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CASE REPORT

Childhood Dupuytren's disease of the interphalangeal joint of the thumb

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Abstract To date, there are only 13 cases of Dupuytren's disease documented in the English literature in children less than 13 years and only seven cases histologically diagnosed in those under 10 years. We present a rare case of childhood Dupuytren's contracture involving only the interphalangeal joint of the thumb and review of current literature of the preferred management of Dupuytren's disease in childhood.

Keywords Childhood Dupuytren's disease · Thumb Dupuytren's disease

Introduction

Dupuytren's disease in children is exceedingly rare [1, 2]. To date, there are only 13 cases of Dupuytren's disease documented in the English literature in children less than 13 years and only seven cases histologically diagnosed in those under 10 years [1, 3–7]. Although the thumb and the first web space are the third most commonly affected sites after the ring and little fingers in adults, there is only one case of childhood Dupuytren's contracture involving the thumb [7, 8]. We present a rare case of childhood Dupuytren's contracture involving the interphalangeal joint (IPJ) of the thumb in isolation without clinical involvement of metacarpophalangeal joint (MCPJ) or the first web space, and review of the literature of the preferred management of Dupuytren's disease in childhood.

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Case report

A right-handed 12-year-old boy presented to our clinic with a one and a half year history of contracture to the IPJ of his non-dominant thumb. The contracture developed gradually over the years with no history of trauma or predisposing factors other than a positive family history. He was otherwise well with no relevant past medical history.

On examination, there was a flexion contracture across the IPJ of his thumb with an extension lag of 60° (Fig. 1). A fibrous band was palpable over the volar radial aspect of his proximal phalanx extending from the middle of the proximal phalanx, crossing the IPJ, and into the base of distal phalanx. The contracture was not correctable on passive extension. The skin over the contracture appeared healthy with no signs of puckering or pitting. There was no palpable nodule. His MCPJ and first web space appeared normal. Examination of his other hand, feet, and penis was unremarkable. Radiography was performed and showed no abnormality.

In theatre, the patient underwent exploration under general anaesthesia. Brunner's incision was made over the palpable band, and the neurovascular bundles were carefully identified and protected. A flat, fan-shaped cord of fibrous tissue resembling a Dupuytren's radial cord was found to extend from adjacent to the sheath of flexor pollicis longus (FPL) to the distal end of the A2 pulley of the thumb (Fig. 2). It was fully dissected and excised. Intraoperatively, manipulation was undertaken to ensure the IPJ achieved full range of movement after the excision. Because there was no sign of skin involvement, the skin was spared, and the wound was closed directly. The IPJ of the thumb was splinted in extension post-operatively, and physiotherapy commenced immediately after the wound was satisfactory. There was no perioperative complication.

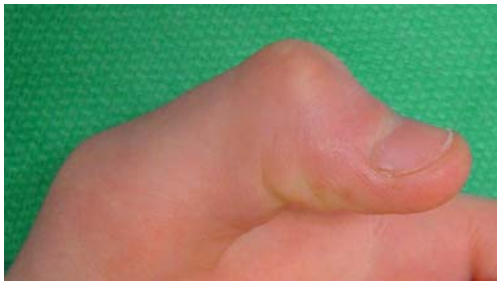

Fig. 1 Pre-operative photograph of flexion contracture of interphalangeal joint of left thumb with extension lag of 60°

The clinical diagnosis of Dupuytren's disease was later confirmed by histology.

A year after the fasciectomy, the patient returned with recurrence of disease on the same site and with skin involvement. The IPJ extension lag was 20°. A dermofasciectomy and full thickness skin graft was performed. The skin was excised from the mid-proximal phalanx level to the middle of the pulp with a clear margin of normal skin. Postoperatively, he was given physiotherapy regime, splinting, and scar management as in adults [2]. Histology finding revealed existing scar issue. Following the second operation, the patient continued to do well with no sign of further recurrence after one and a half year of follow-up (Fig. 3).

Discussion

Dupuytren's disease is a slowly growing superficial fibromatosis. Its aetiology remains unknown with multiple possible factors implicated, particularly its genetic predisposition [2]. It is considered to be a disease of adulthood, mainly involving patients between 40 and 60 years of age [7]. Histologically, it is characterised by proliferation of myofibroblasts in the early stage and production of collagen fibres later in the course of disease [2].

Dupuytren's disease in children was first described in 1954 by Goetzee and Williams [1, 3]. Although Dupuytren himself described this disease in a 6-year-old boy in 1832, there was no histological specimen to confirm diagnosis

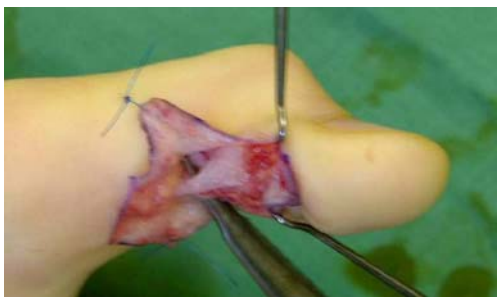

Fig. 2 Intra-operative photograph showing the fan-shaped fibrous cord extending over the interphalangeal joint causing joint contracture

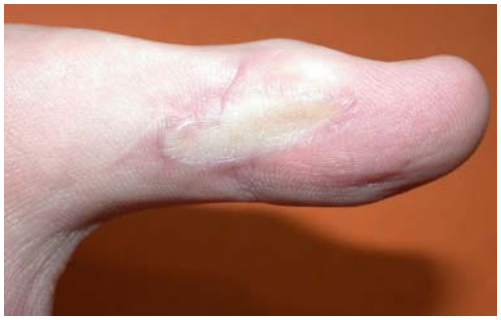

Fig. 3 Post-operative photograph showing the area of full thinkness skin graph following dermofasciectomy of the recurrent Dupuytren's contracture of the left thumb

[1]. At the time of writing, there are only 13 cases of Dupuytren's disease documented in the English literature of patients under 13 years old, with only seven cases histologically proven in those under 10 years [1, 3–7]. Large population studies have shown an incidence of 1% in children under the age of 14 years, but if histological confirmation is taken into account, this incidence would be much lower [7]. The youngest cases documented with histological confirmation occurred in infancy [3, 5, 7].

Because of its rarity, its diagnosis in children should be made with other differential diagnoses in mind. Some of the common causes of childhood finger flexion contracture are camptodactyly, burns, congenital ulnar drift, extra-abdominal fibromatosis, infantile digital fibromatosis (recurring digital fibroma of childhood), epithelioid sarcoma, juvenile aponeurotic sarcoma, infantile fibrosarcoma, giant cell tumour of tendon sheath (localised nodular tenosynovitis), fibrous hamartoma of infancy, and infantile myofibromatosis [1–3, 5, 7].

Although Dupuytren's disease of the thumb accounts for 28% of adult Dupuytren's contracture, it is not associated with Dupuytren's diathesis [8]. Four patterns of disease on the radial side of hand have been described by Milner and Tubiana et al.: a pre-tendinous cord inserting into the sheath of the FPL; a cord on the radial side of the thenar eminence; and two commissural cords, one lying distally and the other lying proximally in the first web space [8, 9]. There was no Dupuytren's contracture affecting the IPJ of the thumb recorded [8]. We describe the first case of isolated Dupuytren's contracture of the IPJ of the thumb in a child. Our case is also one of the only two cases of Dupuytren's contracture involving the thumb recorded in childhood. The first case was a typical contracture of the thumb similar to adult presentation where the child had MCPJ flexion and adduction contracture [7–9].

The management of Dupuytren's disease has been challenging because of its high recurrence rate. The recurrence rate in adults has been reported to be as high as 26% to 80% depending on the operative technique [2]. The risk of recurrence of disease in a paediatric age group

is uncertain [5]. It could be argued that dermatofasciectomy should be considered to prevent recurrence because of the possibility of Dupuytren's diathesis in this age group [2]. Urban et al. advocated aggressive treatment with dermofasciectomy in this age group but note that the 9-year-old boy they treated with dermofasciectomy still developed recurrence after 1 year of surgery [1]. Rhomberg et al. performed only fasciectomy on their patient with Dupuytren's disease of the thumb and skin closure with z-plasty. At 2 years, there was no sign of recurrence. Bebbington and Savage also experienced no recurrence after excision of Dupuytren's tissue on a 6-month-old infant with Dupuytren's disease on the palm and left ring finger. Foucher et al. recommended that fasciectomy was sufficient unless there was skin involvement to warrant dermofasciectomy. It should also be highlighted that the extent of dermofasciectomy should extend as far as each mid-lateral line across the palmar aspect of a finger [10]. On hindsight, the extent of dermofasciectomy on our patient has not extended as far to the ulnar side as advocated, and this could potentially predispose our patient to the need for future scar revision.

Based on the present literature, we do not feel that there is sufficient evidence to warrant an aggressive dermofasciectomy in the child's first presentation unless there is unequivocal evidence of a diffuse disease with skin involvement. Dermofasciectomy is only warranted in cases where recurrent disease or skin involvement are found. In conclusion, one should continue to adopt the same basic principles in the management of adult onset Dupuytren's disease to the management of childhood cases until proven otherwise.

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