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E- CASE REPORT

Iso-Kikuchi Syndrome: Report of 3 Pediatric Cases☆

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KEYWORDS
Iso-Kikuchi;
Anonychia;
Nail;
Index finger;
Phalanx

Abstract Iso-Kikuchi syndrome, or congenital onychodysplasia of the index finger, is an uncommon condition characterized by total anonychia or dysplasia of the nail of the index finger. It is occasionally accompanied by underlying bone abnormalities and is rarely associated with other conditions. Although various hypotheses have been put forward to explain the pathophysiology of the syndrome, its etiology remains unknown.

We report the cases of 3 pediatric patients (2 boys and 1 girl) with nail changes and bone abnormalities consistent with Iso-Kikuchi syndrome. We highlight the importance of recognizing this entity early to avoid the need for additional tests and unnecessary treatment.

PALABRAS CLAVE
Iso-Kikuchi;
Anoniquia;
Uña;
Dedo índice;
Falange

Síndrome de Iso-Kikuchi: 3 casos en la edad pediátrica

Resumen El síndrome de Iso-Kikuchi, onicodisplasia congénita del dedo índice, es una entidad poco frecuente caracterizada por la anoniquia total o displasia de la uña del dedo índice, acompañado, en algunas ocasiones, de alteraciones óseas subyacentes, por lo general, en ausencia de otras anomalías. Si bien se han planteado distintas hipótesis fisiopatogénicas, la etiología sigue siendo desconocida.

Describimos los casos de 3 pacientes pediátricos, 2 varones y una niña, con alteraciones ungueales y óseas compatibles con el síndrome de Iso-Kikuchi. Destacamos la importancia de reconocer esta entidad tempranamente para evitar la realización de estudios complementarios y terapéuticas innecesarias.

Introduction

Iso-Kikuchi syndrome is characterized by anonychia or onychodysplasia of the index finger, accompanied or not by underlying bone abnormalities. It is a benign condition that is not associated with alterations to other organs or systems. Very few cases have been reported to date.

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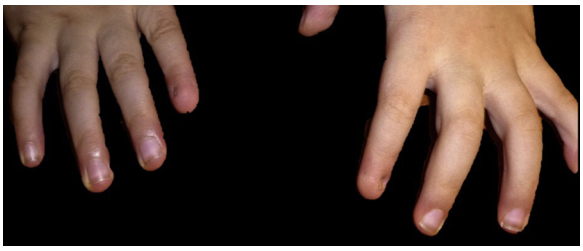

Figure 1 Patient #1. Note the micronychia on the radial side of both index fingers.

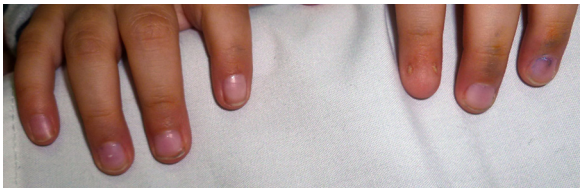

Figure 2 Patient #2. Unilateral involvement of the left index finger with micronychia on the ulnar and radial sides of the nail bed. The rest of the nails are unaltered.

Case Descriptions

Two boys and 1 girl presented with Iso-Kikuchi syndrome for the first time between the ages of 2 and 4 years.

Patient #1, a 2-year-old boy, had bilateral index finger involvement and a Y-shaped deformity of the underlying phalanx (Fig. 1). His past medical history was remarkable for extreme preterm birth (30 weeks), which is why we have included other diseases associated with this condition in Table 1.

Patient #2, a 4-year-old girl, had involvement of the left index finger and a Y-shaped bifurcation of the affected phalanx (Fig. 2).

Patient #3 was 2 years old and had bilateral index finger involvement (Fig. 3) without radiographic changes (Table 1).

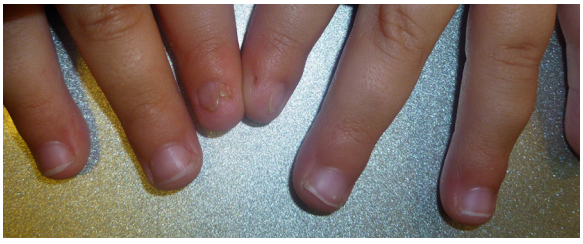

Figure 3 Patient #3. Polyonychia of the left index finger and nail dystrophy of the right index finger.

None of the patients' relatives had Iso-Kikuchi syndrome. All the parents denied consanguinity and reported that their children's nail alterations had been present since birth.

Discussion

Iso-Kikuchi syndrome was first described by Dr. Iso in 1969 and Dr. Kikuchi in 1974,[1–3] and very few cases have been reported since (Table 2). It affects both sexes equally and can be sporadic or familial.

Iso-Kikuchi syndrome runs an indolent course that is characterized by anonychia or onychodysplasia of the index finger possibly accompanied by bone abnormalities in the underlying phalanx.[4] It can affect one or both hands or feet. Based on their observations and analysis of different clinical cases, Baran and Stroud[5] proposed the following diagnostic criteria for Iso-Kikuchi syndrome: *1*) unilateral or bilateral hypoplasia (up to complete anonychia) of the index finger and/or other fingers or toes, 2) radiographic changes in the distal phalanx of the affected digit; and 3) sporadic or hereditary congenital occurrence. Transmission of Iso-Kikuchi syndrome is autosomal dominant. No associations with systemic diseases have been reported.[4,5]

Onychodysplasia has multiple presentations (Table 2), the most common of which are anonychia, micronychia (characteristic deviation of the nail to the ulnar side of the nail bed),[6]polyonychia (2 nails, 1 on each side of the nail bed),

Table 1 Cases Presented.

Patient	1	2	3
Sex	Male	Female	Male
Age	2 y	4 y	2 y
Perinatal history	Born at 30 wk; nail changes present at birth	Term delivery; nail changes present at birth	Term delivery; nail changes present at birth
Digit involved	Both index fingers	Left index finger	Both index fingers
Appearance of other nail	Micronychia on radial side	Micronychia on radial and ulnar sides	One finger with polyonychia, dystrophic homolateral finger
Radiographic changes	Bilateral Y-shaped deformity	Y-shaped deformity in affected finger	None
Other alterations	Grade 3 intraventricular hemorrhage, bilateral inguinal bleeding, bilateral pelviureteric junction stenosis	None	Gastroesophageal reflux
Family history	No	No	No