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Case Report

Iso-Kikuchi Syndrome in A Newborn and His Mother

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Abstract

Iso-Kikuchi syndrome or congenital dysplasia of the index finger is a rare entity that affects primarily the nails of both index fingers, and presents with onychodysplasia or complete anonychia. The etiology remains unknown. We present two familial cases of this unusual entity.

Keywords: iso-kikuchi syndrome, anonychia, onychodysplasia, polyonychia, congenital dysplasia of the index finger

Introduction

Iso-Kikuchi syndrome or congenital onychodysplasia of the index fingers is a rare entity that affects primarily the nails of both index fingers and can compromise the phalanges. It produces total anonychia or dysplasia of the nails [1-3].

In 1969 Iso described seven cases from his own practice and collected two others from the literature with anomalies in the nails of the index fingers which included anonychia, micronychia and polyonychia. He proposed the term of congenital nail defects of the index fingers [2, 4, 5]

In 1974 Kikuchi et al. reported two patients with nail alterations and bone anomalies in the index fingers and suggested the term congenital onychodysplasia of the index fingers [2].

In 1980 Baran and Stroud proposed to name Iso and Kikuchi syndrome because in some cases it affected other fingers in addition to the index ones [4].

The pathogenesis is unknown; some authors suggest a genetic abnormality or ischemia during the fetal period. A complete family history is important to determine if it is a sporadic or a hereditary condition [6-7].

Case Reports

The patient was a 24-hour-old full-term baby, delivered by caesarean section. He presented an anonychia of the left index finger (photo 1) and polyonychia on the right index (photo 2). No other abnormalities on the physical exam were found.

With the suspicion of Iso-Kikuchi-syndrome his 26-year-old mother was evaluated. She presented congenital micronychia on her left index finger (photo 3 and 4) and limited mobility of the distal phalanx. These were her only nail abnormalities. An X-ray showed a slight deviation of the distal phalanx of the affected index which confirmed our diagnosis (photo 5).

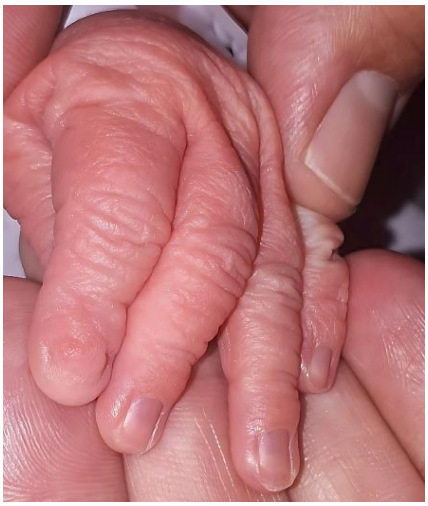

Figure 1: anonychia of the left index finger

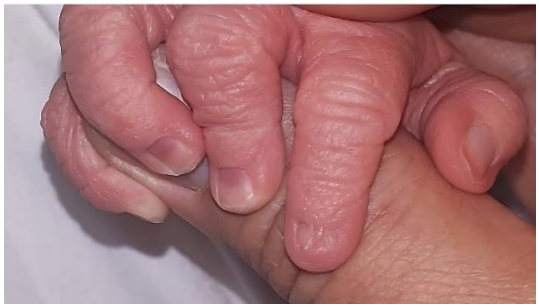

Figure 2: polyonychia of the right index finger

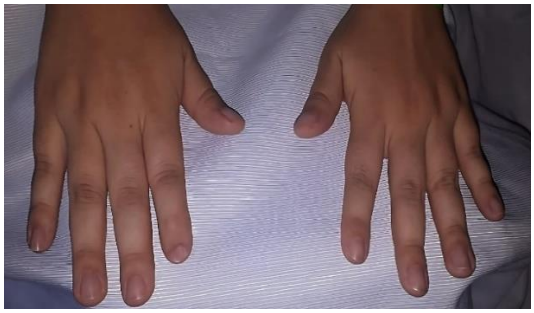

Figure 3: micronychia of the left index finger

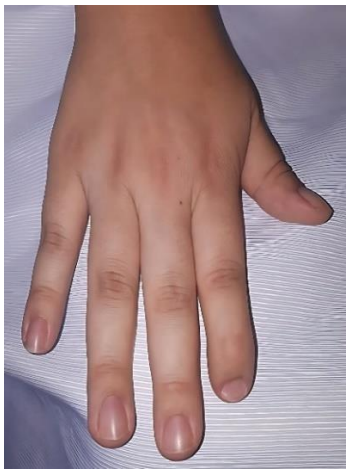

Figure 4: micronychia of the left index finger

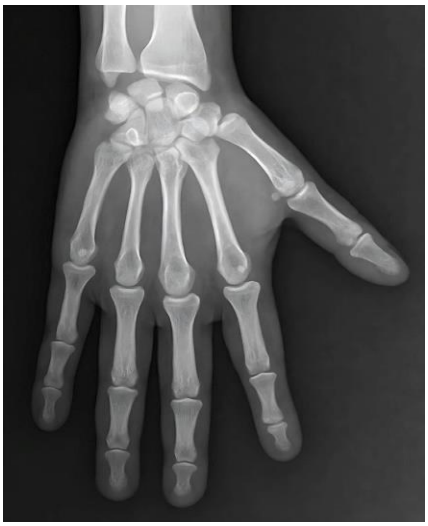

Figure 5: X-ray showed a slight deviation of the distal phalanx of the affected index

Discussion

Iso-Kikuchi syndrome is a benign and infrequent entity. International incidence is about 4.2/100000 births [3]. It was first reported in Japan, and later in other countries such as the USA, England, France, Korea [5], Argentina [1], Brazil [7] and Italy [3, 5, 8]. It affects primarily the nails of the index fingers, especially the medial aspect of the nail and it can also affect the underlying bone [1-4,8].

Anomalies include: polyonychia – where there are two small incomplete nails on one finger with the lateral one being larger than its counterpart–, micronychia – where a small nail is seen in the lateral aspect of the finger, anonychia –meaning the total absence of the nail–, hemionychogryphosis, alterations of the lunula [2] and misalignment [1, 4, 5, 7, 9-11].

Bone changes were first described by Kikuchi et al. 2 and include: narrowing of the distal phalanx and, more frequently, an enlarged nail with a Y-shaped bifurcation. Other associated changes are: brachydactyly, brachymesophalangia of the fifth fingers, congenital flexion alterations of the index fingers and cutaneous syndactyly of an index and middle finger [1, 4, 11].

The diagnosis of this syndrome is clinical 1. The first Iso's description of this entity was as follows: 1) congenital 2) both index fingers and never the toenails affected 3) with variable nail involvement but always more pronounced on the medial aspect rather than on the lateral one 4) non-familial 5) without abnormalities of the bone and joint system [2].

Years later, in 1980, Baran and Stroud suggested a modification of the criteria: congenital occurrence, unilateral or bilateral index finger involvement, variability in nail appearance, possible hereditary involvement and frequently associated bone abnormalities [1, 3, 4, 7, 8, 11].

Finally, Franceschini et al. in 2001 proposed these criteria that more fully encompass this entity: 1) bilateral or unilateral index fingernail involvement 2) congenital occurrence 3) onychodysplasia characterized, with decreasing frequency, by micronychia, polyonychia (split nail: pathognomonic when present) anonychia, hemionichogryposis and malalignment 4) underlying bony phalanx anomalies (mainly bifurcation of distal phalanx only evident in lateral projection) and 5) hereditary [5, 9, 11]. The two patients we describe in this article fit the criteria mentioned above. There are some differential diagnoses to be considered (table 1)

Although it can occur sporadically2, as several members of the same family have been reported to have similar alterations, Harper and Beer suggested an autosomal dominant inheritance with variable expression [6, 8].

The etiology is unknown but several mechanisms have been postulated:

• A fetal grip could cause ischemia of the nail and phalanx leading to dysplastic changes and even resorption of these structures. However, this is not consistent with the fact that the fetus does not have the ability to have a strong grip before the limbs and bones are developed (one of the first hypotheses).

• A selective abnormal fetal vascular supply from the palmar digital artery that causes ischemic injury.

• A dysplastic change in the crescent-shaped cap of the distal phalanx.

• Genetic mutations causing alterations of the WNT signaling pathway.

• Fetal exposure to teratogens such us phenytoin, carbamazepine and valproate [1, 8, 9, 11].

Differential Diagnosis
Congenital anonychia
Onychoheterotropia
Syndactylia and polydactylia
Congenital onychoatrophia
Dyskeratosis congenital
Chondro-ectodermal dysplasia
Nail-patella-elbow syndrome
Congenital ectodermal defect
Deafness-onychodystrophy syndrome

Table 1: differential diagnosis of Iso-kikuchi 1,2

Treatment of this condition is optional and it is usually defined by associated symptoms, such as pain or cosmetic concerns. Urea cream has been used to soften the nail plate. However, if symptoms persist, a partial matricectomy of the curved portion of the nail is suggested. [10].

Conclusion

We present two members of the same family with this infrequent nail anomaly whose diagnosis is eminently clinical. Dermatologist should be aware of this syndrome in order to provide an early diagnosis and avoid unnecessary tests and treatments.

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