Total hip arthroplasty in a patient with neurofibromatosis type I and recurrent spontaneous hip dislocation

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A 26-year-old woman came to the hospital with her third spontaneous posterolateral dislocation of the left hip, which had occurred as this young woman was getting out of a boat (Fig. 1). Her history included a diagnosis at 9 months of age of neurofibromatosis (NF) type I.

Reduction of the hip dislocation with traction was easy under general anesthesia, with no resultant complications, as confirmed by standard radiographs, arthrogram and arthroscan. During the following 2 years she had 3 other episodes of hip dislocation from trivial trauma. A shelf operation and fascia lata tenorraphy led to a symptom-free period of 8 years until a dislocation recurred, with subsequent development of progressive hip arthritis.

She underwent total hip arthroplasty. During that procedure, we noted that her hip muscles were atrophied and her capsulo-ligamentous structures loose, but found was no evidence of neurofibroma in or around the hip joint. After the hip replacement, an intraoperative examination demonstrated poor stability. To tighten the gluteal muscles, we performed a trochanteric osteotomy, fixed with a Downmills hook.

Afterward the patient had another episode of dislocation and developed recurrent instability of her artificial joint. This necessitated the replacement of the acetabular component by a constrained liner (Fig. 2).

At follow-up 18 months later, she has had no further dislocations.

Discussion

To the best of our knowledge, this is the first reported case of recurrent hip dislocation in a patient with NF without any evidence of intra-articular neurofibromas or bony anomalies of the hip joint. A review of the literature revealed only 7 hip dislocations that have been reported in patients with NF I, including 2 gradual dislocations in 2 young children[1] and 5 acute dislocations after trivial trauma in older patients.[2–5]

The following triad was described in acute dislocations by Guillemet and colleagues[2]: skin lesions discovered at birth, hypertrophy of the lower limb in childhood, and spontaneous dislocation of the hip in adult life. All the cases of dislocations reported were associated with intra-articular neurofibromas and bony chan-

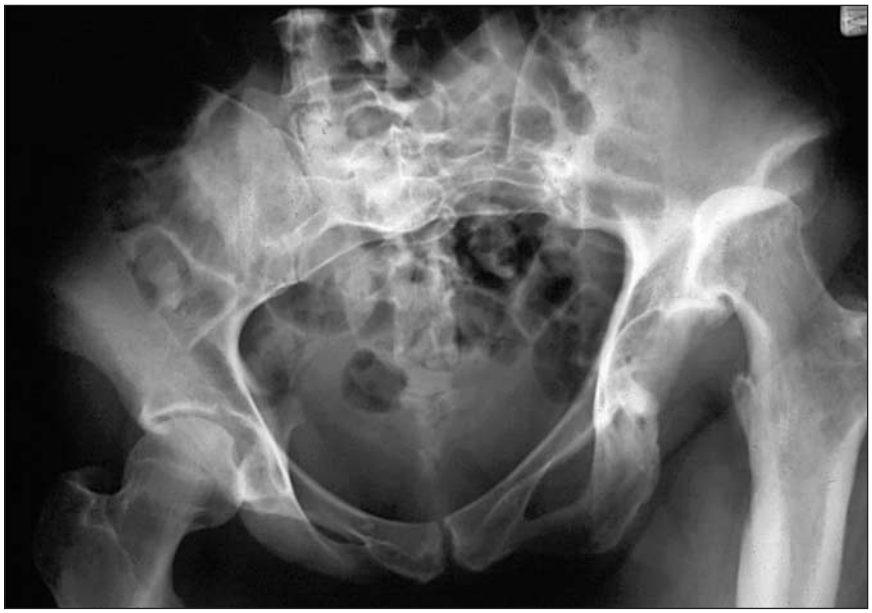

Fig. 1. Posterolateral hip dislocation (third episode).

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ges, including acetabular dysplasia, erosion and growth disturbance with resultant coxa valga caused by neurofibroma invasion. Furthermore, mechanical problems such as weakness of the abductor muscles caused by spinal-cord tumours were also reported as possible predisposing factors.[1]

In our case, we found no anatomical or mechanical abnormalities such as neurofibromas to explain the problem. The evolution toward degenerative changes, instability and dislocation could suggest a deficiency of the normal sensory protection of the hip. This hypothesis was emphasized by Phillips and McMaster[5] because of the absence of pain during progressive dislocation in neuropathic arthropathy.

For our patient, because her soft tissue was not strong enough to restrain the hip joint, further joint dislocation could be prevented only by mechanical stabilization, including a shelf operation and insertion of a constrained-liner acetabular component. Despite the short-term good result, long-term prognosis is of concern because of her high risk of loosening of the acetabular component and early wearing in the absence of normal sensation.

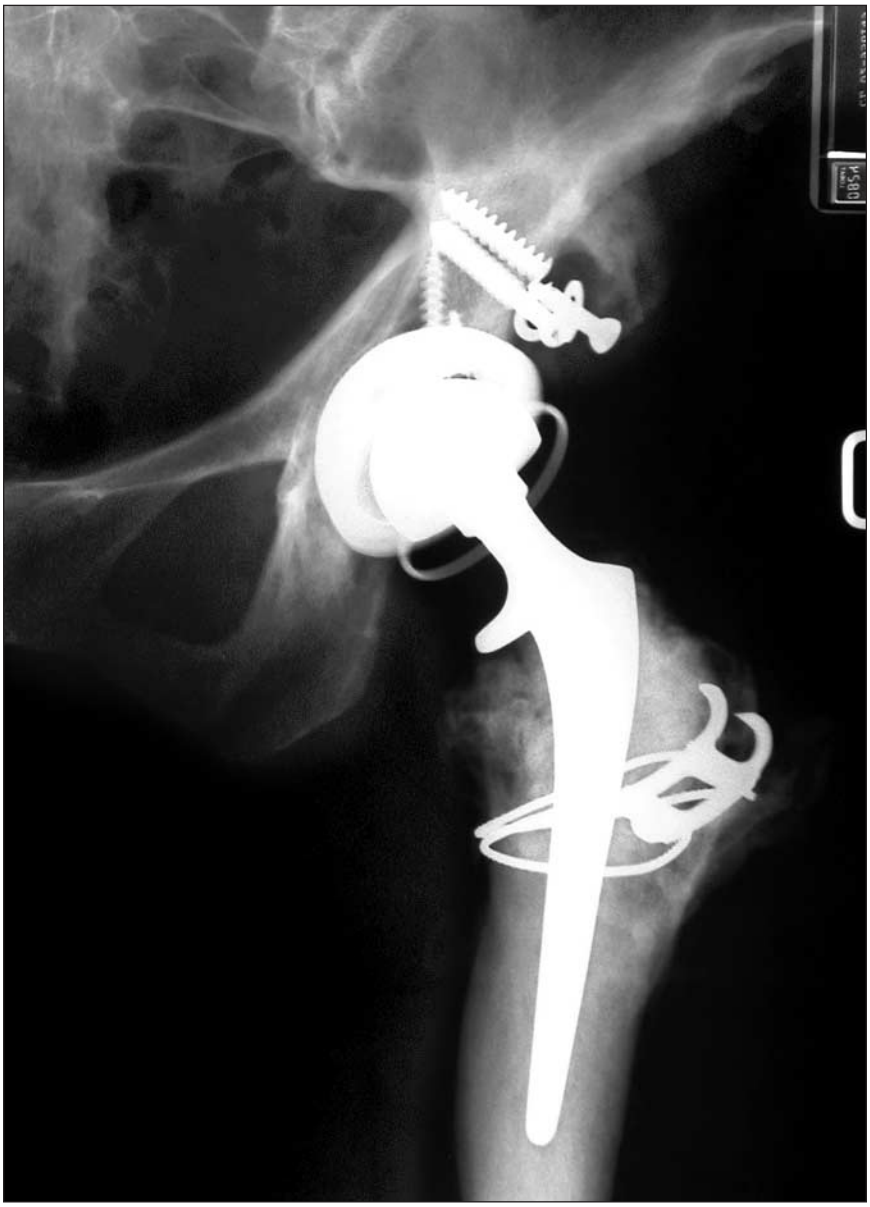

Fig. 2. Total hip arthroplasty with a constrained liner.

Competing interests: None declared.

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