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Case report

Triphalangeal thumb: case report and literature review

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Summary. Triphalangeal thumb (TPT) is a relatively rare congenital anomaly of the hand with evidence for autosomal dominant transmission. Surgical treatment is not always required, but according to Wood's classification it is related to the shape of the extra phalange. In this article, we present a case of a 32 year old woman with isolated bilateral TPT. (www.actabiomedica.it)

Key words: thumb, phalanx, finger, hand, malformation

Introduction

Overview

TPT is a rare anomaly described for the first time by Columbi in 1559 (1). The true incidence of the condition is unknown, but is estimated at 1:25.000 live birth (2). In about two-thirds of the patients there is a hereditary component (3). These cases, characterized by an autosomal dominant transmission (mutation on chromosome 7q36), are mostly non-opposable, bilateral and can occur with other malformations of the hand or foot (4). The sporadic cases are opposable and unilateral. Non hereditary cases may be linked to thalidomide (5).

The opposition of the thumb is a purely human characteristic but not exclusive because cercopithecoids (baboon) and hominidae (gorillas, chimpanzees, orangutans and gibbons) can oppose their thumbs to either grasp objects firmly or lightly if precision is required to complete a task (6). On the other hand, prosimians (tupaia, indri, aye aye, galacone, lori, tarsier) can only grasp. Throughout the evolutionary history of life on Earth, thumb length has been an important issue to allow its opposition with the long fingers of the hand and for in-hand coordination and manipulation of small objects. The gibbon has an extremely short thumb, which limits grasping of larger objects. If the thumb is extremely long, fine motor skills would be impeded. However, in the history of nature there are no reported cases of phylogenetic evolution of species with TPT.

TPT theories

Many theories are reported in the literature to explain the absence of a joint in the thumb in comparison to the long fingers. According to Olivier (7), at 39 days after fertilization (human embryo of 11 mm) the hand appears with 3 radii similar to tridactyla birds. Some authors sustain that in the human hand each finger, including the thumb, normally holds 3 phalanges whereas the metacarpus of the 1st ray has evolved into the modern trapezium. This could explain the curved shape of the 1st metacarpal bone, which resembles to the shape of the phalanges. The nutrient foramen of the 1st metacarpus is also similar to one of the phalanx. Further studies could also determine the amount of the ossification. Other authors consider that a fusion occurred between the 1st metacarpus and the proximal

phalanx. Safety and Poirier (7) argued that the proximal epiphysis of the 1st metacarpus corresponds to the original one and the body of the metacarpus represents the missing phalanx. Finally, Windle's (8) theory of a shortened 1st ray is based on the absence of the distal phalanx.

Anomalies can also be observed in the foot. In fact, the 5th ray often presents with a spontaneous fusion of the middle and the distal phalanges, which resembles to the phenomenon seen in the 1st ray of hand.

Classifications

Many classification systems can be found in the literature. Wood classified this malformation by assessing the shape of the extra phalanx (9). The 3 types of TPT are the delta type with ulnar deviation at the interphalangeal joint (type 1), the trapezoid type (type 2) and a fully developed extra phalanx (type 3). De Mas and Mele classified TPT from a functional point of view (10). The amount of thumb opposition in relation to the long fingers of the hand is assessed and ranges from complete opposition to lateral pinch, i.e. adduction in the same plane of the fingers (non-functional opposition). Buck-Gramcko proposed a more extensive classification, which incorporates additional factors such as intrinsic musculature, web space and length (11). The extended Wassel classification can be used to further classify combined radial polydactyly and triphalangism, if present (12). On the other hand, the International Federation for Societies for Surgery of the Hand (IFSSH) simply suggests considering TPT as a group III of the congenital hand malformation classification, i.e. polydactyly (13).

Treatment

Many techniques and approaches have been proposed in the literature to correct TPT. In 1940 Bunnell (14) initially advised against any type of surgery and a decade later Milch (15) advised to remove the supernumerary phalanx only in children. In 1976 Wood (16) recommended the removal of the supernumerary phalanx in children and osteotomy or arthrodesis in symptomatic adults. More recently, Horii (17) recommended surgical correction of the malformation 1 or 2 years after birth in children when the ossification centers are evident radiographically. In adults, surgery is recommended only when functional limitation is reported, but as mentioned by Gousheh (18), patients generally adapt well to their deformity during their life span and generally do not report difficulties, as it was in our case.

Other authors recommend surgery according to the patient's age and type of deformity (19-23). If the TPT is non-opposable (on the same plane of movement of long fingers), a derotational osteotomy is recommended with Kirschner wire (K-wire) stabilization and plaster immobilization into opposition (7). If the TPT is opposable, the treatment is based on the type of deformity (Wood's classification) with the goal to correct the angle of the deformity , stabilize the new interphalangeal joint and shorten the ray for improved grasp and pinch (17). Type 1 requires the removal of the delta phalanx by lateral access and tensioning of the collateral ligament with a capsular flap and K-wire stabilization. Horii (17) recommends early intervention in order to "reprogram" a normal movement pattern of the thumb. He also recommends this intervention or partial removal of the epiphysis in type 2 TPT, and removal of a subsequent deformity if it occurs at bone maturation. Type 3 requires an osteotomy of claw phalanges, which are fused with the shaft of the middle phalanx in order to have a single new joint. In this latter type of deformity, a dorsal V-shaped access is needed to do the osteotomy and fusion. Just recently, Wang (24) controlled 14 type 1 patients treated with this method and showed good functional and aesthetic evolution. Zuidam (25) compared outcomes between the mere removal of supernumerary phalanx osteotomy and arthrodesis in 33 hands and found no differences at follow-up between the 2 techniques.

Case report

In January 2015 a 32-year-old Brazilian woman presented herself to our attention for an accidental injury to the extensor tendons of the 5th finger of the right hand. During this emergency visit (ER), the admitting physician noticed a longer than normal 1st digit in both hands, with ulnar deviation of the left side (Figure 1). The patient reported to have no limitations

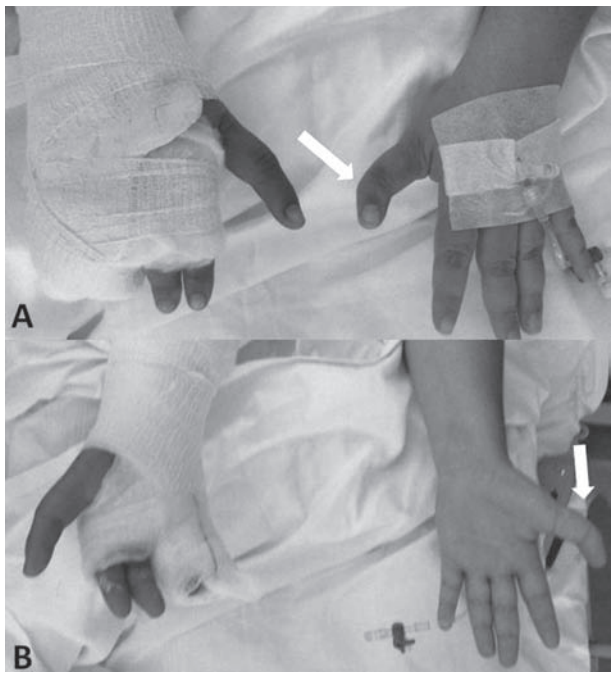

Figure 1. Bilateral TPT. Dorsal (A) and palmar view (B). Ulnar deviation of the left thumb (arrows)

during daily activities (ADL's). Radiographs, taken in the emergency room to exclude bone lesions to the little finger, confirmed the presence bilaterally of an additional phalanx (Figure 2). The patient was in good health, did not present any other type of malformations or concomitant disorders. Similar isolated cases of TPT were reported in her family, which confirms the hereditary dominant autosomal gene. It was not possible to perform a detailed radiographic and functional assessments of her family because of the remoteness of those having TPT.

During her ER visit, the patient was able to oppose the thumb normally, her pinching strength and precision with the tips of the long fingers were judged normal and the muscles of the thenar eminence appeared normal. It was unfortunately impossible to assess in greater depth her thumb's functional capacity because her stay in Italy was temporary and could not return for follow-up. Thus, only a single morphological study through radiographs and Magnetic Risonance Imaging (MRI) were possible in this case.

Radiographically, the first digit of the right hand showed a normally shaped additional middle phalanx (Wood's type 3) (Figure 2A), while contralateral thumb had a trapezoid shaped additional middle phalanx (Wood's type 2) (Figure 2B).

The patient agreed to undergo an MRI (Figure 3) on her hands to assess whether the soft tissue anatomy was altered by the presence of an extra phalanx. The MRI confirmed that the muscles of the thenar emi-

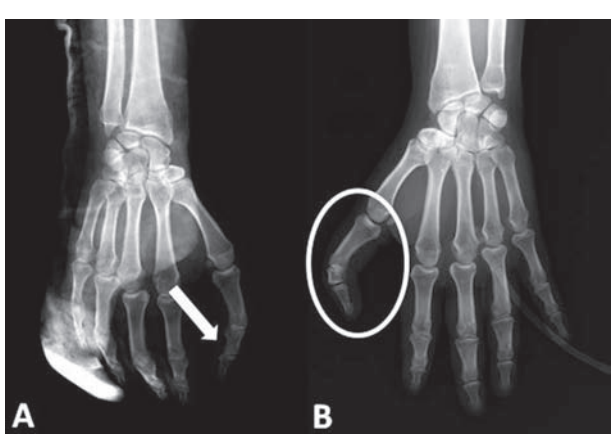

Figure 2. Radiographs of both hands TPT. Fully developed right extraphalanx (A) (arrow) and trapezoid type of the left thumb with ulnar deviation (B) (circle)

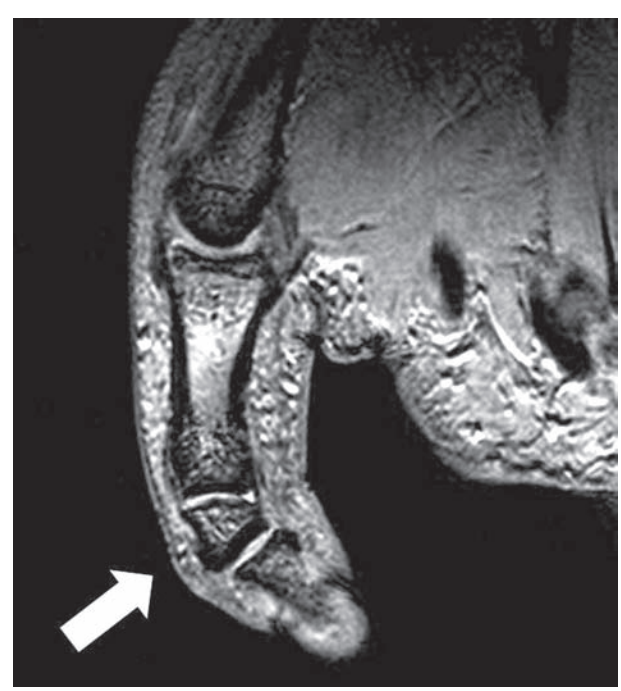

Figure 3. MRI coronal DP fat-sat image of trapezoid type 2 TPT of the left hand with ulnar deviation (arrow)

nence had a normal structure with insertions of the Abductor Pollicis Brevis, Flexor Pollicis Brevis and Adductor Pollicis tendons located at the base of the proximal phalanx to the thumb, and insertions of the Opponens Pollicis at the mid shaft portion of the 1st metacarpus. Extrinsic thumb muscles also had normal origins and insertions. The Flexor Pollicis Longus (Figure 4 and 5) and Extensor Pollicis Longus (Figure 5) tendons inserted at the base of the distal phalanx, the Abductor pollicis Longus at the base of the 1st metacarpus and the Extensor Pollicis Brevis at the base of the proximal phalanx.

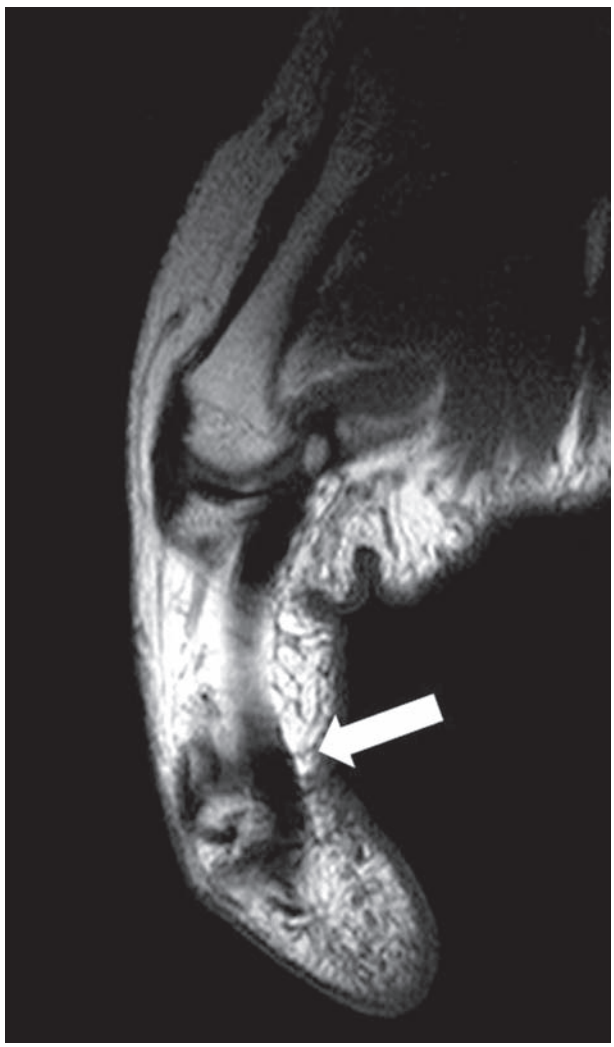

Figure 4. MRI volar coronal SE-T1w image of the left hand with insertion of the flexor pollicis longus tendon at the base of the distal phalanx (arrow)

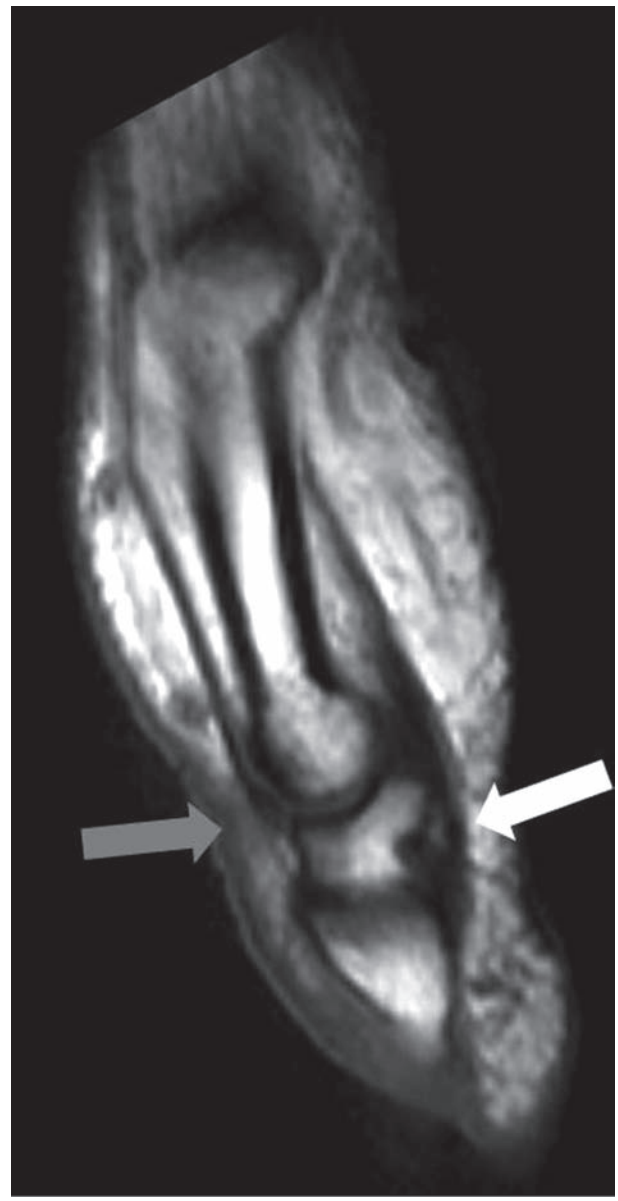

Figure 5. MRI sagittal view SE-T1w of the left hand with insertion of the flexor pollicis longus tendon (white arrow) and of the extensor pollicis longus (grey arrow) at the base of the distal phalanx

The additional phalanx was thus void of any tendon insertion, which may explain why the patient showed normal thumb function and did not seek medical attention for her TPT in the past.

Discussion

Other cases of adult TPT can be found in the literature. Most people with TPT present the malformation bilaterally as an hereditary disease (20). They generally do not show limitations in range of motion and only in a few cases the metacarpalphalangeal joint (MCPj) is unstable (26). Intrinsic movements of the thumb (anteposition, opposition, MCPj flexion) (20) and grip and pinch strength are affected negatively (26,27). Self-reported function is not impeded but the presence of uni- or bilateral TPT does influence social aspects of patients (18,26).

Our TPT patient, as mentioned earlier, was brought to our attention for an unrelated hand problem and could not return for extensive assessment as she became unreachable for follow-up. We therefore could not fully subjectively and objectively assess the patient with standardized tools. However, in the brief time we had at disposal, we were able to classify her TPT as a Wood's Type 3 (right) and Type 2 (left) and rule out any soft tissue anomalies through the MRI. Similarly to most patients with this malformation, our patient was female with a family history of TPT, had bilateral deformities and reported full function. Upon examination, no abnormal movements or instability were noticeable. Our patient mentioned she never sought medical attention for her TPT as she could use it in all her ADL's. Appearance also did not seem to be an issue in our patient, unlike most other cases found in the literature, but we could not truly investigate on possibly the most important aspect of TPT.

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