Unilateral late-onset tibia vara associated with bilateral proximal femoral growth disturbance in monozygotic twins: case report

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Clinical and histopathological similarities and rare association of Blount's disease with various proximal femoral physeal affections (i.e. adolescent coxa vara and slipped capital femoral epiphysis) are well known. Association of tibia vara with another epiphyseal disease of the proximal femur has not been reported previously.

In this paper, a monozygotic set of twins with concordant bilateral epiphyseal growth disturbance of the proximal femur and unilateral late-onset tibia vara is presented. Radiological characteristics of the affected knees revealed a wedging in the proximal tibial epiphysis, depression of the medial joint surface and varus deformity of the tibia. Proximal femurs of both cases showed aspheric congruity, coxa magna, shortness of the femoral neck, and subchondral cystic changes.

The presented cases support the genetic etiology of tibia vara, and association of the two conditions is unique.

Key words: adolescent Blount's disease, tibia vara, proximal femoral epiphysis.

In tibia vara (Blount's disease), enchondral ossification is disturbed in the medial aspect of the proximal tibia (i.e. epiphysis, physis, and metaphysis). Many authors differentiate two main groups of the disease depending upon age at clinical onset, namely, early-onset (or infantile) and late-onset tibia wara[1-4]. Histopathological studies of affected individuals indicate a similar pathologic process in developmental coxa vara and in slipped capital femoral epiphysis (SCFE)[3-5]. Previous manuscripts reported the interesting association of these two proximal epiphyseal affections in the same patient without determining the exact relation of the diseases[6].

A monozygotic set of twins with unilateral late-onset tibia vara associated with proximal femoral growth disturbance is presented in this paper.

Case Reports

The patients were 16-year-old monozygotic twin males. HLA screening indicated they were identical for all locations.

Their chief complaint was progressive bow-leg deformity. At approximately 14 years of age, angulation of their left knees initially appeared. They had mild left knee pain which increased with standing, walking and vigorous activities over the last few months prior to presentation. The patients presented with no other musculoskeletal problems.

The father was heroin addict. The mother had taken some unknown pills to terminate her pregnancy. Prenatal, birth, and early childhood histories were not remarkable. It was reported that both siblings walked when they were 14 months old and did not have any hip pain or difficulties in walking during their childhood.

Review of the systems was normal in both patients. Biochemical and hematological screening revealed no abnormalities. The range of flexion-extension of the hip joints was within normal limits in both patients. There was minimal restriction in rotational movements. No deformities were observed. Minimal waddling was seen when they were waddling.

In the first patient (Patient A), the left knee range of motion was normal. Grade I medial collateral instability was detected when the involved knee was examined in 20° flexion. There were no differences of length in the lower extremities.

In the second patient (Patient B), an apparent lateral thrust of the left knee during ambulation was observed. The left knee demonstrated full extension, with flexion being limited (approximately 20°). When the left knee was examined in 20° flexion, grade II medial collateral instability was detected. An inequality of 1.5 cm in length of the two lower extremities was found (Fig. 1).

Fig. 1. Picture demonstrating clinical features of Patients A and B (at the right side).

On anteroposterior (AP) view of the pelvis (Patient A), growth disturbance at the lateral part of the proximal, shortening of the femoral neck and enlargement of the head with mild increase of the neck-shaft angle was apparent in both hips. There was aspheric congruity and cystic changes at right hip (Fig. 2a). A weight-bearing AP X-ray of the left knee showed a wedging in the proximal tibial epiphysis, depression of the medial joint surface (metaphyseal-diaphyseal angel [MDA] of 25°), and varus angulation of the tibia (Fig. 2b).

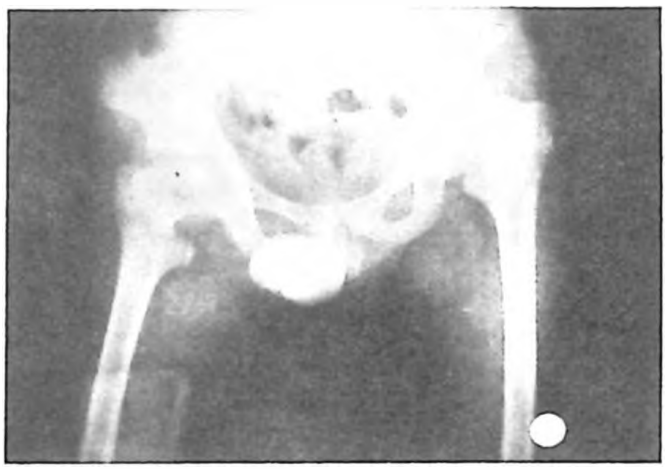

Fig. 2a. Anteroposterior (AP) view of the pelvis (Patient A).

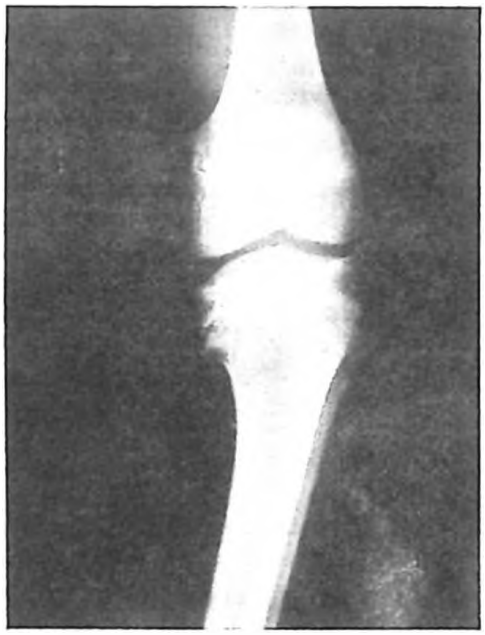

Fig. 2b. Weight-bearing anteroposterior (AP) X-ray of the left knee (Patient A).

Pelvis AP view of Patient B showed bilateral coxa magna with deformation of the femoral heads with short femoral necks, very similar to his brother. On AP weight-bearing X-ray of the left knee, we observed severe wedging of the medial tibial epiphysis and metaphysis, severe depression of joint surface (MDA of 30°), and adaptive overgrowth in the medial femoral condyle (Fig. 3).

The X-ray examinations of the knees and hips of the other members of the family (except one distant relative with tibia vara) disclosed no deformities of the tibia or proximal part of the femur. This observation suggested that this was a new mutation.

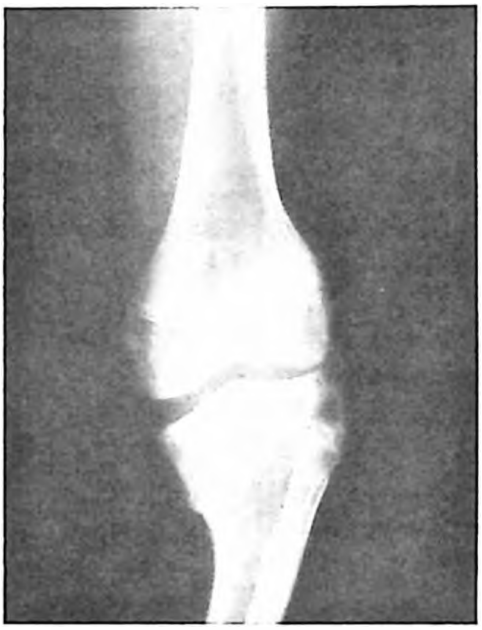

Fig. 3. Anteroposterior (AP) weight-bearing X-ray of the left knee (Patient B).

Discussion

The similarity between the clinical courses of tibia vara and developmental coxa vara was first reported by Lagenskiöld[1,2]. Later research showed that this similarity also existed at a microscopic level[3]. Although Say et al.[7] reported a family with congenital coxa vara and late adaptive degeneration of the proximal tibia, coexistence of adolescent tibia vara and coxa vara has not been previously published. Lovejoy and Lovell[8] described two cases in which adolescent tibia vara presented with SCFE. Wenger et al.[6] also reported one similar case.

Although trauma, infection, and nutritional and genetic factors have been implicated as causative factors, the etiology of tibia vara still remains to be elucidated.

Proximal physis of the femur consists of three parts: longitudinal growth plate, trochanteric growth plate and femoral neck isthmus. Separate or combined growth arrest of these plates eventually leads to different types of deformities. In line with such deformities, the acetabulum shows adaptive changes.

Growth disturbance of the proximal femur may occur due to a multitude of factors such as Legg-Perthes-Calvé disease, trauma, infection, and congenital and developmental conditions. Each condition (except the genetic and insidious developmental diseases) presents its clinical symptoms with varying radiological features in relation to the etiology and the severity of the epiphyseal damage[9]. In the presented cases, broader femoral neck, relative overgrowth of the trochanter major and close-to-normal appearance of the medial side of the neck suggested growth arrest at the lateral side of the longitudinal growth plate.

Certain studies investigating tibia vara draw attention to the high probability of genetic etiology, though currently the mode of inheritance has not been determined[10-13]. Concurrence of the same disease in monozygotic twins with the absence of any defined predisposing factors like marked obesity, black race, or early walking supports the genetic etiology. In this report the association of the two distinct conditions in twins also supports the genetic etiology and genetic relation of these conditions. While there is no clear evidence for the common etiology of these two conditions, it seems that there is a variable coexistence for the affections of the proximal epiphysis of the femur and tibia.

Tibia vara coexisting with SCFE has been reported previously in three cases. This is the first report mentioning coexistence of a different epiphyseal growth problem of the proximal femur. We suggest that the proximal femur should be examined for possible epiphyseal problems in patients presenting with adolescent tibia vara.

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