TIBIA VARA DUE TO FOCAL FIBROCARILAGINOUS DYSPLASIA

THE NATURAL HISTORY

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We report five patients with tibia vara due to focal fibrocartilaginous dysplasia of the medial aspect of the proximal tibia. In three patients spontaneous correction occurred, while in one of the remaining two treated by operation, a valgus deformity and neurological complications resulted. Conservative management is therefore recommended.

In 1985 Bell et al. reported three patients with tibia vara in association with a characteristic radiological appearance of the medial aspect of the proximal tibia; two of their patients were treated by operation, while spontaneous resolution had occurred by the age of seven years in the third. From the histology specimens of the lesions in these three patients they derived a name for the condition: focal fibrocartilaginous dysplasia.

We report another five patients, two of whom were treated surgically and three in whom spontaneous resolution occurred. From these patients the aetiology may be postulated.

CASE REPORTS

Our five patients were all referred either to The Hospital for Sick Children, Great Ormond Street or to Queen Mary's Hospital for Children, Carshalton, between 1974 and 1985. The details of our patients and the three originally described by Bell et al. (1985) are summarised in Table I.

All the patients presented with unilateral tibial bowing (Figs 1 and 2). The age at presentation varied from nine to 28 months, and distribution of the condition by sex or side seemed random. Radiographs revealed a

cortical defect in the metaphyseal region of the medial tibia with an area of surrounding sclerosis (Fig. 3), a characteristic appearance.

Two of our patients underwent corrective surgery and biopsy was performed on one of them. Sections showed a mass of fibrocartilaginous tissue merging with dense fibrous tissue at the margins. Near the centre, the cells lay in lacunae in a manner suggesting fibrocartilage, while peripherally the appearance was more suggestive of tendinous tissue. There were no giant cells and no evidence of osteoid tissue or bone within the lesion

Fig. 1

Fig. 2

Case 2. Figure 1 – The patient at presentation, aged 24 months. Figure 2 – Resolution of the deformity.

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Table I. Details of the patients with tibia vara in association with fibrocartilaginous dysplasia

Case	Sex	Side	Age at presentation (months)	Management	Comments
1	F	L	15	Tibial osteotomy and biopsy at 28 months	
2	M	R	24	Conservative	Resolution almost complete by 4 years
3	M	R	17	Biopsy aged 28 months but no corrective surgery	Resolution almost complete by 4 years 7 months
4	F	L	28	Conservative	Resolving (now aged 3 years 7 months)
5	F	R	16	Tibial and fibular osteotomies at 25 months	Surgery followed by peroneal nerve palsy; at age 14 years 7 months persistent valgus deformity
<i>Cases reported by Bell et al. 1985</i>					
6	M	R	9	Tibial and fibular osteotomies at 24 months	Deformity first noted at 3 months
7	M	R	15	Tibial and fibular osteotomies at 24 months	
8	F	R	27	Conservative	Deformity first noted at 1 year; resolution by 7 years

Fig. 3

Radiograph of Case 1 at presentation, aged 15 months.

Fig. 4

Photomicrograph showing dense fibrous tissue. On the left, cells lie in lacunae in a manner resembling fibrocartilage. This merges with cellular collagenous tissue on the right which resembles tendon (H&E $\times 125$).

(Fig. 4). The histological appearance in this case, and in that of a third patient who underwent biopsy only, was identical with that described by Bell et al. (1985).

In three of our patients (Cases 2, 3 and 4) there was improvement by the age of three years without surgical correction.

DISCUSSION

An analysis of the details of our patients and those reported by Bell et al. (1985) allows us to suggest the

probable natural history of this condition. In both series, the children's parents had noted the deformity in the first 18 months, although presentation at hospital was later than this for three patients. Initial deterioration had sometimes been observed. In this context it is noteworthy that patients treated by operation presented younger and it was the initial deterioration which prompted operation.

Spontaneous improvement in the bowing and the radiological lesion was observed in four patients. In these

Fig. 5

Fig. 6

Case 4. Figure 5 – Radiograph of the patient at presentation, aged 28 months; the mother reported that resolution was already occurring. Figure 6 – The same patient aged 3 years 7 months: spontaneous resolution is almost complete.

patients the resolution started at about the age of two years and almost complete correction had occurred in three patients by the age of four years. The only patient in whom follow-up has been sufficient to show total spontaneous correction is Case 8.

From these patients it is impossible to state the incidence of this condition as both The Hospital for Sick Children and Queen Mary's Hospital for Children are tertiary referral centres; however, the fact that only five patients have been seen between 1974 and 1985 indicates its rarity.

We believe that resolution would have occurred in all the patients had they been given the opportunity. Operation is to be avoided as it is unnecessary and has attendant hazards, as illustrated by Case 5 who, after operation, had a valgus deformity (Fig. 7) as well as neurological problems.

This variety of tibia vara should be added to the list of skeletal deformities in children in which spontaneous correction can be anticipated (Walker 1972).

Fig. 7

Case 5. A photograph taken when the patient was aged 14 years 7 months: there is persistent valgus deformity and a mild peroneal nerve palsy.

REFERENCES

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