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CASE REPORT

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Sjögren's syndrome with bilateral spontaneous fracture of the femoral neck following aseptic necrosis of the femoral head

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Abstract A woman with Sjögren's syndrome concurrently suffered bilateral fractures of the femoral neck without any proximate cause, while she had aseptic necrosis of the femoral head resulting from the treatment for Sjögren's syndrome. According to previous reports, fractures of the femoral neck following aseptic necrosis of the femoral head are attributed to primary diseases such as systemic lupus erythematosus (SLE) or idiopathic thrombocytopenic purpura (ITP), and steroids are administered in most cases. Heterogeneous bone regeneration after necrosis causes irregularity in its mechanical strength depending on which regions of frailty are generated. Patients with aseptic necrosis of the femoral head whose bone density is considerably reduced need careful medical attention to avoid fractures of the femoral neck caused by weak forces.

Key words Aseptic necrosis of femoral head · Bilateral fracture · Femoral neck fracture · Sjögren's syndrome · Spontaneous fracture

Introduction

Idiopathic aseptic necrosis of the femoral head rarely involves fractures of the femoral neck prior to the decompression of the femoral head, and its etiology remains unknown. We studied a patient under treatment for Sjögren's syndrome who suffered aseptic necrosis of the femoral head and consequent bilateral fractures of the femoral neck almost simultaneously. Here, we report this case together with comments based on related references.

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Case report

A 60-year-old woman was administered steroids equivalent to 15 mg prednisolone per day, starting on May 15, 2003, following a diagnosis of Sjögren's syndrome by a dermatologist. The dose of medication was increased to 20 mg on September 17, and subsequently she took a maintenance dose. On October 14 she experienced pain in her right hip joint without any cause. On October 15 she came to our hospital. A plain X-ray film at her first visit did not indicate any fracture or necrosis (Fig. 1). A magnetic resonance imaging (MRI) scan on October 23 revealed a band-like low-signal intensity in the subcapital region of the left femur, and a patchy signal intensity in the region from the right femoral head up to the neck on the T1-weighted image (Fig. 2). Accordingly, she was diagnosed as having aseptic necrosis of the femoral head. She was advised to use crutches and was followed up. However, she continued to experience pain in her left hip. On November 12 she was admitted to the hospital with difficulty in walking because of aggravated pain since November 5. At the time of hospitalization, her body mass index (BMI) was 22.4, and the positions of her hip joints were normal. However, pain on motion and tenderness at the bilateral Scarpa's triangle were found. A plain X-ray film after admission revealed a left subcapital fracture and a right neck fracture of the femur, Garden IV and III (Garden classification), respectively (Fig. 3).

Prior to the administration of steroids, her serum calcium was 9.1 mg/dl (normal range 9–10.5 mg/dl), phosphate 3.6 mg/dl (normal range 3–4.5 mg/dl), chloride 108 mEq/l (normal range 98–108 mEq/l), potassium 3.5 mEq/l (normal range 3.4–4.9 mEq/l), and alkaline phosphatase (ALP) 442 U/l (normal range 85–300 U/l). At the time of admission, these levels were 9.1 mg/dl, 4.1 mg/dl, 104 mEq/l, 4.1 mEq/l, and 469 U/l, respectively. After admission, her bone ALP was 125 U/l (normal range 31–123 U/l) and her urine NTx was 991.1 (normal range <999). Total protein, including albumin, liver function, and renal function, were normal. Lumbar bone mineral density was measured by

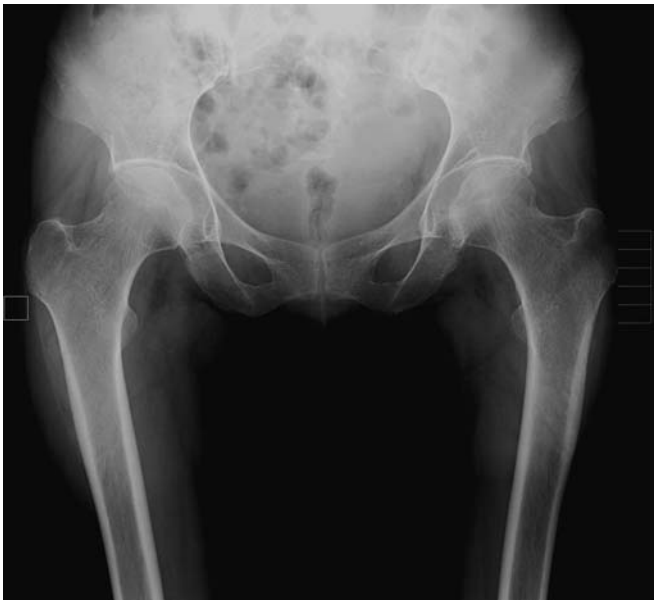

Fig. 1. There was no indication of fracture or aseptic necrosis of the femoral head on the plain X-ray film taken at the first visit

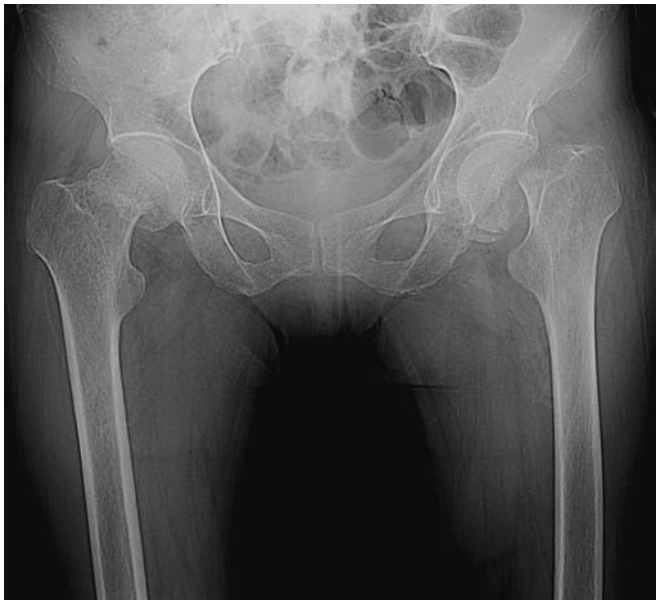

Fig. 3. A plain X-ray film taken after admission showing a subcapit fracture on the left and at the neck on the right. Garden classifications were stage IV and III, respectively

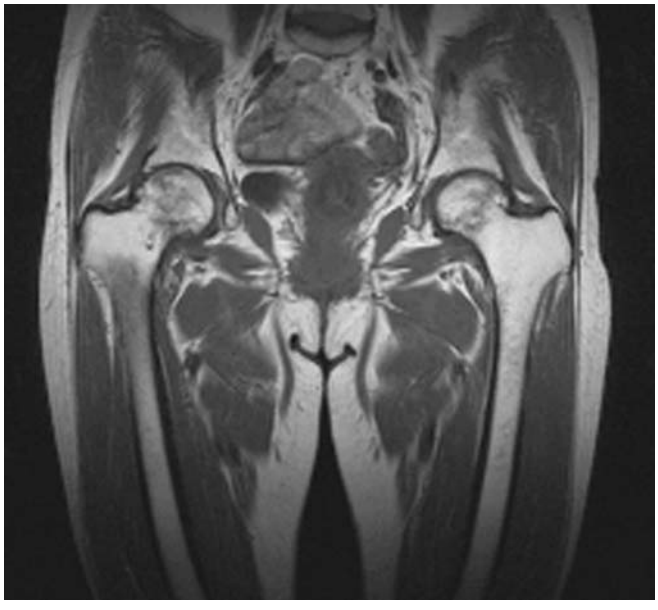

Fig. 2. T1-weighted magnetic resonance imaging showing a low-signal-intensity area centered around the subcapital zone in a band on the left hip, and porphyricity in the whole head to neck area in the right hip

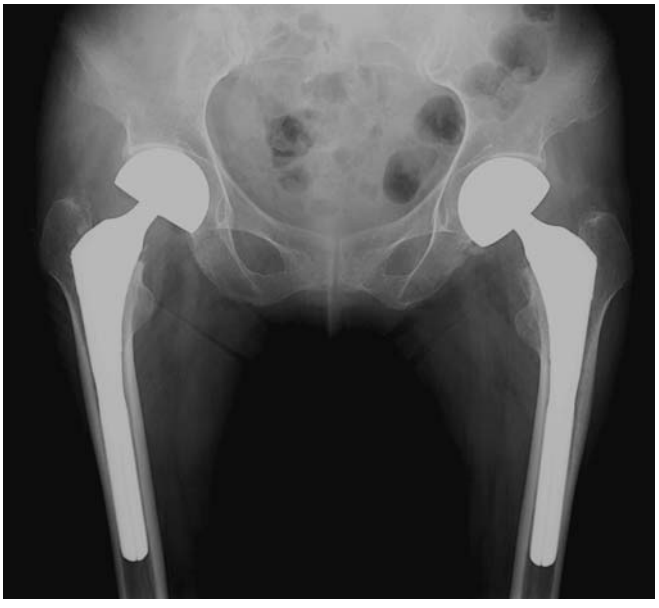

Fig. 4. A plain X-ray film taken after bilateral bipolar hemiarthroplasty

dual-energy X-ray absorptiometry (Stratec XR26S, DEXA). The young adult mean was 41.3% and bone mineral density (BMD) was 0.430 g/cm^2.

Bipolar hemiarthroplasty was first performed on her left hip on November 21 because the pain in the left hip was more intense than that in the right. Subsequently, bipolar hemiarthroplasty was performed on her right hip on December 12 (Fig. 4). At surgery, a moderate amount of yellow joint effusion was observed in the left hip, and a slight amount of a similar effusion was also observed in the right hip. Joint surfaces were normal on both the acetabular roof and the femoral head. The right femoral neck fracture was partly united. Sitting was allowed on December 19, and bisphosphonates were introduced for osteoporosis treatment. Full weight-bearing was allowed on December 26. The patient could walk with a T-cane 2 months after the first operation.

The histopathological findings of the resected right femur were necrosis of the trabeculae and bone marrow near the proximal femoral head, granulomatous replace-

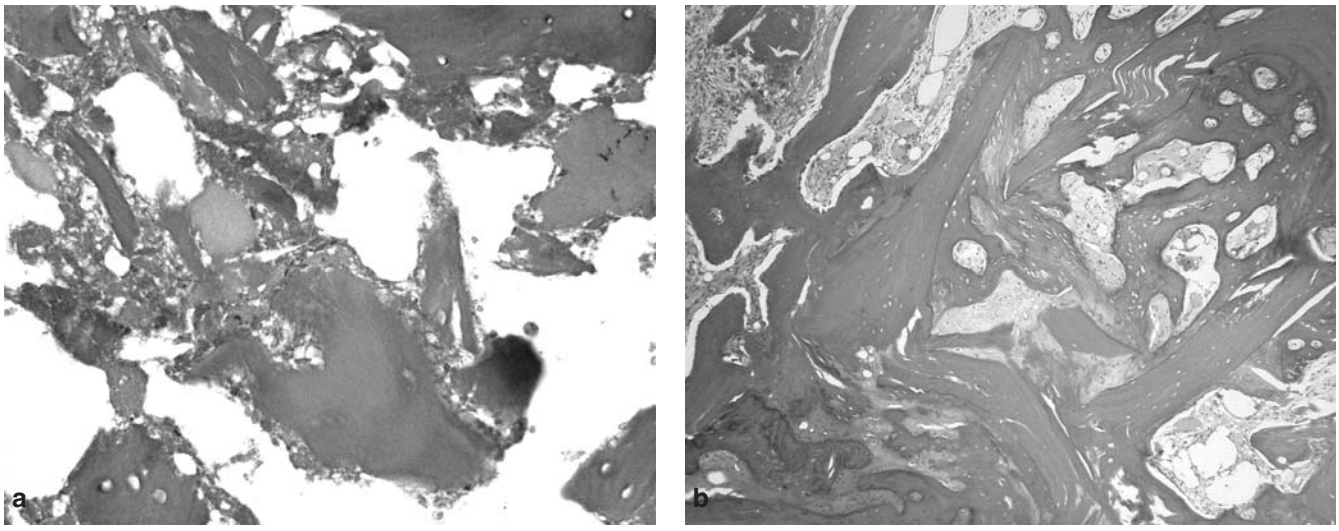

Fig. 5. **a** Histopathological observations of the extracted right femoral head showed necrosis of bone trabecula in the proximal head and on part of the bone marrow. **b** The distal bone trabecula showed an increase in the manner of regeneration or hyperplasia

ment and scarring, and regenerative or hypertrophic trabeculae around the necrosis (Fig. 5).

Discussion

Femoral neck fracture without any history of significant trauma, which is generally referred to as spontaneous fracture,[1] accounts for only 3.6% of femoral neck fractures according to the study of Hagino and Yamamoto.[2] The incidence of bilateral fractures is almost 5%. Accordingly, this case was a very rare one because the bilateral fractures occurred almost concurrently, no history of trauma was reported, and the neck fracture was caused without prior decompression of the bone head. Including this case, a total of 28 patients[3–9] with femoral head fracture or neck fracture following aseptic necrosis of the femoral head have been reported. However, this is the first report of a patient who had suffered bilateral neck fractures almost simultaneously.

The independent risk factors for femoral neck fracture are a reduction in bone density, a fall, decreased BMI, previous fractures, smoking, and consumption of meat, caffeine, and alcohol.[10] This case was not related to any risk factor except for a considerable reduction in bone density, which was considered to be the cause of the fractures.

Screening for the primary disease that causes osteoporosis is necessary in patients with bone density reduction and/or fragile fracture. In other words, secondary osteoporosis should be taken into consideration. Sjögren's syndrome frequently accompanies distal renal tubular acidosis (approximately 15% of all cases). Distal renal tubular acidosis inevitably causes systemic acidosis, leading to calcium reabsorption from the bones, which in turn results in an acceleration of osteoporosis. In this case, laboratory data did not support the possibility of renal acidosis or endocrinopathy. Secondary osteoporosis due to the administration of steroids was strongly suspected.

There are many reports of a reduction in bone density due to steroid use.[11–13] According to the general practice research database by Van Staa et al.[14] and a metaanalysis of clinical research, steroid administration alone significantly decreases bone density with or without a primary disease. Thus, the risk of fracture is strongly increased. They reported that the risk of nonvertebral bone fracture is considerably increased by 6–9 months of steroid administration, and that of vertebral bone fracture is increased by 3–6 months administration. The dose of steroids administered is correlated with the risk of fracture, and the total dose is strongly correlated with the reduction in bone density. Steroid administration equivalent to less than 5 mg glucocorticoid per day slightly increases the relative risk of nonvertebral bone fracture by 1.2 times. The risk increases with an increase in the daily dose. The relative risk is increased to nearly 1.6 times by the administration of 20 mg glucocorticoid. The total dose is significantly correlated with the bone density of the lumbar bones and the femoral head ($r = -0.58$, $r = -0.55$).

In this case, a daily dose of 15–20 mg had been administered for approximately 5 months until the fracture occurred. The total dose was nearly 2.5 g. The administration of steroids mainly decreased the density of the spongy bones, which resulted in aseptic necrosis of the femoral head. The neck fracture appeared to be related to the aseptic necrosis. Aseptic necrosis is not generally correlated with osteoporosis. Steroid pulse therapy for collagen disease involves serious bone damage.[15] Minamisawa[16] have reported the mechanism of femoral neck fracture after aseptic necrosis of the head. According to this mechanism, aseptic necrosis initially causes a fragile region in the bone. In this region, repetitive trabecular fractures, in combination with weak stresses inflicted by everyday activities, result in the neck fracture. In this case, the MRI scan before

the fracture showed that the aseptic necrosis extended throughout the head of the right hip and to the subcapital region of the left hip. The borders between the normal and necrosed regions coincided with the fracture lines. We presumed that the weak stresses caused the fractures, because the borders were rendered fragile and the bone density was considerably decreased.

According to the histopathological findings in the resected right femoral head, a reparative process appeared to follow the necrosis near the proximal head. Distal active osteogenesis caused the difference in the mechanical strength at the borders. Heterogeneous mechanical strength, in combination with a considerably reduced bone density and the weak force, caused the fractures at the borders.

Patients with aseptic necrosis of the femoral head caused by steroid use sometimes suffer femoral neck fracture induced by slight stress. Such developments should be monitored in these patients. Moreover, inadvertent steroid use should be avoided. According to the recommendations of the American College of Rheumatology (ACR),[17] bisphosphonates should be administered for primary prevention to patients receiving steroids equivalent to more than 5 mg of prednisolone for 3 months.

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