Avascular Necrosis of the Spine in a Patient with Systemic Lupus Erythematosus and Antiphospholipid Syndrome

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Abstract

Avascular Necrosis (AVN) lesions affecting the long bones, especially the femoral and humeral heads, are observed with increased frequency in patients with lupus and antiphospholipid syndrome. However, AVN involving the spine is exceptionally unusual. We report a case that illustrates the importance of considering the diagnosis of AVN when evaluating lesions involving the spine in those patients and the characteristic findings.

Introduction

Systemic Lupus Erythematosus (SLE) and Antiphospholipid Syndrome (APS) are known risk factors for Avascular necrosis (AVN). AVN commonly occurs in femoral and humeral heads. The imaging findings can mimic what may be in other processes such as infections, malignancy or other marrow replacing processes and may lead to extensive work up. Although rare, AVN can occur in the spine and needs to be considered in the differential diagnosis.

Case Report

3 year old caucasian female with an established history of Systemic Lupus Erythematosus (SLE), Antiphospholipid antibody syndrome (APS), deep vein thrombosis on chronic anticoagulation was evaluated for pleuritic type chest pain. A CT pulmonary angiogram was performed (Figure 1). The exam revealed no pulmonary embolus, but did describe two lytic lesions at T9 and T10 which were new since the comparison study from nearly 4 years earlier. Both had an irregular shape with sclerotic margins. The radiologist differential diagnosis included infection and metastatic disease, with the latter considered less likely given the patient’s young age.

MRI was later obtained which demonstrated heterogeneous low T1 and high T2 signal within T9, while T10 showed predominantly high T1 signal with T2 signal being high peripherally and low centrally (Figures 2a, 2b and 3). Similar findings were also seen in the lumbar spine, particularly at L5 which had appeared abnormal on a lumbar MRI from 4 years prior. Differential diagnoses included a marrow infiltrative process, myelofibrosis, or malignant infiltration, and bone scan was recommended.

On Tc 99m MDP bone scan, photopenia was seen in the T9 and T10 vertebral bodies corresponding with the lesions (Figure 4). Heterogeneous uptake in the left proximal femur was suggested to possibly represent an early lytic lesion, and radiographic correlation was recommended.

The patient’s pleuritic chest pain resolved and she was asymptomatic in terms of these lesions. She did not report any history of trauma to the spine. She was not on prednisone for at least 6 months prior to her presentation. Based on the imaging studies and patient’s clinical history, AVN of the spine secondary to APS / SLE is considered the likely diagnosis. She is being monitored clinically and radiologically, she has not required any escalation of her therapy and is stable at 2 years with minimal progression of her lesions.

Discussion

Avascular necrosis is reported in association with many entities including excessive alcohol use, glucocorticoid use, SLE and APS [1-6]. AVN has been reported in 3 to 30 percent of patients with SLE [4,5]. 20 percent of patients were found to have asymptomatic AVN in patients with primary APS based on a study by Tektonidou et al. [3]. AVN of the spine is an uncommon disease in general [7] and in the setting of SLE and APS. Only one case in this setting was reported based on a search of the English literature [8]. The case presented above reflects some of the characteristic radiological findings that are seen with AVN using various imaging modalities and the varied manifestations of AVN based on location and phase. For example, the characteristic sclerotic line surrounding a bone infarct occurs late in the disease course [9,10]. While the appearance is often very characteristic, differential diagnoses

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may include an involuting fibroxanthoma or enchondroma. When occurring in a subchondral location, subchondral cystic changes of osteoarthritis and osteochondral defects may be considered [11]. Earlier in the evolution, the sclerotic rim is absent with the lesion appearing either normal or as a lucency, typically in the subchondral bone [12]. When the infarct is predominantly an ill-defined lucency, osteomyelitis or neoplastic etiologies such as myeloma and metastasis can also enter the differential along with insufficiency fracture.

The sclerotic line seen radiographically correlates to a low signal line on T1 weighted MRI, which on T2 sequences demonstrates low signal with an adjacent hyperintense inner border known as the double line sign. A pseudomass appearance may result, with preservation of internal marrow fat distinguishing the infarct from tumor [13,14]. During the acute phase of infarction associated edema results in increased T2 signal. Differential diagnoses for the MR appearance include the same entities as radiographically.

On Tc99 MDP bone scan, bone infarcts can appear normal or result in a photopenic defect [15]. This is a nonspecific finding, with differential diagnoses including myeloma, metastatic disease, some sarcomas, postradiation changes, and artifact from overlying metal or orthopedic hardware. In the acute phase, there may be increased tracer uptake.

The imaging findings in AVN can mimic many other etiologies and may lead to extensive work up. Although rare, AVN can occur in the spine and needs to be considered in the differential diagnosis.

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References


