Case Report

Bilateral distal femoral bone infarction revealing polymyositis

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Abstract

Orthopedic manifestations, particularly bone infarctions, remain exceptional and unusual during polymyositis. They are classically related to systemic corticosteroids or other associated thrombotic conditions. We report an original observation of bilateral distal femoral bone infarction revealing polymyositis in a 42-year-old woman. Our observation is, to the best of our knowledge, the first to report bilateral femoral infarction as a possible manifestation of polymyositis apart from any prolonged corticosteroid therapy and any associated anti-phospholipid antibody syndrome.

Keywords: aseptic necrosis of bone, bone necrosis, osteonecrosis, polymyositis

Introduction

Polymyositis integrates with dermatomyositis and inclusion body myositis in the nosological framework of primitive inflammatory myositis of dysimmune origin [1,2]. Of all primary inflammatory myopathies, polymyositis is the rarest with an estimated overall incidence of 0.6 to 1.0/100,000 [2,3]. This polymyositis can sometimes have unusual clinical presentations and represent a real diagnostic challenge for clinicians [2,3].

Orthopedic manifestations, particularly bone infarctions, remain exceptional and unusual during polymyositis [4,5]. They are classically related to systemic corticosteroids or other associated thrombotic conditions [4,5]. In this paper, we report an original observation of bilateral distal femoral bone infarction revealing polymyositis without any other underlying thrombotic condition.

Case

A 42-year-old woman, with no pathological medical history, was explored for asthenia and proximal and distal muscular weakness of progressive onset for two weeks, associated with spontaneous and diffuse myalgia. The somatic examination noted an afebrile patient, myalgia caused by palpation of the different muscle groups of the upper and lower limbs, and a proximal and distal muscle deficit more marked in the pelvic girdle. Muscle testing was at 3-4 in the majority of skeletal muscles. The neurological examination was normal and
no skin lesions were noted. The ear, nose, throat, articular, cardiac, and pulmonary examinations were also without abnormalities. The biology showed marked rhabdomyolysis with creatine phosphokinase (CPK) at 11,000 IU/L, lactate dehydrogenase at 9,870 IU/L, aspartate-aminotransferase at 340 IU/L, alanine-aminotransferase at 289 IU/L, and a high erythrocyte sedimentation rate at 100 mm/H1. The rest of the basic biological tests were within normal limits: leukocytes at 8,200/mm³, hemoglobin at 14.6 g/dL, platelets at 450,000/mm³, creatinine at 98 μmol/L, calcium at 2.52 mmol/L, serum sodium at 148 mmol/L, serum potassium at 4.6 mmol/L, total cholesterol at 5.23 mmol/L, triglycerides at 1.48 mmol/L, fasting glucose at 5.2 mmol/L, and thyroid stimulating hormone at 2.05 μIU/mL. Electrocardiogram and chest X-ray were without abnormalities. The electromyogram revealed a severe myogenic syndrome of the muscles of the lower and upper limbs, without neurogenic signs. Muscular magnetic resonance imaging (MRI) revealed a discreet fatty invasion of the muscular of both thighs with intramedullary bone infarction at the distal ends of both femurs (Figure 1, 2, and 3).

Figure 1. T1-weighted coronal MRI of the thigh muscles: irregular and heterogeneous medullary bone infarction of distal ends of the two femurs.

Figure 2. T2-weighted coronal MRI of the thigh muscles: bilateral and distal femoral medullary bone infarction.

Figure 3. T2-weighted axial MRI of the thigh muscles: right femoral medullary bone infarction with bilateral muscular involution of thighs.

The infection investigation was negative. Immunological tests were positive for anti-nuclear autoantibodies and anti-Jo1 autoantibodies. Biopsy of the right quadriceps muscle confirmed the diagnosis of polymyositis. Subsequent investigations for this bone infarction were negative (anti-native DNA antibodies, anti-nucleosome antibodies, anti-cardiolipin antibodies, anti-β2-glycoprotein-1 antibodies, anticoagulant lupus, ANCA, cryoglobulin, protein C, protein S, antithrombin III, factor-V Leiden, electrophoresis of serum proteins, tumor markers, and thoracoabdominopelvic CT-scan). The diagnosis was that of polymyositis complicated by bilateral avascular necrosis of distal femurs.

The evolution was favorable under systemic corticosteroid therapy at the dose of 1 mg/kg/day augmented with hydroxychloroquine (400 mg/day) and salicylate acid at platelet anti-aggregating dose (100 mg/day). The CPK was 9,870 IU/L after one week, then 1,620 IU/L after two weeks, and 62 IU/L after one month of treatment.

Discussion

Bilateral osteonecrosis of the femurs is exceptional [6,7]. It is particularly reported in systemic lupus erythematosus and sickle cell disease [6,8,9] and appears to be favored by prolonged systemic corticosteroid therapy and the presence of antiphospholipid antibodies [6,7,10].
This aseptic bone necrosis predominates classically at the level of the femoral heads; distal femoral osteonecrosis is much rarer [6]. Medullary bone infarction remains mostly asymptomatic and would be fortuitously discovered on radiological examinations [9]. Conventional bone radiographs do not contribute to the diagnosis (often remain normal), and MRI is the test of choice for detecting bone infarction [9].

In the course of polymyositis, bone infarctions have been reported only twice previously [4,5]. In both cases, the direct cause was prolonged systemic corticosteroids (bone infarction was a complication of steroid treatment) [4,5].

Our observation is, to the best of our knowledge, the first to report bilateral femoral infarction as a possible manifestation of polymyositis apart from any prolonged corticosteroid therapy and any associated anti-phospholipid antibody syndrome. Bone infarction during polymyositis requires special attention because malignant degeneration remains a possible complication as evidenced by the observation of Dua et al. reporting the case of bone sarcoma secondarily developed on bone infarction in a 56-year-old woman followed for polymyositis [4].

**Conclusion**

Bone infarction remains an exceptional and unusual complication during polymyositis. It is often considered a complication of prolonged systemic corticosteroids therapy. Our observation is, to our knowledge, the first reporting bilateral femoral bone necrosis as a possible and revealing manifestation of polymyositis. This complication, as exceptional as it is, deserves to be known by clinicians because it presents a potential carcinogenic risk.

**Conflict of interest**

All authors declare that they have no conflict of interest.

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**References**