Mirror-image Trigger Thumb in Dichorionic Identical Twins

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abstract

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The congenital vs acquired etiology of pediatric trigger thumb is the subject of considerable debate. Existing case reports of bilateral presentation in identical twins and first-degree familial association support the congenital hypothesis. However, prospective studies have yet to report a neonate presenting with this anomaly at birth. This article describes the first known set of dichorionic, monozygotic identical twins with unilateral trigger thumbs, affecting contralateral (mirror-image) hands and with asynchronous age at presentation (11 months and 18 months, respectively).

Pediatric trigger thumb is caused by a mismatch between the flexor pollicis longus tendon and its A1 synovial pulley. Four sets of twins have been previously reported in the literature with trigger thumb. Of these, 3 sets were monozygotic twins who had bilaterally affected thumbs. Together with the absence of trauma, a congenital etiology was suggested. The fact that pediatric trigger thumb is generally seen several months after birth was felt to be due to infants holding their thumbs clutched in their palms until 6 months. However, no confirmed cases of trigger thumb have been diagnosed at birth in several large prospective studies of newborns.

In the current case, the asynchronous presentation of unilateral trigger thumbs in identical twins does not support a solely congenital cause. Furthermore, the mirror-image presentation contradicts current embryological understanding of the temporal course of twinning and the determination of laterality. Thus, a multifactorial etiology is supported with both a genetic and acquired component affecting the development of this condition.

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Trigger thumb was first described in the literature by Notta in 1850. Patients present with a nodule on the flexor tendon at the level of the metacarpal head, which interferes with normal motion. In the pediatric population, the thumb is the digit most commonly afflicted with triggering. The reported incidence of trigger thumb in children aged 1 year ranges from 0.5 to 3 per 1000 live births. Pediatric trigger thumb is a disorder caused by a mismatch between the flexor pollicis longus tendon and its sheath. It is felt to be due to either thickening or synovial proliferation in the flexor pollicis tendon more commonly than from a constricting fibrocartilagenous band in the annular sheath.

Pediatric patients often present many months after birth when a parent notices that the thumb cannot be fully extended. It is speculated that the late presentation is due to the tendency for infants younger than 6 months to hold their thumbs flexed in their palms. The fixed flexion deformity of the interphalangeal joint of the affected thumb is typically the first recognized symptom. Physical examination demonstrates that passive extension of the affected digit is difficult beyond 30° and can rarely be accompanied by triggering, snapping, or a digit locked in extension. A hard, palpable nodule (Notta’s node) at the base of the metacarpal is noted and felt to represent the thickened, bunched up flexor pollicis longus tendon at the base of the A1 pulley. It has been reported that patients presenting soon after birth have a 30% rate of spontaneous resolution, whereas those presenting after 6 months of age have an overall spontaneous resolution rate of <12%. More recently, authors have reported a higher rate of spontaneous resolution in older children. The definitive treatment for this condition is felt by most to be operative in patients in whom it has not resolved by age 1.

The deformity was initially thought to be congenital in nature due to case reports describing its occurrence in twins and a positive family history. However, recent large prospective studies have failed to demonstrate neonates born with this condition, suggesting an acquired etiology. The true etiology of trigger thumb remains unknown. This article presents a case of dichorionic monozygotic twin boys presenting with asynchronous mirror-image (contralateral) trigger thumbs, a unique presentation that, to the authors’ knowledge, has not been previously reported in literature. These phenotypic differences in identical twins can be caused by a combination of genetic and environmental factors. Mirror imaging in identical twins is a rare phenomenon that is felt to be caused by a late cleavage of monozygotic twins.

CASE REPORT

Identical twin boys, patient A and patient B, presented to the hand surgery clinic aged 12 and 18 months, respectively. Patient A’s pediatrician referred him after his mother first noticed at approximately 11 months that his right thumb was becoming stuck in a flexed position. The diagnosis of trigger thumb was made on examination, which demonstrated a flexion contracture and a palpable Notta’s node (Figure A). The parents declined surgery and elected to proceed with conservative treatment. While patient A was being observed, the mother noted that patient B’s left thumb started to become stuck in flexion beginning at 17 months. The diagnosis of trigger thumb was made on examination, which demonstrated a flexion contracture and a palpable Notta’s node of the left thumb (Figure B). Both twins had intact neurovascular function and no history of traumatic injury to the hand. They were able to fully flex and extend their contralateral thumbs with no triggering or evidence of a Notta’s node. No other hand, upper extremity, or congenital anomalies existed. No history of trauma, significant illness, or family history of trigger thumb existed. No other evidence of mirror imaging was noted.

Nonoperative therapy failed to resolve the triggering in either twin, so the parents decided to proceed with surgical release. Both twins were successfully treated by surgically releasing the A1 pulley of the involved thumb. Follow-up at >1 year postoperatively confirmed recovery of full extension in both twins without development of a contralateral trigger thumb. Further genetic testing confirmed monozygosity with >99% confidence.

DISCUSSION

To the authors’ knowledge, the presentation of mirror-image trigger thumbs in monozygotic twins has not been previously reported; however, 4 case reports of twins presenting with pediatric trigger thumb have been reported in the literature. The earliest reported case occurred in dizygotic twins, 1 with bilateral involvement and the other with unilateral involvement. Three cases of monozygotic twins have been reported in literature, with both twins affected bilaterally. Two cases had bilateral trigger thumbs, and 1 case had bilateral trigger thumbs and digits. The exact timing of presentation in these case reports relied on parental recollection and thus could not be reliably ascertained. The bilateral identical twin concordance suggested a genetic factor or a congenital predisposition.

Monozygotic twins develop from a single fertilized ovum through fission (division) shortly after fertilization. If division of the embryonic cells occurs within 2 to 3 days, 2 amnions, 2 chorions, and...
2 placentas (25% of the time) will result, known as dichorionic monozygotic twins. If cell division occurs within 4 to 8 days, 2 amniotic sacs, 1 chorion, and 1 common placenta (75% of the time) results, known as monochorionic monozygotic twins.17

Mirror image is a poorly understood phenomenon that causes phenotypically opposite anomalies or differences in monozygotic twins. It is felt that the craniocaudal and dorsoventral axis are defined earlier in the zygote than left-right orientation (laterality). Mirror image is felt to occur later in cell division when some differentiation of the inner cell mass has occurred (4 to 8 days) and laterality has started to be defined. At this stage, division of the zygote will require redefining the right–left axis, which may lead to mirror image. Because mirror image is more common in conjoined twins than in identical twins, it is felt that the later the embryo divides, the higher the rate of mirror image. In identical twins, mirror imaging has been described in handedness, malformations, and other congenital conditions.18-20

In the current case, the exact contribution of the mirror imaging phenomenon to the contralateral presentation of trigger thumbs is unknown. Although it is extremely unlikely given the low incidence of this condition, the authors recognize the possibility that this presentation may be due to pure chance.

Recent studies have challenged the idea that pediatric trigger thumb is truly a congenital condition. Four existing prospective studies of a total of 14,581 newborns failed to find trigger thumbs at birth.5,14-16 Furthermore, pathological reports of 2 cases demonstrated changes consistent with an acquired traumatic etiology. The microtrauma associated with flexed position of the thumbs during the perinatal and neonatal period has been suggested as a possible mechanism for the development of the condition.5

The exact contribution of a congenital vs an acquired etiology in trigger thumb remains unknown. The literature remains sharply divided, arguing that it is either congenital or acquired. The fact that the current identical twin patients had an asynchronous presentation of contralateral (mirror-image) thumbs would indicate an acquired component; otherwise, a simultaneous presentation would be expected. Because the trigger thumbs did not occur at birth, they cannot be said to be truly congenital. This case does not exclude the possibility of a congenital contribution but lends less credence to a truly congenital etiology as an explanation for previously observed bilateral trigger thumbs in identical twins. Given this and the fact that no case report exists in which only 1 identical twin developed a trigger thumb, a solely acquired etiology is unlikely. Based on this case and the existing literature, it appears that pediatric trigger thumb has a multifactorial etiology.

REFERENCES