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Orthopaedic Clinic, Medical University of Pécs, Hungary

DYSPLASIA EPIPHYSEALIS HEMIMELICA

OTTO BARTA, ANTAL SCHANZL & JÁNOS SZEPESI

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Dysplasia epiphysealis hemimelica (D.E.H.) is a rarely observed disorder of ossification. It occurs mostly in the distal femoral and distal tibial epiphyses. It is rarely seen in the upper extremities. The chief characteristic of D.E.H. is appearance of isolated, irregular ossification centres on one side of the epiphysis. Ossification of these centres to each other and to the main ossification centre results in asymmetric enlargement of the epiphysis. This disorder of ossification is usually limited to one side of a single limb (Young 1960); the medial side is affected twice as often as the lateral.

The lesion was first observed in the talus and was named "Tarsomegalie" by Mouchet & Belot (1926). Fairbank (1956) separated this disorder from dysplasia epiphysealis multiplex and dysplasia epiphysealis punctata and recommended calling it D.E.H. A variety of names has been attached to this disorder, including: "Tarso-epiphysealis aclasis" (Trevor 1950), "Chondrodystrophie epiphysaire" (Ingelrans et al. 1953), "Epiphysealis hyperplasia" (Aegerter et al. 1968), and "Epiphyseal osteochondroma" (Goldenberg 1966).

The etiology is unknown and genetic factors probably play no role. One of identical twins suffered from this disease but the other one was healthy in Donaldson's case (Donaldson et al. 1953). Developmental abnormality is considered responsible for the disorder (Mouchet & Belot 1926), whereas according to Wheble & Connell (1958) the cause is probably viral infection (rubella, measles). In Fairbank's opinion (1956) the causative agent has an effect on the apical ectoderm of the limb bud for a very short time in the fifth week of foetal development. This would explain the monomelic appearance of the disease.

Neither the etiology nor the pathomechanism has been clarified. It is possible that a proliferative chondric centre is the starting point of the disturbance. The cartilage cells placed perichondrially lose their polarity at the top of the mitotic annulus, and the cells wandering in the

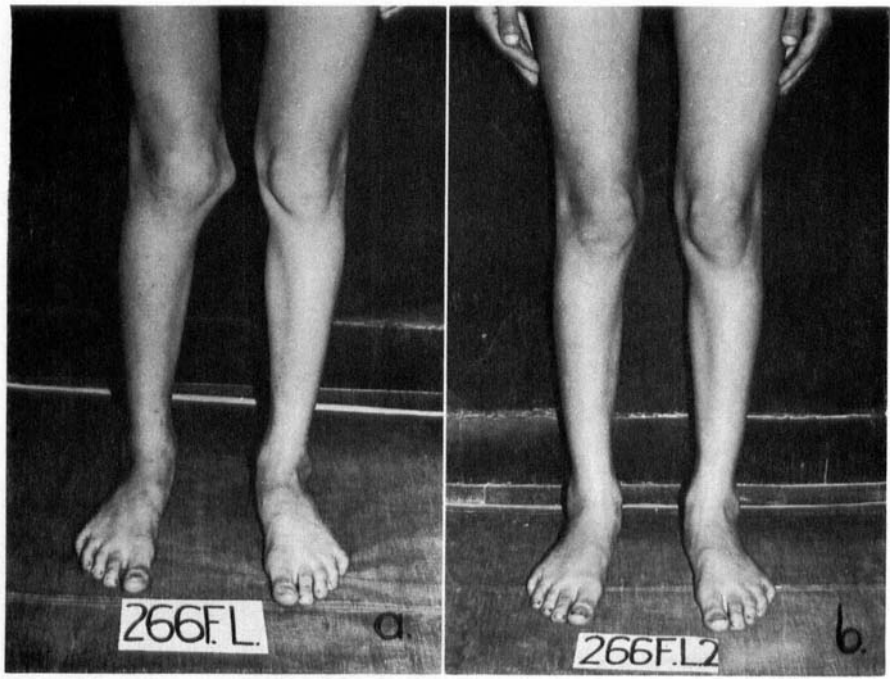

Figure 1. Boy aged 12 years. (a) Swelling at the inner side of the right knee (preoperative picture). (b) Picture after operation.

direction of the epiphysis produce an ectopic centre. Division of cells continues here. Later they degenerate and become calcified, causing an atypical ossification centre inside the epiphysis. A number of ossification centres arise if cells placed peripherically among the concentrically localised cells around the epiphyseal ossification centre preserve their capacity to divide and ossify. The ossification centres develop in different directions if the cells also lose their polarity (Trevor 1950).

Local hyperaemy caused by disturbance in innervation may explain the illness of one side of epiphysis (D'Angio et al. 1955).

Clinical signs

This disorder is usually recognised during the growth period. In the majority of published cases D.E.H. has been most common between two and fourteen years of age.

The chief clinical sign is the bony hard swelling on the extremities (usually on the inner or outer aspect of the knee or ankle). Regional muscular atrophy may occur as well as restriction of motion at the

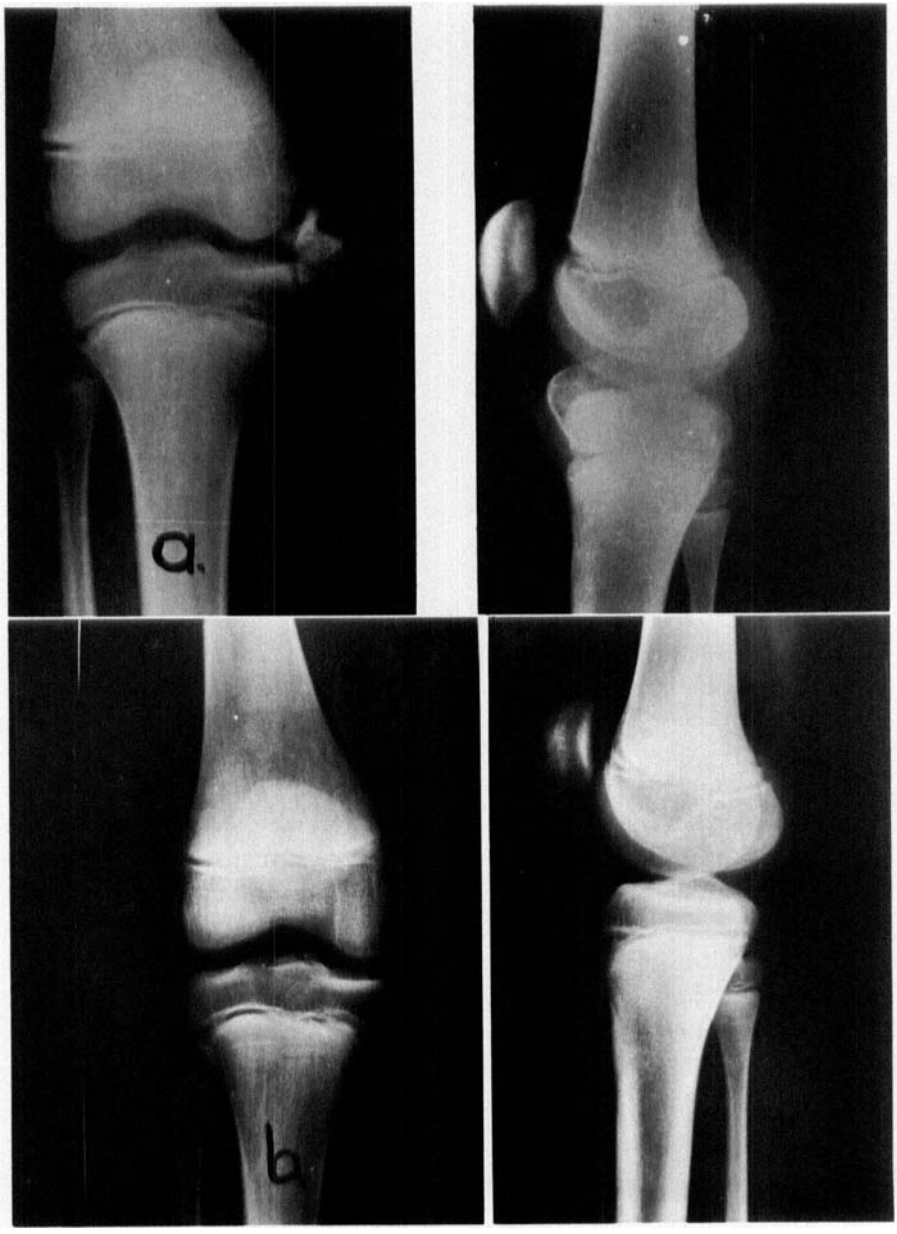

Figure 2. X-ray pictures of the right knee. (a) X-ray picture before operation. (b) X-ray picture after operation.

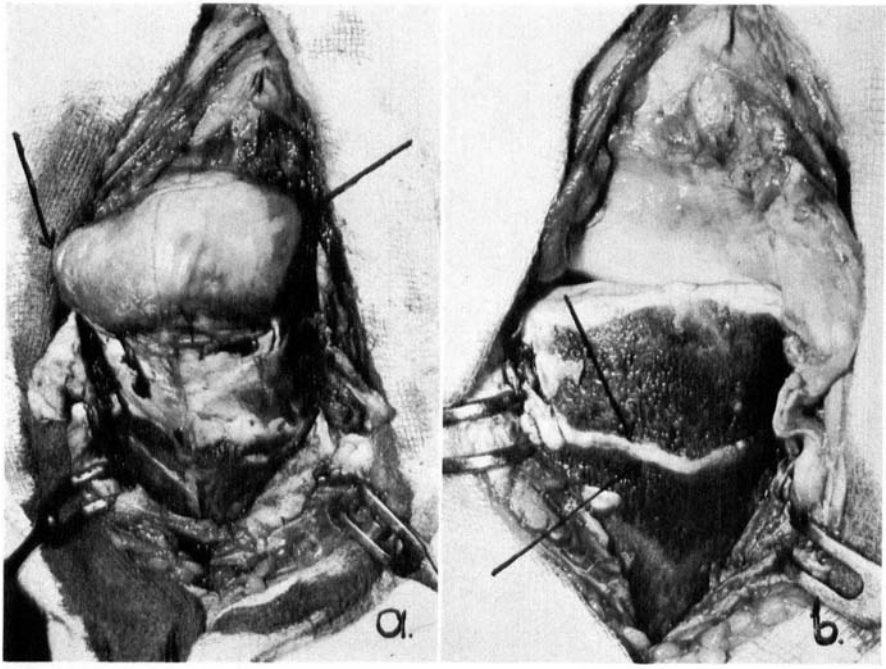

Figure 3. Exposure at exploration. (a) Arrow shows the bony lesion. (b) The growth cartilage of the tibia can be seen after excision.

affected joint. The latter is observed if the epiphyseal prominence impinges on the opposing articular surface. Because of this varus or valgus deformity may be present. There is usually no pain and inequality of limb length. Macroscopically the lesion is found to be covered by smooth cartilage, as it is seen in osteocartilaginous exostosis. There is no sharp borderline between the swelling and the epiphysis.

CASE HISTORY

F.L. This 12-year-old boy was admitted with a complaint of slowly growing swelling on his right knee of 3–4 years' duration. On physical examination a nut-sized, bony hard, painless swelling was palpable on the inner aspect of the right knee. Motion was free at the knee joint. The laboratory findings were normal. The X-ray picture showed supernumerary growth cartilage on the inner aspect of the epiphysis of the right tibia. It was seen some mm proximally from the growth cartilage. A sharply outlined accessory epiphysis (32 × 35 mm in size) could be seen in part between the surfaces of the joint (Figure 2 a).

At exploration we observed that the smooth, cartilage-covered nut-sized swelling was attached ventrally to the growth cartilage of the tibia by a cartilagic bridge

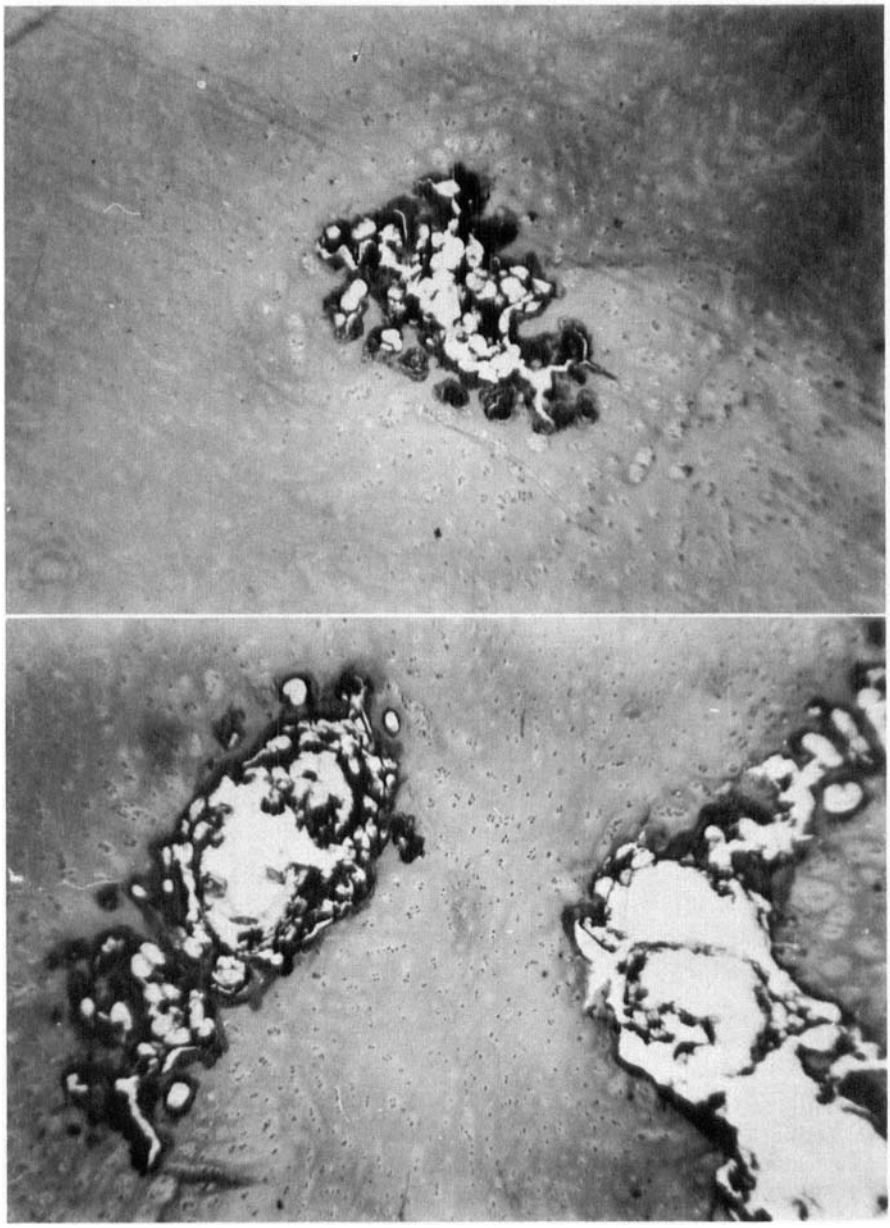

Figure 4. Ossification centres can be observed in the accessory epiphysis.

Figure 5. On one side of the supernumerary growth cartilage (localised between the true epiphysis and the accessory one) the cartilage cells produce lines.

(Figure 3 a). As the accessory epiphysis and the cartilagic bridge were excised (Figure 3 b), the ligamentum mediale became loose, so we had to strengthen it with the ligament of musculus semitendinosus. At follow-up a year later the patient was free of complaints. The X-ray picture showed the place of resection (Figure 2 b).

Microscopically several ossification centres of different sizes could be observed in the accessory epiphysis. These centres were fused with each other and in part with the main ossification centre (Figure 4). An epiphysis plate was found between the epiphysis of the tibia and the accessory epiphysis. It could clearly be observed that cartilage cells lined up, as is characteristic of the growth cartilage. Calcified areas could be noticed in primary trabeculae (Figure 5). Hypertrophied cartilage cells were found in the cartilage of accessory epiphysis: somewhere in a group, elsewhere irregularly (Figure 6). The findings are similar to those of osteocartilaginous exostosis.

DISCUSSION

About 60 cases of D.E.H. have been reported in the literature. The special characteristic of the case reported here is an epiphyseal plate between the proximal epiphysis of the tibia and the accessory epiphysis. It is probable that the polarity of cells of the ectopic, prolifera-

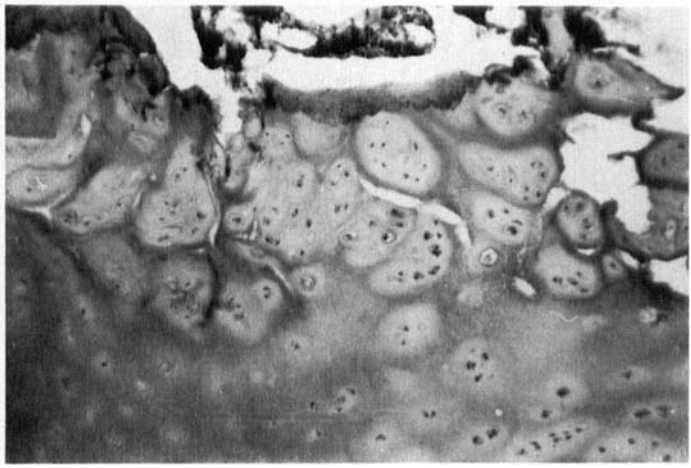

Figure 6. Hypertrophic cartilage cells can be seen in the cartilage cap of the accessory epiphysis.

tive cartilage has turned 180° in the vicinity of this area. This has resulted in a supernumerary epiphyseal plate, because the cells have had different divisional and degenerative tendencies on the opposite sides of the accessory epiphyseal plate.

The bony lesion has grown medially rather than proximally. This explains why neither valgus deformity nor restriction of motion has occurred at the knee joint. The purpose of surgical procedure in this case was to prevent deformity of the joint, as well as consecutive arthrosis.

SUMMARY

A successfully operated case of D.E.H. (localised to the medial condylus of the right tibia) is reported and the literature is reviewed. Etiologic and pathogenetic factors and some hypotheses about the pathomechanism are considered. The characteristic of the author's case is the presence of a nearly normal growth cartilage between the epiphysis and the accessory epiphysis.

REFERENCES

Aegerter, E. & Kirkpatrick, J. A. (1968) *Orthopaedic diseases*, pp. 99, 103. 3rd ed. W. B. Saunders, Philadelphia.

D'Angio, G. J. D., Ritvo, M. & Ulin, R. (1955) Clinical and roentgen manifestations of tarso-epiphyseal aclasis. *Amer. J. Roentgenol.* **74**, 1068.

Donaldson, J. S., Sankey, H. H., Girdany, B. R. & Donaldson, W. F. (1953) Osteochondroma of the distal femoral epiphysis. *J. Pediat.* **43**, 212.

Fairbank, T. J. (1956) Dysplasia epiphysealis hemimelica (tarso-epiphyseal aclasis). *J. Bone Jt Surg.* **38**-B, 237.

Goldenberg, R. R. (1966) Remarks made to Kettelkamp's article: Dysplasia epiphysealis hemimelica. *J. Bone Jt Surg.* **38**-B, 765.

Ingelrans, P. & Lancheretz, M. (1953) A propos d'un cas de chondrodystrophie epiphysaire. *Rev. Chir. orthop.* **59**, 242.

Kettelkamp, D. B., Campbell, C. J. & Bonfiglio, M. (1966) Dysplasia epiphysealis hemimelica. *J. Bone Jt Surg.* **48**-A, 746.

Mouchet, A. & Belot, J. (1926) La tarsomegalie. *J. Radiol. Electrol.* **10**, 289.

Rechnagel, K. (1960) Dysplasia epiphysealis hemimelica. *Acta orthop. scand.* **29**, 237.

Rubin, Ph. (1964) *Dynamic classification of bone dysplasias*, p. 159. Year Book Medical Publishers Inc., Chicago.

Saxton, H. M. & Wilkinson, J. A. (1964) Hemimelic skeletal dysplasia. *J. Bone Jt Surg.* **46**-B, 608.

Shanks, S. C. & Kerley, P. (1971) Dysplasia epiphysealis hemimelica. *A text-book of X-ray diagnosis*, Vol. VI, p. 613. H. K. Lewis & Co., Ltd., London.

Theodorou, S. & Lanitis, G. (1968) Dysplasia epiphysealis hemimelica (epiphyseal osteochondroma). *Helv. paediat. Acta* **23/2**, 195.

Trevor, D. (1950) Tarso-epiphyseal aclasis. *J. Bone Jt Surg.* **32**-B, 205.

Wheble, V. H. & Connell, M. C. (1958) Dysplasia epiphysealis hemimelica. *Brit. J. Radiol.* **31**, 637.

Young, H. H. (1960–1961) Dysplasia epiphysealis hemimelica. *The year book of orthopedics and traumatic surgery*, p. 41. Year Book Medical Publishers, Chicago.

Correspondence to:

Prof. dr. Otto Barta
Orthop. Clinic of Univ. Pécs
H–7643 Pécs, Ifjusag u. 31. Hungary