TIBIA VARA CAUSED BY FOCAL FIBROCARTILAGINOUS DYSPLASIA

THREE CASE REPORTS

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We present three cases of a previously undescribed condition characterised by unilateral tibia vara associated with an area of focal fibrocartilaginous dysplasia in the medial aspect of the proximal tibia. The three children affected were aged 9, 15 and 27 months respectively. Two required tibial osteotomy, but in one the deformity resolved without treatment. The pathogenesis of the focal lesion remains conjectural; the most likely explanation is that the mesenchymal anlage of the tibial metaphysis has, for unknown reasons, developed abnormally at the insertion of the pes anserinus.

Gross unilateral bowing of the tibia is rare in early childhood. The common postural bowing of the toddler is sometimes unilateral but the varus deformity is never as severe as in the three children discussed here, nor is local bony pathology a feature (Levine and Drennan 1982). Unilateral tibia vara may be due to Blount’s disease (Kessel 1970), dyschondrosteosis (Dawe, Wynne-Davies and Fulford 1982), fibrous dysplasia, Ollier’s disease, neurofibromatosis or trauma. The three children presented here showed none of the features of any of these conditions.

CLINICAL FEATURES

Three white children (two boys and one girl) presented with unilateral tibia vara. There was slight shortening of the affected limb (1 cm and 0.5 cm) in two of the children, but no abnormality in any other limb and there was no skin blemish overlying the proximal tibia. The children were healthy and each had a normal antenatal and postnatal history. No member of any of their families had suffered from a similar condition.

Radiographs of all three children showed a lesion in the proximal tibia on its medial aspect. In each case the lesion was at the same distance from the growth plate. The varus angulation was at the site of this lesion which consisted of a punched-out area of the cortex with some surrounding sclerosis extending distally.

Biopsies were taken of two of the lesions and both consisted of dense hypocellular tissue resembling fibrocartilage in some areas and tendon in others; we have called the lesion focal fibrocartilaginous dysplasia.

CASE REPORTS

Case 1. A boy aged nine months presented because his parents had noticed that his right leg was bowed. The deformity had increased since it was first observed at the age of three months. Radiographs were taken (Fig. 1) and the condition was observed. When he was seen six months and 12 months later (Fig. 2) the deformity was clinically and radiologically more severe.

On examination at the age of two years his right leg had 20° of varus deformity and 30° of medial tibial torsion; it was 1 cm short (Fig. 3). Osteotomies of the proximal tibia and distal fibula were performed; these corrected both the varus and the torsional deformity. At operation the lesion was exposed and found to be a plug of white shiny tissue of cartilaginous appearance, into which the pes anserinus was inserted. There was a plane of cleavage between the lesion and the normal bone. The entire lesion was shelled out.

The osteotomies united in six weeks; the appearance and function of the leg were normal and were still normal four and a half years after operation (Fig. 4). At this time there was no longer any discrepancy in leg length.

Case 2. A boy aged 15 months presented because he had recently been noticed to have unilateral tibia vara. Radiographs disclosed the characteristic lesion in the right leg (Fig. 5). Six months later the deformity had increased both clinically (Fig. 6) and radiologically (Fig. 7). In addition to the varus deformity there was 30° of medial tibial torsion.

At the age of two years osteotomies of the proximal tibia and distal fibula were performed to correct both elements of the deformity. A white cartilaginous lesion with

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a clearly defined edge was seen deep to the pes anserinus (Fig. 8). The entire lesion was removed. Fourteen months later the limb looked normal (Fig. 9).

Case 3. This girl presented at the age of two years and three months. When she was aged 12 months it had been noticed that her right leg was bowed; the deformity initially worsened but recently had improved. On examination the right leg was varus and was 0.5 cm shorter than the left. Radiographs showed a lesion in the medial cortex of the proximal tibia with scalloping and sclerosis extending down the medial cortex, and with marked varus angulation (Fig. 10). When reviewed one year later her leg looked normal and radiographs showed less tibia vara with less scalloping but with persistent sclerosis (Fig. 11). At the age of seven her legs were clinically and radiologically normal (Fig. 12).

PATHOLOGY

Case 1. Sections of the resected tissue showed a mass of sparsely cellular collagenous tissue with inactive fibrocytes. In places the cells in the tissues were plump, lay in lacunae and resembled chondrocytes. The ground substance stained patchily for acid mucopolysaccharide especially adjacent to the lacunae. There was no elastin. At the margins this fibrocartilaginous tissue merged with fibrous tissue of nondescript appearance. There were no giant cells, osteoid or bone within the lesion (Fig. 13).

Case 2. Sections showed a circumscribed and sharply demarcated rounded mass of dense and relatively acellular collagen. Near the centre the constituent cells lay in lacunae, an appearance suggesting fibrocartilage, but peripherally the appearance more closely resembled tendon. Surrounding this structure, and sharply demarcated from it, was dense fibrous tissue of nondescript appearance. There were no giant cells, osteoid or bone within the lesion (Figs 14 and 15).

DISCUSSION

The three cases presented here all had severe unilateral tibia vara. The parents of one child (Case 1) had noticed the deformity when the child was aged three months; in the other two children it was noticed at a slightly older age, but in both this was under one year. Radiographs showed a punched-out circumscribed lesion at the site of insertion of the pes anserinus.

Microscopically, the two lesions which were biopsied looked similar. They were composed of dense fibrous tissue, reminiscent of tendon in its structure and arrangement, with foci in which the fibroblasts lay in lacunae, producing an appearance that resembled fibrocartilage. This histological appearance is very unusual. It does not resemble a metaphyseal fibrous defect, a lesion with which it could be confused radiologically. Although close to the epiphysis, the lesion was obviously not epiphysial since it was not composed of hyaline cartilage; in fact there was no hyaline cartilage within the lesion at all. For this reason periosteal chondroma, which has a similar radiological appearance (Rockwell, Salter and Enneking 1972), was excluded.

The intimate relationship of the lesions to the insertion
Case 2. Figure 5—Radiographic appearance at presentation, when aged 15 months. Figure 6—Clinical appearance at the age of 21 months. Figure 7—Radiographic appearance at the age of 21 months. The tibia vara has increased and the metaphysial lesion has enlarged. Figure 8—Operation photograph. The distal end of the incision lies to the left of the figure and the anterior aspect of the limb is at the top. The pes anserinus has been split and reflected anteriorly and posteriorly to disclose the shining white lesion with a curved distal edge which occupies the central one-third of the photograph. Figure 9—Radiographic appearance 14 months after operation. There is no recurrence of either the tibia vara or of the metaphysial lesion.
Fig. 10
Case 3. Figure 10—Radiographic appearance at presentation, when aged two years and three months. Figure 11—One year later, the metaphyseal lesion is smaller and the tibia vara has decreased. Figure 12—At the age of seven both tibiae are normal.

Fig. 13
Figures 13 to 15. Histological appearance of the lesions from Cases 1 and 2. Case 1. Figure 13.—The mass of abnormal tissue is composed of dense fibrous tissue in which cells lie in lacunae, an appearance resembling fibrocartilage (haematoxylin and eosin, ×320). Case 2. Figure 14—A photograph of the plug of abnormal tissue. On the right side of the picture the cells lie in lacunae and the tissue has a fibrocartilaginous appearance; on the left side, wavy rows of dense collagen separated by fiброcytes resemble tendon (haematoxylin and eosin, ×70). Figure 15—Detail from Figure 14 of the fibrocartilaginous tissue (haematoxylin and eosin, ×140).
of the pes anserinus was striking. Similar tissue is found normally at the site of insertion of tendons into cortical bone (Cooper and Misol 1970); a narrow zone of tissue occurs between tendon and bone that closely resembles fibrocartilage and is almost identical, except in amount, to the material seen in the lesions we examined. It therefore seems possible that in these children abnormal development of fibrocartilage has occurred at the site of insertion of the pes anserinus.

The reason for this maldevelopment is unknown but in view of the early age of onset in Case 1 it is presumably a developmental anomaly beginning in utero. It seems possible that the mesenchymal anlage in the area of the pes anserinus fails to differentiate normally and forms too much fibrocartilage; this in turn would interfere with growth on the medial aspect of the proximal tibia to produce the varus deformity. We have no histological proof that the lesion in Case 3 was identical to those in Cases 1 and 2 but this seems likely since the clinical and radiological features were identical. As this third lesion resolved spontaneously, presumably the abnormal tissue had been resorbed or it had differentiated into normal bone.

Surgical correction was carried out in Cases 1 and 2 because the deformity was increasing and was unacceptable to the parents; the fact that spontaneous resolution occurred in Case 3 raises the question as to whether operation is necessary and should encourage the clinician to adopt a conservative approach unless the appearance of the limb is too bizarre to be acceptable.

REFERENCES


