CASE REPORT

Dupuytren’s disease in an infant

A. Bebbington, R. Savage

From Royal Gwent Hospital, Gwent, Wales

Dupuytren’s disease has only been rarely reported in children and is rarer still in infants. We report a case in a six-month-old infant who required surgery when aged ten months. Histology confirmed the diagnosis of Dupuytren’s disease.

Dupuytren’s disease is generally considered to be a disease of adults. It is rarely reported in children. Dupuytren himself reported a case in a six-year-old boy in 1832. The first reported case of histologically-proven Dupuytren’s disease in a child was in a 14-year-old boy who had flexion contractures of the ring and little fingers with a similar affection in the plantar fascia.

In a study of nine cases by Urban et al., eight had histologically-proven Dupuytren’s disease before the age of 13 years, but five of these were from unpublished communications. The youngest published case in this study was nine years of age. A review of the literature found Dupuytren’s in 13 other cases before the age of 13 years, and a further 109 patients who presented with the onset of Dupuytren’s before the age of 20 years, but without histological confirmation. Recently, three further cases of histologically-proven Dupuytren’s have been reported in children aged 2.5, nine and ten years, although later review of the histology of the case of the nine-year old revealed features which were suggestive of epitheloid sarcoma.

Only one case of Dupuytren’s occurring in an infant has been reported. This was in a ten-month-old boy with a flexion deformity of the distal interphalangeal joint. Due to a progressive contracture of the finger, also involving the proximal interphalangeal joint, surgery was performed at 18 months of age and histology of the excised tissue showed features typical of Dupuytren’s disease. We report a case of histologically-proven Dupuytren’s in a six-month-old.

Case report

A six-month-old boy was referred by his general practitioner with a swelling in the left palm and an associated palmar cord to the ring finger. There was no family history of Dupuytren’s disease or epilepsy. Examination revealed a palpable plaque of tissue deep to the skin at the base of the ring finger with a cord extending distally to the radial side of the proximal phalanx. There were two shallow skin pits visible in the palm. The finger extended to the straight position passively but could not be hyperextended. The features were thought to be similar to Dupuytren’s in an adult. The boy was reviewed three months later and the swelling had increased slightly. The parents noticed a slight reluctance to use the hand. The infant underwent an excision biopsy at ten months of age.

A Bruner incision was used in the palm, extending into the ring finger. The mass was excised. It did not involve the digital nerves or the vessels, but appeared to involve the flexor tendon sheath at the A1 pulley and part of the A2, the radial surface of the flexor digitorum profundus tendon and the edge of the third lumbrical. These were excised together with the mass.

Histological examination showed a fibrocellular lesion consisting of thick bundles of fibroblasts separated by collagen fibres and slit-like compressed blood vessels. The lesion had diffusely infiltrated the fibrofatty and muscular tissue. It showed no evidence of encapsulation. There were no malignant changes. The features were consistent with Dupuytren’s fibromatosis. Independent review of the histology by a regional soft-tissue tumour pathologist (Professor Fisher, Royal Marsden Hospital) confirmed infantile fibromatosis consistent with Dupuytren’s.

At 21 months’ follow-up, the child had no functional disturbance of the hand. There was full active flexion but a 10˚ loss of active extension of the ring finger. There was no restriction.
of passive extension but there was a visible and linear subcutaneous scar on the radial side of the finger. No nodule or mass could be felt (Fig. 4).

Discussion

Dupuytren’s disease in infants is exceedingly rare. This is only the third recorded histologically-proven case in a child less than nine years of age and only the second in an infant. Other causes of flexion contracture of the fingers include camptodactyly, burns, congenital ulnar drift and infantile digital fibromatosis. Infantile digital fibromatosis (also known as ‘recurring digital fibroma of childhood’) typically presents in the first year of life as a single, firm, pink or red nodule on the dorsum of the fingers or toes. These nodules have the characteristic histological appearance of eosinophilic inclusion bodies in the cell cytoplasm. Fibrous tissue tumours are more commonly reported than these other causes of contracture.

Dupuytren’s disease in children can resemble tumours and may have a similar, infiltrative, histological appearance ‘with bands of proliferative spindle cells that blend with the surrounding tissue’. Tumours that need to be considered in the differential diagnosis include extra-abdominal fibromatosis, calcifying aponeurotic fibroma, infantile fibrosarcoma, fibroma of the tendon sheath, localised nodular tenosynovitis (giant cell tumour of the tendon
It should not be assumed that a swelling is a tumour without a tissue diagnosis. A diagnosis of Dupuytren’s disease should not be made without histology. One reason for operating in our case was to exclude malignancy. Despite the infiltrative nature of the tissue plaque the pathologist was certain of the histological diagnosis of Dupuytren’s. Despite the fact that removal was probably incomplete, recurrence has not occurred. In a review of 66 excised childhood fibrous tumours, only three proved to be palmar fibromatosis but no further specific details were mentioned.

The aetiology of Dupuytren’s disease is multifactorial and has been recently reviewed. The fact that it has now been described in two infants suggests a genetic causation. In the treatment of childhood Dupuytren’s disease, it has been suggested that aggressive dermofasciectomy should be performed. Urban et al described a nine-year-old boy who was treated initially by fasciectomy but required a dermofasciectomy 19 months later for recurrence. The only other reported case in an infant was treated by fasciectomy and no recurrence was noted at 27 months’ follow-up. We treated our case with fasciectomy and no recurrence has been observed at 21 months.

The optimal timing for surgery of this rare condition in infancy has yet to be established.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References