DYSPLASIA EPIPHYSEALIS HEMIMELICA OF THE POSTERIOR MEDIAL ANKLE

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STATEMENT OF PURPOSE:

Dysplasia Epiphysealis Hemimelica (DEH) is a rare skeletal condition that forms osteochondromas in the epiphysis of long bones. This occurs mainly in pediatric patients with a predilection for the lower extremities. The purpose of this study is to review a case of DEH in a pediatric patient involving the lower extremity.

LITERATURE REVIEW:

DEH was first described by Mouchet and Belot in 1926, describing the condition as ‘‘tarsomegaly’’ (1). In 1950, Trevor described 8 cases of “tarsoepiphyseal aclasis” in which he believed was a congenital error of epiphysial development, coining the name “Trevor’s disease” (2). Finally, in 1956 Fairbank named the condition dysplasia epiphysealis hemimelica (3). The incidence of DEH has been estimated at 1 in 1,000,000 individuals in the general population (4). However, others believe this number is much lower due to improper diagnosis (5). DEH is usually diagnosed in children between two and eight years old. DEH has been shown to occur three times as often in males than in females (6), with the medial side of the epiphysis affected twice as often as the lateral side (7).

CASE STUDY:

A nine year old boy with history of asthma presented to clinic with a long history of left ankle pain. Radiographs and an MRI were obtained demonstrating a left intra-articular osteochondroma (Figure 1) consistent with Trevor's disease as well as extensive tenosynovitis. Therefore, recommendation was made for left ankle arthrotomy, tenosynovectomy and resection of the large talus intra-articular osteochondroma.

CASE STUDY CONTINUED:

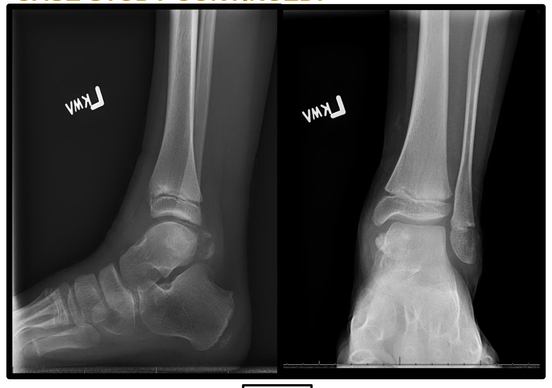

Figure 1

In the operative procedure, an anteromedial ankle incision was made followed by dissection down to the fascia. Upon incising the deep fascia, the medial malleolus was exposed. A 0.5 inch osteotome was then used to perform a medial malleolar osteotomy. The medial malleolus was then inverted inferiorly to expose the ankle joint. The very large posteromedial intra-articular osteochondroma was thus identified. There was extensive ankle synovitis as well as tenosynovitis of the tibialis posterior, flexor digitorum longus, and flexor hallucis tendons on along the posteromedial aspect of the ankle. The inflamed synovium was removed with a scalpel and rongeur. The large osteochondroma was removed in fragments using a series of osteotomes, rongeurs and curettes. The specimen was sent to pathology for evaluation. The ankle was then copiously irrigated. There was no evidence of further intra-articular loose fragments. The ankle joint was found to be stable. The medial malleolus osteotomy was then repaired using two Synthes 4.5 mm cannulated screws. This provided stable fixation (Figure 2A).

CASE STUDY CONTINUED:

Upon stressing the ankle for plantarflexion, dorsiflexion, inversion and eversion, the ankle was found to be stable. There was no restriction to ankle movement. The tendon sheaths, ankle capsule and skin edges were re-approximated in the usual manner. The patient underwent application of a short leg cast.

The patient tolerated the procedure without difficulty and was taken to the recovery room in stable condition.

Following surgery, the patient was placed in a below knee cast for four weeks. At four weeks post operatively, the patient began 6 weeks of physical therapy (12 sessions) which was successfully completed. At three months postoperatively (Figure 2B), the patient was pain-free and returned to normal activity as tolerated, which included organized sports such as baseball. At ten months postoperatively, the patient underwent an elective removal of the hardware that was utilized for fixation of the medial malleolus. At 19 months postoperatively, the patient has continued pain-free normal activities and organized sports with no evidence of growth arrest or recurrence of the lesion.

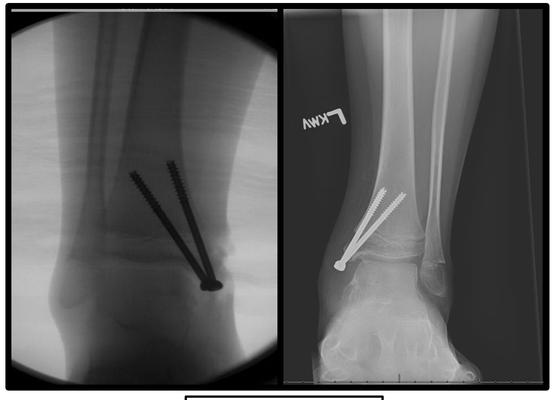

Figure 2A (left) and 2B (right)

DISCUSSION:

DEH usually presents in children as a unilateral mass affecting the medial aspect of the ankle joint. Pain may occur due to irritation of the soft tissue from shoe wear from an underlying mass or due to the osseous mass affecting the joint. Osseous masses affecting the joint can cause not only pain, but also gait abnormalities and limb length discrepancies at which point surgical resection should be performed (8).

CONCLUSION:

DEH is a rare condition that affects mainly pediatric patients with predilection for the lower extremity. A thorough physical exam with proper imaging is key to the diagnosis. Although conservative care may be successful in patients, surgical intervention is often warranted to prevent premature arthrosis of the foot and ankle, limb length discrepancy and/or gait abnormalities.

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