerosis/granulation tissue.

Double line sign pathognomonic of bone infarct seen at border between the viable and non-viable bone. Inner bright line due to granulation tissue and outer dark line due to sclerosis. The outer line is continuous and inner line may be continuous / discontinuous This sign is reported in 80-85% cases. Osteonecrosis can be differentiated from 'rim sign' which is seen due to osteochondral fragment with fluid surrounding it. This is noted in AVN. Other differential is osteomyelitis and is differentiated by obtaining contrast enhanced scan. In osteonecrosis there is peripheral thin linear/rim- enhancement whereas in OM there is more geographic /irregular thick peripheral enhancement at centre. Defect in the cortex may be appreciated in OM with edema in marrow and adjacent soft tissue

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