Unusual presentation of more common disease/injury

Acutely presenting kissing lesions of the ankle: an atypical Trevor's disease and literature review of other unusual presentations of the disease

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Summary

Trevor's disease, an intra-articular variant of osteochondroma, has been reported previously in various unusual forms. The present article describes acutely presenting, intra-articular kissing lesions in a 4-year-old girl. A high index of suspicion and elaborate awareness on the part of the treating surgeon are essential to diagnose as well as manage such cases.

BACKGROUND

Osteochondromas are common benign lesions of the skeleton characterised by typical cartilage-crested bony excrescences that originate from the physal plate, allegedly arising as an aftermath of localised or generalised osseous maldevelopment.¹ The literature reveals diverse variants of these anomalous bony protuberances. Trevor's disease or dysphasia epiphysialis hemimelica or Fairbank's disease has been a prominent structurally and histologically cognate lesion thus far described. Originally described in 1926 by Mouchet and Berlot,² 'tarsomegalie' (as named by the discoverers) has time and again been reported by orthopaedicians worldwide, with different typical and atypical presentations.³⁻⁵ The present article describes one such patient with a typically described classic Trevor's disease with an unusual presentation. The peculiarity in the clinical presentation, the diagnostic dilemma faced by the treating surgeons, management controversies and a review of other atypical presentations of the condition in the literature have been expatiated upon.

CASE PRESENTATION

A 4-year-old girl presented to our paediatric orthopaedic emergency services with a 2-day history of acute onset, severe pain over the right ankle joint accompanied by significantly erythema and swelling. The swelling and pain had completely restricted all the activities of the child and caused her to cry incessantly for a few hours, which had prompted the parents to bring the girl to our hospital. The parents were unsure of any preceding traumatic episode or febrile illness. The child was allegedly playful and healthy prior to the onset of these symptoms. The ankle joint was extremely tender, especially over the anteromedial joint line, and all movements of the child were restricted. The main differential diagnoses that were considered at this stage included infective arthritis, fractures or soft tissue injuries of the ankle joint.

INVESTIGATIONS

The child immediately underwent investigations, including complete blood counts, erythrocyte sedimentation

rate and roentgenograms of the involved ankle joints. The radiographs demonstrated a calcified area over the antero-medial aspect of the medial malleolus and another such lesion over the anterior aspect of the talus that resembled 'kissing lesions' representing complementary protuberances arising from the adjacent bones. The CT picture showed a much clearer picture demonstrating intra-articular, pedunculated osseous lesions arising from the two bones, with a discontinuity observed in the proximal bump arising from the distal tibia. This partial break in the mass arising from the tibia, which possibly resulted from some injury, was responsible for the acute painful presentation in our patient. The MRI showed a cartilage cap that is typical of osteochondromatous outgrowth. The osseous discontinuity was well evident on the MRI with a high signal intensity corresponding to the oedematous region. A skeletal survey failed to show similar lesions elsewhere in the body. Retrospectively, when enquired, the mother felt that the child probably had been developing some fullness over the anteromedial part of the ankle in the last few months. No other similar illness had been identified in any of the patient's relatives so far. The child was sedated, limb splinted, medicated adequately with antioedema and analgesic drugs and planned for excisional biopsy of the lump (see figures 1 and 2).

TREATMENT

Under general anaesthesia, the child's ankle was exposed using the anterior approach. The approach was developed between the anterior tibial and extensor hallucis longus tendons: with the neurovascular bundle, retracted laterally with the long extensor tendons of the toes, and the anterior tibial tendon, retracted medially. Intraoperatively, a large osteocartilaginous mass consistent with our pre-operative diagnosis was found to be attached below the medial malleolus. A similar mass was present on the dorsal surface of the talus. Both were excised and sent for histopathological examination. Histologically, the tissue consisted of normal bone and hyaline cartilage. The basic

Figure 1 Clinical photograph.

pathological tissue appeared to be abnormal cartilage proliferation associated with enchondral ossification. The diagnosis thus conclusively arrived at was intra-articular osteocartilaginous growth consistent with dysplasia epiphysealis hemimelica (DEH) (see figures 3–6).

Figure 2 Radiography showing the mass.

Figure 3 Intraoperative picture showing lesions arising from the medial malleolus.

Figure 4 Lesions arising from the talus.

Figure 5 Picture following excision of lesions.

DISCUSSION

DEH is an uncommon epiphyseal growth abnormality that is encountered as rarely as 1 in 1 000 000 among the paediatric population.³ Around 24 years after the initial description, Trevor *et al* described 10 patients with this disorder and replaced the initially proposed term 'tarsomegalie' by 'tarso-epiphyseal aclasis'.⁶ In 1956, Fairbank *et al* reported 14 more cases and renamed it 'dysplasia epiphysealis hemimelica',⁷ a term that is still unanimously used to define this anomaly.

One needs to understand the embryological miscreation that has been proposed currently to explicate the aetiological background of the condition, although the picture

is hardly limpid hitherto.³ Trevor hypothesised a congenital error or insult perpetrating the defect in the lower limb buds during early fetal life. He also speculated that the peculiarity in vascular arrangement of the epiphysis was responsible for the tendency of this lesion to be asymmetrically confined to only one side of the joint, preferentially the medial side.⁶ Bhosale *et al*,⁸ in 1933, conjectured that the cartilage of an epiphysis of a long bone is formed from a narrow zone 'mitotic annulus', where cartilage cells are actively dividing. These cells in the mitotic annulus eventually disintegrate, and together with the matrix they give rise to the main constituent of synovial fluid. He theorised that in DEH, the process is altered such that the cells do not become senile and flat but retain their power of division, so that a mass of cartilage is produced. Fairbank,⁷ in his article, had surmised that the lesion was a true dysplasia or a faulty growth of the epiphysis. No hereditary or genetic aetiological background has been established heretofore and the commonly involved joints are the ankle and knee.⁹

The lesions typically involve children and young adults, with a relatively higher predilection for male predominance.⁹ Azouz *et al*¹⁰ had classified the disease based on the extent of involvement as: A. localised form, B. classic form (more than one area of osseous involvement in a single extremity) and C. generalised or severe form. The classic form is the most commonly reported variety, accounting for more than two-thirds of cases. Our patient also presented with a classic type of DEH, affecting the distal tibial and talar epiphyses.

The peculiarity in our patient that we wish to foreground is the acute presentation in a 4-year-old following fracture of one of the 'kissing' lesions. In such young children, the ability to extract an adequate history or conduct appropriate clinical examination is hugely curtailed, which imparts greater ambiguity to the existing diagnostic dilemma at such times. We had considered two major differential diagnoses in our patient—infected or traumatic, considering the mode of presentation. The availability of radiological investigative modalities could, however, help us approach the diagnosis, which was confirmatively established following histopathological examination. Considering the rarity of the disease (with only three reports from the subcontinent so far), it definitely poses a significant difficulty and discombobulation to the treating clinician.

Management of these lesions has been discussed elaborately in the literature and is considered appropriate to tailor the management protocol according to the morphology of the lesion, patient profile and symptoms.^{11–13} In asymptomatic, intra-articular lesions in immature patients, an observant, waitful approach is advocated unanimously. Early surgeries in these patients may result in secondary osteoarthritis and, therefore, best avoided. Some surgeons have advocated early surgical intervention in the asymptomatic, extra-/juxta-articular lesions (which are less prone to postsurgical complications), although present literature favours a conservative approach with supervised neglect until skeletal maturity in all lesions unless symptomatic (malignant transformation of these lesions has never been reported).

These lesions may present as innocuous, asymptomatic swellings or with pain, movement restriction or

Figure 6 Excised osteochondromatous lesions.

Table 1 Atypical presentations of Trevor's disease

Sl. no.	Article	Authors, year	Journal	Comments
1.	Dysplasia epiphysealis hemimelica of the patella	A. Sadykhov AG <i>et al</i> (1962) B. Louis <i>et al</i> (1968) C. Enriquez J <i>et al</i> (1981) D. Araujo CR <i>et al</i> (2006)	A. <i>Ortop Travmatol Protez</i> B. <i>Bull Soc Med Afr Noire</i> C. <i>Clin Orthop Relat Res</i> D. <i>RadioGraphics</i>	
2.	Epiphyseal osteochondroma in an adult	Shinozaki T (1999)	<i>Clin Imaging</i>	
3.	Upper extremity involvement	A. Heiple KG (1961) B. Lamesch AJ <i>et al</i> (1983) C. Azouz <i>et al</i> (1984) D. Maylack FH <i>et al</i> (1988) E. Rao SB <i>et al</i> (1994) F. Poli GG <i>et al</i> (1995) G. Levi N <i>et al</i> (1998) H. Taniguchi Y <i>et al</i> (1998) I. Vanhoenacker F (1999) J. Takeuchi H (2003)	A. <i>J. Bone Joint Surg</i> B. <i>J. Bone Joint Surg Am</i> C. <i>J Can Assoc Radiol</i> D. <i>J Hand Surg Am.</i> E. <i>Clin Orthop Relat Res</i> F. <i>Chir Organ Mov</i> G. <i>Acta Orthop Belg</i> H. <i>J Hand Surg Br</i> I. <i>Eur Radiol</i> J. <i>J Shoulder Elbow Surg</i>	A. Carpal B. Carpal bones D. Metacarpophalangeal joint E. Upper limb with ass. osteochondromas F. Radius G. Distal radius H. Carpal instability I. Scaphoid J. Coronoid process ulna
4.	Bilateral lesions	A. Maurer HJ <i>et al</i> (1963) B. Gregory PR <i>et al</i> (1993) C. Merzoug V <i>et al</i> (2002)	A. <i>Fortschr Geb Rontgenstr Nuklearmed</i> B. <i>Foot Ankle</i> C. <i>Pediatr Radiol</i>	A. Symmetrical patellar osteochondroma

functional limitation, compressive or mechanical complaints, intra-articular loose body, ankle deformation, peroneospastic flatfoot, limb length discrepancies or rarely in adults with secondary osteoarthritis.^{11–14} Although fractures are known complications of pedunculated osteochondromas, our literature review surprisingly yielded hardly any information on similar acutely presenting cases.

We believe that morphological parameters of these outgrowths like size, presence or absence of a peduncle, intra-articular location (increased mobility around the lesion causing increased chances of breakage), deformity of the adjoining joints, etc, are important risk factors predictive of developing complications. Another significant factor observed in our case was the presence of large, oppositionally located, 'kissing' lesions, which may accentuate the possibility of fractures and other complications. We strongly recommend that these additional factors must be assessed in detail in every patient prior to planning a conservative management, so as to obviate any possible ensuing adversities.

The literature review on all atypical presentations of Trevor's disease has been tabulated (table 1).

Learning points

- The purpose of this article is to report an unusual presentation of intra-articular variant of Osteochondroma (Trevor's disease).
- High index of suspicion is necessary in diagnosis and management of such lesions.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. Kuo RS, Bellemore MC, Monsell FB, *et al*. Dysplasia epiphysealis hemimelica: clinical features and management. *J Pediatr Orthop* 1998;**18**:543–8.
2. Mouchet A, Berlot J. La tarsomegalie. *J Radiol Electrol* 1926;**10**:289–93.
3. Bansal P, Khare R, Lal H, *et al*. Dysplasia epiphysealis hemimelica or Trevor's disease of proximal tibia mimicking loose body. *J Chirur Orthop Traumatol* 2010;**1**:105–6.
4. Azouz EM, Slomic AM, Archambault H. Upper extremity involvement in Trevor disease. *J Can Assoc Radiol* 1984;**35**:209–11.
5. Araujo CR, Montandon S, Montandon C, *et al*. Dysplasia epiphysealis hemimelica of the patella. *RadioGraphics* 2006;**26**:581–6.
6. Trevor D. Tarso-epiphyseal aklasis: a congenital error of epiphyseal development. *J Bone Joint Surg Br* 1950;**32-B**:204–13.
7. Fairbank TJ. Dysplasia epiphysealis hemimelica (tarso-epiphyseal aklasis). *J Bone Joint Surg Br* 1956;**38-B**:237–57.
8. Bhosale SK, Dholakia DB, Sheth BA, *et al*. Dysplasia epiphysealis hemimelica of the talus: two case reports. *J Orthop Surg* 2005;**13**:79–82.
9. Wynne-Davis R, Hall CM, Apley AG. Dysplasia epiphysealis hemimelica. In: Wynne-Davis R, Hall CM, Apley AGeds. *Atlas of skeletal dysplasia*. New York: Churchill Livingstone, 1985:539–43.
10. Azouz RM, Slomic AM, Marton D, *et al*. The variable manifestations of dysplasia epiphysealis hemimelica. *Pediatr Radiol* 1985;**15**:44–9.
11. Azzoni R. Dysplasia epiphysealis hemimelica of the talus. *J Orthopaed Traumatol* 2009;**10**:43–6.
12. Kuo RS, Bellemore MC, Monsell FB, *et al*. Dysplasia epiphysealis hemimelica: clinical features and management. *J Pediatr Orthop* 1998;**18**:543–8.
13. Glick R, Khaldi L, Ptaszynski K, *et al*. Dysplasia epiphysealis hemimelica (Trevor disease): a rare developmental disorder of bone mimicking osteochondroma of long bones. *Hum Pathol* 2007;**38**:1265–72.
14. Arkader A, Friedman JE, Moroz L, *et al*. Acetabular dysplasia with hip subluxation in Trevor's disease of the hip. *Clin Orthop Rel Res* 2006;**457**:247–52.

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