Unusual association of diseases/symptoms

Perthes' disease and femoroacetabular impingement in a child with proximal femoral focal deficiency

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Summary

A girl with known proximal femoral focal deficiency presented with Perthes' disease at 5 years of age. Her treatment involved a Salter osteotomy. This in conjunction with articular incongruence, due to deformity of the femoral head, resulted in mixed type femoroacetabular impingement when she was 10 years old. Surgical hip dislocation and femoral neck osteochondroplasty successfully relieved her symptoms of impingement. This is the first reported case of Perthes' disease in a patient with proximal femoral focal deficiency. The case highlights the importance of thoroughly investigating pain in patients with proximal femoral focal deficiency, a condition which is normally painless. Timely diagnosis of Perthes' disease and containment procedures can prevent collapse of the femoral head and the resultant sequelae. Acetabular over-coverage should be avoided in pelvic osteotomy to prevent the development of femoroacetabular impingement.

BACKGROUND

This case is unique in reporting the occurrence of Perthes' disease in a girl with proximal femoral focal deficiency and serves to alert clinicians to the possible association of the disorders.

CASE PRESENTATION

This female, Chinese patient was referred, at birth by the paediatricians who attended to her in the postpartum period, due to the presence of asymmetrical groin creases. Clinical evaluation revealed a bowed right femur that was shortened by 2 cm compared with that in the contralateral limb. Passive right hip abduction was limited to 45°. The examination was otherwise unremarkable. The diagnosis of proximal focal femoral deficiency (PFFD) was confirmed on plain radiograph (figure 1). Interval plain radiographs subsequently showed normal femoral head morphology, normal acetabular development, a continuous bony connection between femoral head and diaphysis and a mild varus deformity in the subtrochanteric region. These features are consistent with an Aitken type A¹ PFFD (figure 2). Her right hip and knee were stable, her foot was plantigrade and she exhibited none of the other associated abnormalities.

Initially, the management of this patient was observant and she was followed-up annually. She remained asymptomatic until the age of 5 years, when she began to complain of mild, self-limiting hip pain. A scanogram was performed and judged to have no abnormal features other than the known PFFD. The discrepancy in limb length had progressed to 4.2 cm by 5 years of age. Paley's² multiplier method predicted that this would reach approximately 7 cm by the time the patient reached skeletal maturity. At the age of 5 years she was commenced on a shoe raise, with the anticipation to surgically lengthen the limb at a future date. The patient and family were

reassured with regard to hip pain and annual follow-up was continued.

The following year, when she was 6 years old, hip pain had worsened and she walked with an asymmetrical gait. Thomas' test revealed a 10° fixed flexion contracture. Internal rotation and abduction were moderately reduced in range. A plain radiograph of the right hip showed features consistent with established Perthes' disease, in addition to coxa vara and coxa brevia (figure 3). Connective

Figure 1 Plain radiography of the patient's right hip at 5 months showing a shortened right femur, with varus angulation proximally, increased teardrop to metaphysis distance and a normal acetabulum.

Figure 2 Plain radiograph of the patient's right hip at 3 years showing normal femoral head and acetabulum development, a bony connection between femoral head and the diaphysis, and a shortened right femur with a varus deformity in its proximal portion.

tissue diseases and skeletal dysplasias were excluded clinically, biochemically and radiologically. At the time of diagnosis she was Herring's C³ and in the late fragmentation stage.⁴

The diagnosis of Perthes' disease prompted the scano-gram, performed when she first complained of hip pain a year earlier, to be revisited. On retrospective evaluation,

Figure 3 Plain radiograph of the patient's right hip at 6 years showing fragmentation, greater than 50% collapse and lateral extrusion of right femoral capital epiphysis.

the femoral capital epiphysis had already shown signs of early fragmentation, flattening and lateral extrusion (figure 4).

To prevent further extrusion and deformation of the femoral head, containment surgery was contemplated.^{5 6} A hip arthrogram showed the right femoral head was optimally contained with the hip in flexion and abduction. There was no hinged abduction. A Salter osteotomy⁷ was performed to increase anterior and lateral coverage of the acetabulum. A varus femoral osteotomy was not performed due to preexisting coxa vara. Resultant femoral remodelling followed, and at 4 years postoperatively the femoral head was aspherical, but congruent with the acetabulum (figure 5). She did well clinically in the short to medium term and at 3 years follow-up her range of right hip movement was 120° of flexion, 35° of abduction and 20° of internal rotation.

The patient began to complain of recurrent right groin pain at 4 years postoperative follow-up. Pain could be reproduced when the hip was put into 100° of flexion, 20° of adduction and 20° of internal rotation. The finding was compatible with a positive anterior impingement sign of femoroacetabular impingement.⁸ The plain radiograph revealed an aspherical femoral head, with an impingement bump on the femoral neck and acetabular retroversion (figures 6 and 7). A MR arthrogram showed a reduced α angle, an increased anterior femoral offset and a labral tear. The mixed cam and pincer femoroacetabular impingement in this patient was likely to be caused by proximal femoral growth disturbance as a sequela of Perthes' disease and iatrogenic anterior acetabular over-coverage from Salter osteotomy. The severity and persistence of her symptoms necessitated surgical intervention. Femoral neck osteochondroplasty, debridement of the labral tear and distal trochanteric advancement were

Figure 4 Scanogram of the patient's right hip at 5 years, when she first complained of hip pain, showing some early features of Perthes' disease.

Figure 5 Plain radiograph of the patient's right hip at 7 years, 1 year after a Salter osteotomy was performed for containment.

performed following an open approach and formal hip dislocation (figure 8). One year after surgery, she was pain free with a full range of hip motion. Her leg length discrepancy was measured to be 5.2 cm.

DISCUSSION

PFFD has an incidence of 2 per 100 000 children.⁹ In the Asian population, the incidence of Perthes' disease is 3.8

Figures 6 Plain radiograph of the patient's right hip 4 years after containment surgery showing an aspherical femoral head, an impingement bump on the femoral neck and features of acetabular retroversion including crossover sign (posterior wall of acetabulum crossovers instead of lying lateral to the anterior wall of the acetabulum) and posterior wall sign (center of femoral head lies lateral to the posterior wall of the acetabulum).

Figures 7 Plain radiograph of the patient's right hip 4 years after containment surgery showing an aspherical femoral head, an impingement bump on the femoral neck and features of acetabular retroversion including crossover sign (posterior wall of acetabulum crossovers instead of lying lateral to the anterior wall of the acetabulum) and posterior wall sign (center of femoral head lies lateral to the posterior wall of the acetabulum).

per 100 000 children.¹⁰ This is the first reported case, in the literature, of Perthes' disease in a patient with known PFFD.

PFFD is a rare, congenital malformation of the femur, of unknown aetiology, characterised by the failure of normal proximal femoral development. The spectrum of the deformity ranges from slight shortening to complete amelia. While Aitken's¹ classification subdivides PFFD based upon the severity of osseous abnormality, there are associated muscular and vascular abnormalities which have been described. Knowledge of these abnormalities is important if surgery is to be performed. Pirani *et al*¹¹ reported most muscles around the hip joint to be present, but hypoplastic, from MRI studies. Chomiak *et al*¹²

Figure 8 Plain radiograph of the patient's right hip following surgical intervention for femoroacetabular impingement.

performed CT angiographic studies and reported that many of their PFFD patients had external iliac, femoral and deep femoral arteries of reduced length and diameter. The deep femoral artery arose more proximally than normal. In cases of PFFD, there is a high incidence of fibular haemimelia, dysplastic subluxed patella, genu valgum, absence of lateral rays, tarsal coalition and vertical talus. The number of associations and the variable severity of PFFD necessitates highly individualised management. Patients may present with leg length discrepancy, hip, knee or ankle joint instability, proximal femoral deformity, pseudoarthrosis and deformity associated with fibular haemimelia. In the case reported, the patient suffered from a mild form of PFFD with approximately 7 cm of expected leg length discrepancy at skeletal maturity, stable hip and knee joints, a plantigrade and functional foot. Prior to the development of Perthes' disease, the goal of treatment was, therefore, the equalisation of leg lengths. PFFD is thought to be generally painless, despite the presence of pseudoarthroses and soft tissue abnormalities.¹³ In patients with new onset groin pain, the treating clinician should, therefore, be alert to concomitant hip or sacroiliac pathologies. In this case, the earlier diagnosis of Perthes' disease, with appropriate monitoring and treatment may have arrested femoral head collapse, extrusion and deformity.

Perthes' disease is an idiopathic avascular necrosis of femoral capital epiphysis in an otherwise normal child. Children with congenital abnormalities, as in this case, may be regarded as not conforming to the diagnosis of classical Perthes' disease. The diagnosis should not, however, be refuted because the radiological findings and clinical presentation were typical of Perthes' disease. The abnormality was unilateral. There was no evidence of a metabolic disorder, a connective tissue disorder or a skeletal dysplasia. Furthermore, Hall *et al*¹⁴ reported a high incidence of congenital abnormalities in patients with Perthes' disease. In Perthes' disease, the resultant femoral head shape, proximal femoral growth disturbance and remodelling of acetabulum can create complex deformities.¹⁵ In this reported case the impingement was contributed to by an aspherical femoral head and worsened by iatrogenic anterior acetabular over-coverage from a Salter osteotomy. Myer *et al*¹⁶ reported that anterior femoroacetabular impingement was not uncommon after periacetabular osteotomy and there is no well-established guide as to how to judge the appropriate amount of anterior coverage with Salter's osteotomy. It is suggested that after the acetabular fragment is displaced to the desired position, an anterior T-capsulotomy of the hip joint is required to inspect and palpate for any impingement with the hip flexed and internally rotated. Osteoplasty of the femoral head-neck junction may then be needed to improve femoral anterior offset.¹⁶

Learning points

This case is useful in highlighting that:

- Groin pain in patients with hypoplastic femur warrants thorough evaluation.
- Perthes' disease may occur in patients with PFFD.
- Timely diagnosis of Perthes' disease and containment surgery may prevent long-term morbidity of femoral head deformity.
- Acetabular overcoverage should be avoided in pelvic osteotomy to prevent the development of femoroacetabular impingement.

Competing interests None.

Patient consent Obtained.

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