Injury Extra (2008) **39**, 279–280

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CASE REPORT

Intracapsular femoral neck fracture in a patient with Hypomelanosis of Ito

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Accepted 26 February 2008

Introduction

The case of a child with a diagnosis of Hypomelanosis of Ito — a systemic neurocutaneous disease with features akin to neurofibromatosis and tuberous sclerosis — who sustained an intracapsular femoral neck fracture following a fall is presented here, together with an overview of this interesting syndrome.

To the authors knowledge, there are no other reports in the literature of a patient with this syndrome presenting with an intracapsular femoral neck fracture.

Case report

A 13-year-old girl with learning difficulties, normally ambulant without walking aids, presented to the emergency department of our institution following a fall down a flight of stairs. It was not possible to obtain a history from the child herself, but her accompanying parent had noted that the child was unable to weight bear on the right lower limb following the episode of trauma. Furthermore it transpired that the child had previously been diagnosed to have a syndrome referred to as "Hypomelanosis of Ito", which in her case manifested itself in the form of cutaneous hypopigmentation, severe intellectual developmental delay, epilepsy and thoraco-lumbar scoliotic deformity.

Primary and secondary surveys revealed an isolated injury—an undisplaced intracapsular fracture of the femoral neck. Diagnosis purely on clinical grounds was made more difficult by the fact that the child was incapable of expressing to the examining physician whether certain movements (including movements of the hip) were painful or not. Radiological examination of the right hip however confirmed the clinical suspicion of a hip fracture as being the cause of the child's reluctance to weight bear on the right lower limb (see Fig. 1a and b).

On the next scheduled operating list, the patient's fracture was stabilized by means of two 4 mm partially threaded cannulated screws (Smith & Nephew Inc., Memphis, TN, USA) passed across the fracture line but not violating the physis. Post-operatively the patient was not able to comply with weight bearing instructions and according to her parents was back to her pre-injury mobility status, fully weight bearing on the operated limb 3 weeks after surgery. Follow up X-rays in clinic approximately 6 weeks following surgery revealed a satisfactory result (see Fig. 2a and b).

Discussion

The disorder Hypomelanosis of Ito first described in 1952[3] is characterised by macular cutaneous hypopigmentation in a distinctive pattern of whorls, streaks and patches which may be unilateral or bilateral. This phenotype is believed to result from the random distribution of two populations of melanocytes with different pigment producing potential along the primitive streak early in embryogenesis and from their subsequent dorsoventral migration.[2] The paths of migration known as Blaschko Lines[4] are not dermatomal in origin and do

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1572-3461
doi:10.1016/j.injury.2008.02.035

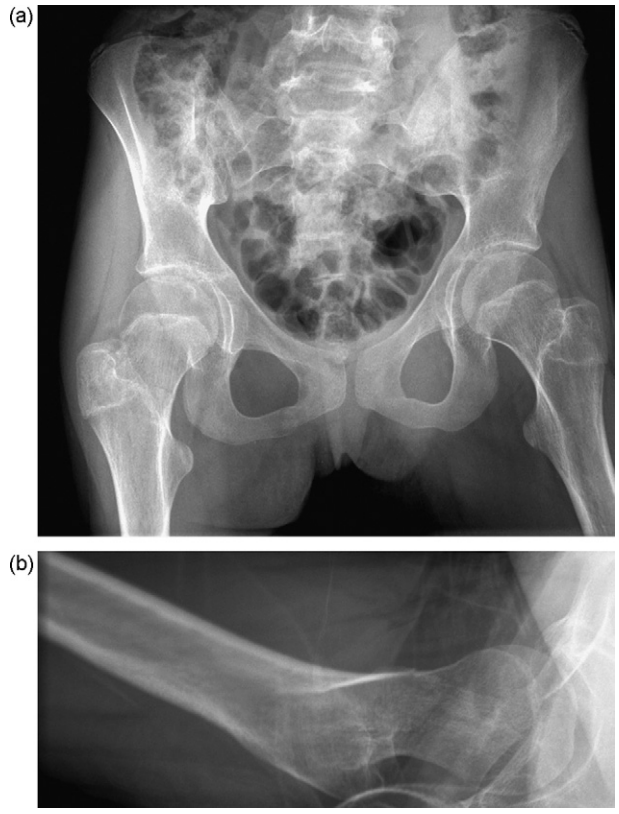

Figure 1 (a) Pre-operative antero-posterior radiograph of patient's pelvis and (b) pre-operative lateral radiograph of patient's right hip.

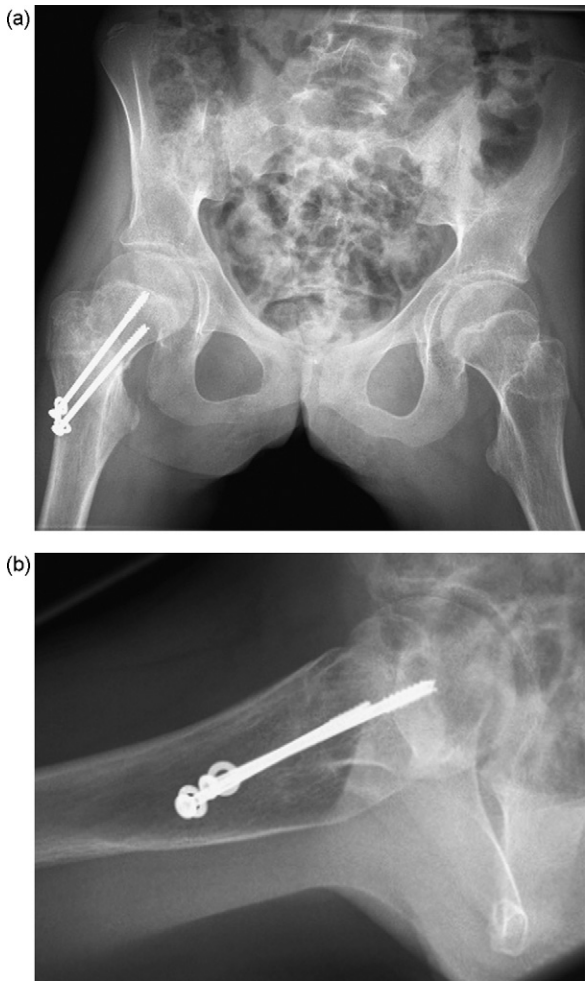

Figure 2 (a) Post-operative antero-posterior radiograph of patient's pelvis and (b) post-operative lateral radiograph of patient's right hip.

not conform to Langer's lines. The typical skin lesions are often noticeable at birth and are usually detected in the first year of life. In children with fair skin (such as the one described in this report) the use of a Woods Lamp (ultraviolet light) is useful in demonstrating the characteristic lesions[1] which occur on the trunk, legs, arms and face. (Unfortunately it was not possible to persuade the patient in this report to be examined under a Woods Lamp. Pictures of the characteristic lesions seen however may be viewed on the world wide web).

Although an autosomal dominant mode of inheritance has been suggested, [5] the vast majority of cases are sporadic and arise de novo.[1] Chromosomal mosaicism is believed to be the reason why patients with this syndrome are so varied in phenotype.[6] The disorder occurs more commonly in females, and in some individuals cutaneous hypopigmentation may be the only manifestation of the disorder.[1] There is no consistent pattern of associated anomalies although, when they do occur central nervous system dysfunction, particularly seizures and delayed (intellectual) development occur especially frequently.

From a musculo-skeletal perspective, patients with this syndrome may present to an orthopaedic surgeon in a number of other guises in addition to the presentation presented in this report including hemihypertrophy which is usually ipsilateral to the hypomelanotic lesions, arm and leg length discrepancies, with abnormalities of hand anatomy including atrophy, syndactyly, polydactyly, clinodactyly or bifid thumb, and with abnormalities of the lower limb including luxatio coxae and genu valgum.

In conclusion, although intracapsular femoral neck fractures are rare in children, a thorough and systematic approach to detect such injuries through the use of primary and secondary surveys should be adopted, especially in situations where patients may have learning difficulties resulting in them being poor historians or unable to effectively participate in the clinical examination to detect such injuries.

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