Transient Osteoporosis of the Hip in Association With Osteogenesis Imperfecta: Two Cases, One Complicated by a Femoral Neck Fracture

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Transient osteoporosis of the hip, a relatively uncommon cause of disabling hip pain, occurs spontaneously in men during the fourth and fifth decades of life and in childbearing females during the third trimester of pregnancy. It has been described in conjunction with osteogenesis imperfecta in 18 cases to date. Meta-analysis has suggested that people with osteogenesis imperfecta (vs the general population) are at increased risk for developing transient osteoporosis, suggesting the role of microfractures in the etiology of transient osteoporosis.

In this report, we present 2 additional cases of transient osteoporosis of the hip in conjunction with osteogenesis imperfecta, including the first case of a spontaneous pathologic fracture in a male patient with both disorders.

Case Reports

Case 1

A man in his late 40s with osteogenesis imperfecta presented with a 5-week history of severe left hip pain. He reported spontaneous onset of left hip pain, which over 5 weeks progressed to incapacitation. There were no precipitating traumatic incidents or other identifiable predisposing factors. Past medical history was significant for a tarda form of osteogenesis imperfecta compounded by multiple previous fractures. Of significance, the patient had sustained a left femoral neck fracture at age 15, a right femoral shaft fracture treated with intramedullary fixation in his late 30s, and a left patella fracture treated with open reduction and internal fixation at age 42. The patient had immediate family members who also had been diagnosed with osteogenesis imperfecta.

On examination, the patient had a moderate limp favoring the left lower extremity, his sclerae had a blue tinge, he had shortened small finger metacarpal bones bilaterally, and range of motion (ROM) of the left hip was moderately restricted and painful at extremes of motion.

"[In men this] occurs spontaneously...during the fourth and fifth decades of life"

The patient was initially evaluated at an outside institution, where plain x-rays showed osteopenia of the left femoral head and neck (Figure 1). No fractures or dislocations were noted. A magnetic resonance imaging (MRI) scan of the left hip showed a mild effusion of the hip joint, decreased signal intensity in the left femoral head and neck on T_1-weighted images, and significantly increased signal intensity of the femoral head and neck on T_2-weighted images (Figures 2A, 2B). The patient was then referred to our institution for definitive management.

The patient was diagnosed with transient osteoporosis of the left hip. A regimen of protected weight-bearing with use of 2 crutches was recommended, and he was placed on mild analgesic medications. A week later (or 6 weeks in the course of the disorder), he slipped and sustained a "torquing" force to the left hip. At that time, he also noted a "snapping" sensation and intense pain in the left hip. He proceeded immediately to the emergency department at our institution, where plain x-rays of the left hip revealed a minimally displaced transcervical femur fracture (Figure 3). He underwent percutaneous fixation with three 7.3-mm cannulated screws and was kept on a minimal weight-bearing postoperative regimen.

Two weeks after the operative procedure, the patient was still suffering from intense pain both during activity and at rest. Plain x-rays of the left hip revealed satisfactory alignment of the fracture fragments; however, the femoral

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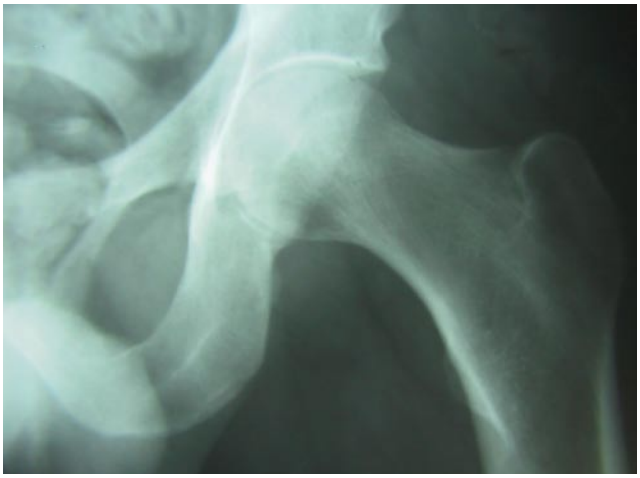

Figure 1. Case 1—Anteroposterior x-ray of left hip reveals osteopenia of femoral head and neck.

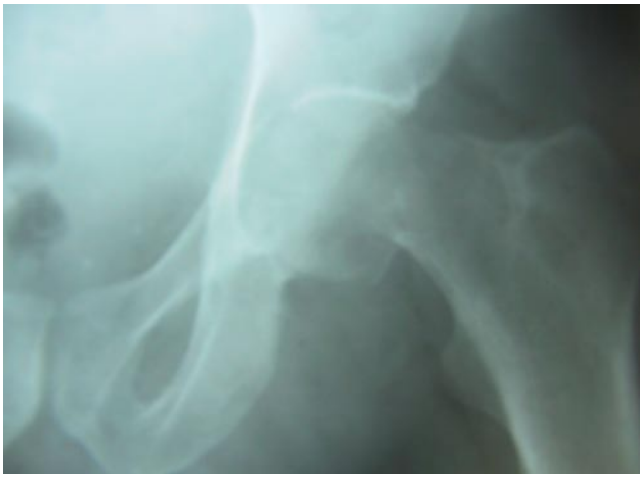

Figure 3. Case 1—Anteroposterior x-ray of left hip shows transcervical femoral neck fracture.

head and neck were notably more osteopenic than in prior studies. The patient subsequently underwent a metabolic bone workup that was significant for an increased level of urinary deoxypyridinolene cross-links and a DEXA (dual-energy x-ray absorptiometry) scan that revealed a bone mineral density that was less than 3 SD below normal for his weight and age. He was placed on a course of intravenous pamidronate therapy.

Three months after the operative procedure, the pain in the left hip had subsided substantially, and motion had improved to a normal range. However, plain x-rays continued to show evidence of inadequate consolidation of the fracture fragments despite adequate alignment. Weight-bearing status was gradually increased, and the patient underwent a course of electromagnetic bone stimulation. Two months later, plain x-rays showed some evidence of fracture consolidation, and he was advanced to 50% weight-bearing. Eight months after the operative procedure, he was fully weight-bearing without pain or use of assistive devices. Plain x-rays revealed full consolidation of the fracture (Figure 4).

The patient, now 5 years after the operative procedure, stated that he had no discomfort in the right hip. He was capable of ambulating without assistive devices for an unlimited distance without pain. There was no perceivable limp or leg-length discrepancy. ROM was 120° flexion, 5° extension, 40° abduction, 20° adduction, 15° internal rotation, and 40° external rotation. Harris Hip Score was 100 points (100 possible).

Case 2

A man in his mid-60s with a history of osteogenesis imperfecta was referred to our institution with a 4-month history of severe left hip pain. The sudden-onset pain was not associated with antecedent trauma. Within the first week of onset, the pain was severe enough that the patient began to limit weight-bearing. He had been diagnosed with osteogenesis imperfecta at approximately 3 years of age. He noted that he had sustained as many as 34 fractures of multiple bones by age 13. All of his children had had multiple fractures, had blue sclerae, and had been diagnosed with osteogenesis imperfecta.

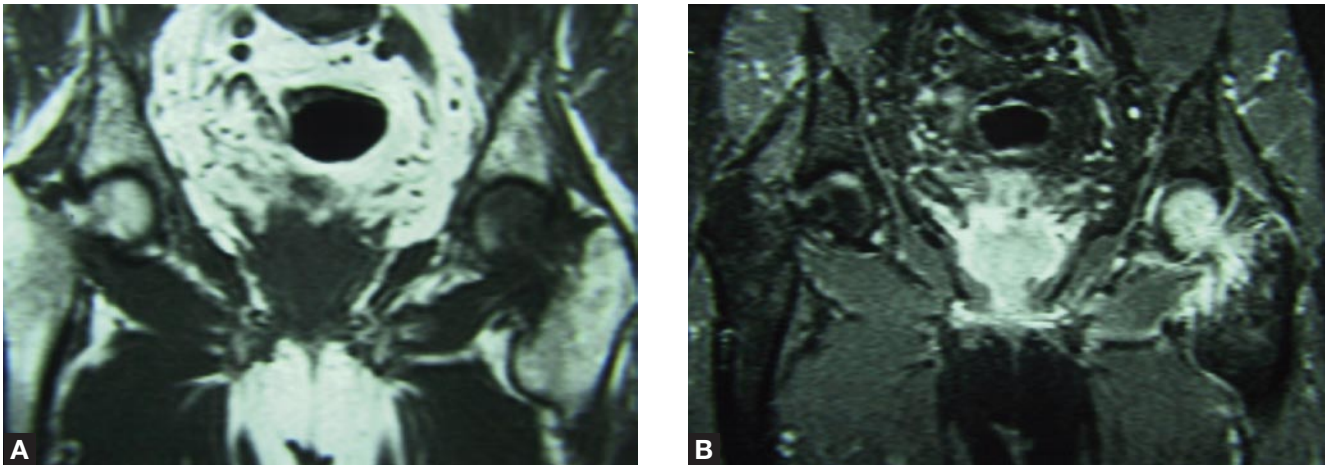

Figure 2. Case 1—**(A)** T_1-weighted magnetic resonance image of hip shows decreased signal intensity in femoral head and neck suggesting edema. **(B)** T_2-weighted magnetic resonance image of hip shows increased signal intensity in femoral head and neck suggesting edema.

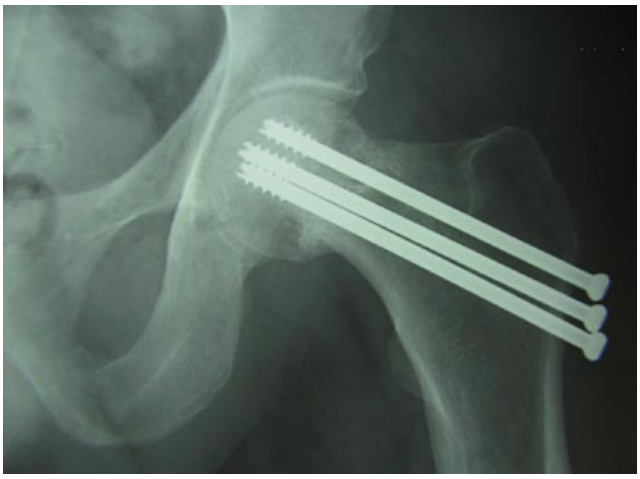

Figure 4. Case 1—Anteroposterior x-ray of left hip at 8 months follow-up shows healing of femoral neck fracture.

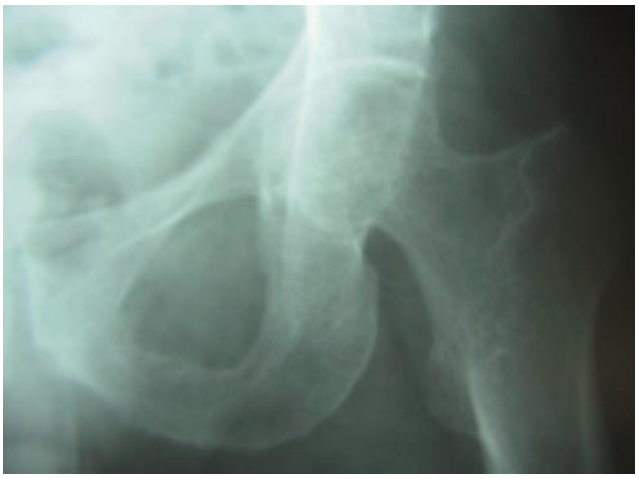

Figure 5. Case 2—Anteroposterior x-ray of left hip reveals osteopenia of femoral head and neck.

On examination, the patient had blue sclera, ambulated with an antalgic gait favoring the left lower extremity, had 20° external and 10° internal left-hip rotation limitations (vs the contralateral, unaffected side), and had significant discomfort at the left-hip rotation extremes.

Plain x-rays of the left hip were notable for osteopenia involving the left femoral head and neck (Figure 5). MRI showed decreased signal intensity on T_1-weighted images and increased signal intensity on T_2-weighted images of the left femoral head and neck, as well as a small effusion of the hip joint (Figures 6A, 6B). Bone scintigraphy demonstrated increased uptake in the left femoral head and neck.

A diagnosis of transient osteoporosis of the left hip was made, and the patient was treated nonoperatively with protected weight-bearing using crutches and analgesics. The patient continued to have severe pain despite these measures. Five months after disorder onset, he underwent core decompression to facilitate resolution of this severe pain. A specimen sent for pathologic examination showed hypervascular marrow; undecalcified sectioning was not performed. Four to 6 weeks after decompression, the patient noted spontaneous resolution of the hip pain. Plain x-rays of the hip revealed resolution of the osteopenia of the femoral head and neck.

Three years after resolution of the transient osteoporosis, the patient reported that the left hip was asymptomatic, except for occasional fleeting discomfort after prolonged immobilization. He ambulated with a slight limp secondary to a left knee flexion contracture that developed after treatment for a distal femur fracture. Examination of the hip revealed 120° flexion, 5° extension, 15° internal rotation, and 40° external rotation. Harris Hip Score was 87 points (100 possible).

Discussion

Transient osteoporosis of the hip is an uncommon cause of disabling hip pain. Since it was first described by Curtiss and Kinkaid[1] in 1959, approximately 500 cases have been documented in the literature.[2] The disorder afflicts males nearly 3 times more commonly than females.[3] The typical male patient is in his 30s or 40s and has no apparent predisposing conditions. The disorder in females occurs almost exclusively in pregnancy during the third trimester.[4]

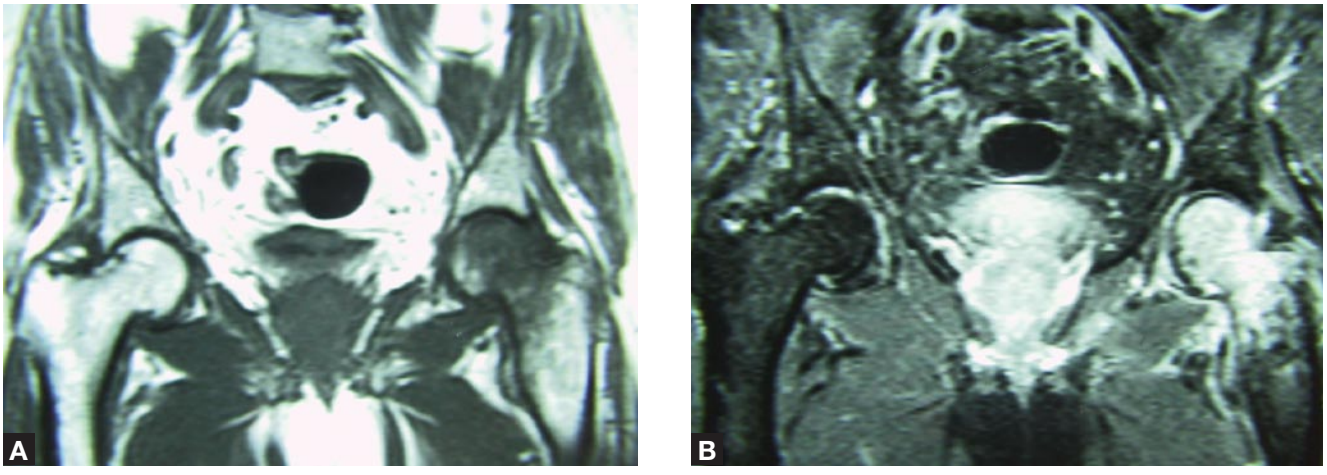

Figure 6. Case 2—**(A)** T_1-weighted magnetic resonance image of hip shows decreased signal intensity in femoral head and neck suggesting edema. **(B)** T_2-weighted magnetic resonance image of hip shows increased signal intensity in femoral head and neck suggesting edema.

The afflicted patient classically presents with acute onset of hip and/or groin pain without antecedent trauma. ROM of the involved hip is typically slightly restricted and painful at the extremes of motion.

Imaging Findings. Plain x-rays are normal early in the course of the disease process. Within 4 to 6 weeks of symptom onset, localized osteopenia of the affected femoral head and neck can be seen. MRI can reveal changes acutely, within a few days of symptom onset.[5] On T_1-weighted images, the affected extremity reveals decreased signal intensity diffusely throughout the femoral head and often extending into the femoral neck to the level of the intertrochanteric line. Conversely, on T_2-weighted images, the femoral head and neck reveal dramatically increased signal intensity, consistent with bone marrow edema. A joint effusion is also typical on T_2-weighted images.

Management. The pain is usually self-limited, resolving in 6 to 8 months in most cases and without surgical intervention. Conservative management with protected weight-bearing is recommended for most cases. Core decompression, which has been advocated by some, particularly in Europe, reportedly results in earlier resolution of symptoms according to one study.[6] Efficacy of core decompression in management of transient osteoporosis of the hip is limited to symptomatic relief. We do not advocate it as a standard of care, as supportive evidence is lacking, and the risks associated with its use (pathologic fracture, in particular) are significant.

To our knowledge, transient osteoporosis of the hip has been reported in conjunction with osteogenesis imperfecta in only 20 cases, including the 2 presented here.[7,8] Incidence of osteogenesis imperfecta is estimated to be 1:20,000 to 1:50,000.[9] A meta-analysis performed by Noorda and colleagues[7] suggested a statistically significantly ($P<.0001$) increased risk for transient osteoporosis in patients with osteogenesis imperfecta versus the general population. Our experience supports their analysis. The senior author, Dr. Marvin E. Steinberg, who developed a tertiary referral practice targeting osteonecrosis and transient osteoporosis of the hip, has evaluated approximately 40 patients with transient osteoporosis of the hip over the past 15 years. Of these patients, 2 were diagnosed with osteogenesis imperfecta before onset of transient osteoporosis (their cases are presented in this report). Although the relationship between transient osteoporosis and osteogenesis imperfecta is unclear, several authors have hypothesized that microfractures may have a role in the etiology of transient osteoporosis.[7,8] This hypothesis seems particularly plausible in transient osteoporosis cases that develop in patients with osteogenesis imperfecta.

Our literature search turned up 8 case reports of a femoral neck fracture in association with transient osteoporosis of the hip.[1,2,10-14] Seven of the 8 cases involved pregnant women; the eighth case was that of a male patient who sustained the fracture during a biopsy procedure.[14] In addition, Dr. Steinberg, the senior author, treated 2 pregnant women who sustained femoral neck fractures as a complication of transient osteoporosis of the hip: One woman healed without sequelae after open reduction and internal fixation, and the other eventually required total hip arthroplasty (unpublished data). To our knowledge, the patient described in case 1 of our report is the first nonperioperative case of a femoral neck fracture in a male patient with transient osteoporosis of the hip, as well as the first femoral neck fracture in a patient with both transient osteoporosis and osteogenesis imperfecta. The fracture presented was sustained through a low-energy mechanism, thus illustrating the importance of maintaining a restricted weight-bearing status until resolution of the transient osteoporosis has occurred, particularly when it occurs in conjunction with osteogenesis imperfecta.

This report also represents the first published case of delayed union in a patient with transient osteoporosis of the hip. Delayed union and nonunions are known to occur in patients with osteogenesis imperfecta, whereas they are exceedingly rare in otherwise healthy children.[9] It is unclear whether the delayed healing of the pathologic fracture sustained by the aforementioned patient is attributable to the transient osteoporosis, the underlying osteogenesis imperfecta, or a combination of the two, though we suspect that osteogenesis imperfecta was dominant in its role.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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This paper will be judged for the Resident Writer's Award.