Monozygotic twins discordant for pediatric trigger-locked thumbs

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Published online: 4 May 2012
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Introduction

Pediatric trigger-locked thumb (PTLT) is a condition frequently encountered by hand surgeons. The term pediatric is more accurate than congenital because most cases do not present at birth. It is possible however that some children are born with the condition but it manifests after birth. The etiology, natural course, ideal treatment, and the age for surgical intervention were all debated in the literature. Children with PTLT typically present under the age of four with the interphalangeal (IP) joint of their thumb locked in flexion. The condition however is seldom preceded by catching, snapping, or triggering of the thumb, which may cause dysfunction and pain. A disparity in the size between the flexor tendon and the surrounding retinacular pulley system, most commonly at the level of the first annular (A1) and partly the oblique pulleys, results in difficulty of flexor pollicis longus tendon gliding hence the triggering/locking phenomenon. This abnormality typically presents at a mean age of two with the IP joint locked in flexion with a palpable nodule in the flexor pollicis longus tendon called Notta's node. One study observed that in 8.5 % of the cases the condition was diagnosed in the first 2 months following birth, in 53.2 % of the cases between 2 months and 2 years, and in the remainder after 2 years of age [1].

There is some debate about the presentation and whether this condition is truly congenital and present at birth, the pathology is present at birth but manifests later in infancy and childhood, or truly acquired and develops after birth.

For these reasons, the term pediatric trigger-locked thumb is more encompassing.

The incidence of acquired or pediatric trigger thumb in children under 1 year of age is 33 per 10,000 live births. Bilateral involvement is reported between 25 and 50 % [1, 4]. The treatment of PTLT is releasing the stretched A1 and part of the constricting oblique pulley and restoring flexor tendon excursion and thumb IP joint extension.

There has been limited number of literature publications on the etiology of PTLT. The few articles available in the literature on this topic suggest that genetic factors are important in its etiology [5–8, 10, 11]. There are three case reports of “congenital trigger thumb” in twins [3, 4, 8] that support the genetic factors in the etiology of the condition. These reports are based on the fact that the condition occurred in “identical” twins but without a proof of the monozygotic nature of the twins with DNA analysis. We report a case of monozygotic twins discordant for the condition: one with bilateral congenital trigger-locked thumbs and the other with no thumb abnormalities.

Case Report

Twin A presented to our clinic as a 22-month-old female with bilateral locked trigger thumbs of 3 months duration. There were no previous interventions or formal physical therapy. Her mother stated that the twin girls are identical twins based on the monochorionic pregnancy. The twins were delivered at 34 weeks without complications.

Examination of twin A showed bilateral locked trigger thumbs, with palpable Notta's nodules and hypermobile metacarpophalangeal (MP) joints. Examination of twin B showed no detectable musculoskeletal abnormalities in the hand or thumb.

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At age of 2 years, twin A underwent bilateral release of A1 pulley and partial release of the oblique pulley. A transverse skin incision was made over the palmar flexor crease of the MP joint. Full passive IP joint extension was restored to neutral position in both thumbs. Notta's node within the flexor tendon was large and palpable but required no debulking which is rarely indicated.

At 9-month follow-up, twin A had full IP joint extension without limitation or palpable Notta's nodule in both thumbs. Twin B again was examined at the same time and found to have no abnormalities in both thumbs.

Postoperatively, DNA analysis was performed for both twin sisters (Fig. 1). DNA was extracted from both twins and genotyping was performed. This strongly supports the monozygosity of the twins.

Discussion

Monozygotic twins status should not be based on the phenotype and identical appearance. Genetic testing is necessary to confirm the monozygotic nature of twins. Previous case reports about congenital trigger thumb among twins lacked information about DNA analysis. Therefore DNA from both twins in our case report was analyzed with several markers. This confirmed their monozygotic nature and hence the discordance of this condition. There is no agreement among investigators about the pathophysiology of this condition. In 1949, Sprecher [7] suggested that PTLT has similar pathology to the adult form. In 1974, Hueston and Wilson [2] stated that the Notta's node was caused by spiraling of the fiber components within the stenosed pulley sheath. Van Genechten [9] felt that the condition was complicated by the proximity of the sesamoid bones to the MP

joint. Some authors also discussed the pathology as a problem simply arising from the tendon, sheath, or pulley. All these pathologies can be used as an argument for an acquired nature of the condition. Herdem et al. [1] discussed the possibility of the genetic etiology of PTLT and supported the argument by the positive family history, multiplicity of pathology in twins, or other genetic anomalies including syndactyly or cleft lip and palate [4]. While much has been discussed about the pathophysiology of the congenital trigger thumb, the debate continues about its cause: a genetic versus acquired etiology. Some studies in the literature lean towards a genetic cause for the condition [3, 8–11], whereas others argue that it may be acquired in nature [12, 13]. Regardless of the true cause of the condition, it is possible that some children are born with the condition but it manifests later in infancy or childhood.

The majority of patients with pediatric trigger-locked thumb present with locked thumb in flexion rather than triggering and do not present at birth but later in infancy [14]. Hence the term pediatric trigger-locked thumb is preferable to congenital trigger thumb.

Our case is the first reported of monozygotic twins with phenotypically different thumbs with one sister having bilateral trigger-locked thumbs and the other free of this abnormality. We cannot make definitive conclusions from one case report, but our case may argue against the notion that there is a genetic component to pediatric trigger-locked thumbs. Perhaps acquired factors are more important than genetic factors in the etiology of this condition.

Acknowledgments The authors would like to thank Mr. Joshua Cavett for helping with the genotyping experiments and Ms. Annette Kezbers for her administrative help in preparing this manuscript for publication.

Conflict of interest The authors declare they have no conflicts of interest, commercial associations, or intent of financial gain regarding this research.

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Fig. 1 Postoperative DNA analysis performed for both twin sisters

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