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Abstract: The Iso-Kikuchi Syndrome is a rare condition characterized by nail dysplasia involving the index fingers, including micronychia, polyonychia, anonychia, irregular lunula, malalignment and hemionychogryphosis. On the antero-posterior image, radiologic examination reveals a narrowing of the distal phalanx. The lateral image shows a Y-shaped bifurcation of the distal phalanx. We report a case of a patient with typical clinical and radiologic signs of Iso-Kikuchi Syndrome.
Keywords: Congenital abnormalities; Nails; Nails, malformed

CASE REPORT

A 13-year-old patient born and living in Sao Paulo sought the dermatology clinic of the Hospital do Servidor Público Municipal de São Paulo with a complaint of birth-onset nail changes on the index fingers. The patient denied a history of consanguinity or consumption of drugs or teratogenic molecules during pregnancy. Dermatological examination revealed micronychia and changes in the lunula of the second right finger, and micronychia, malalignment of the nail, and changes in the lunula of the second left finger (Figure 1). Parents denied any associated malformations or cutaneous and systemic diseases. On the antero-posterior image, radiologic examination revealed a narrowing of the distal phalanx. (Figure 2). The lateral image shows erosion and Y-shaped deformity of the dorsal side of the free margin of the distal phalanx of the second finger and enlargement of the ulnar side (Figure 3).

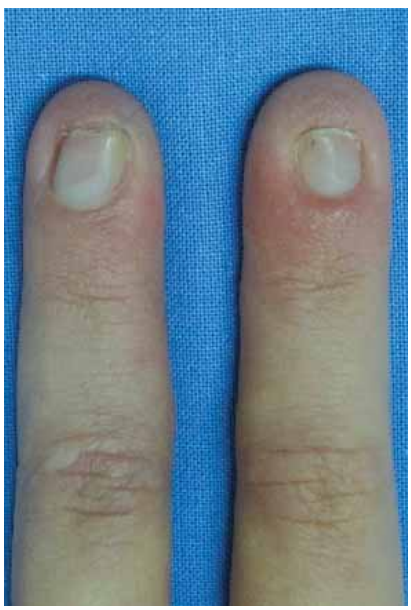

FIGURE 1: Onychodysplasia of the index fingers. Right index finger: micronychia and changes in the lunula. Left index finger: micronychia, malalignment of the nail and changes in the lunula

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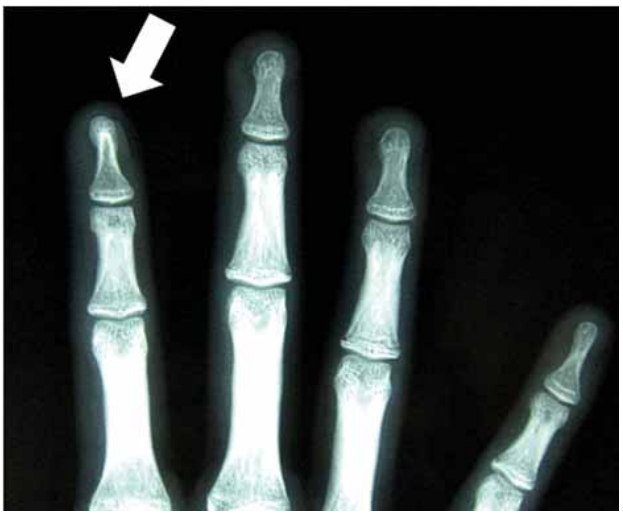

Figure 2: Radiography of the right hand. Narrowing of the distal phalanx of the index finger

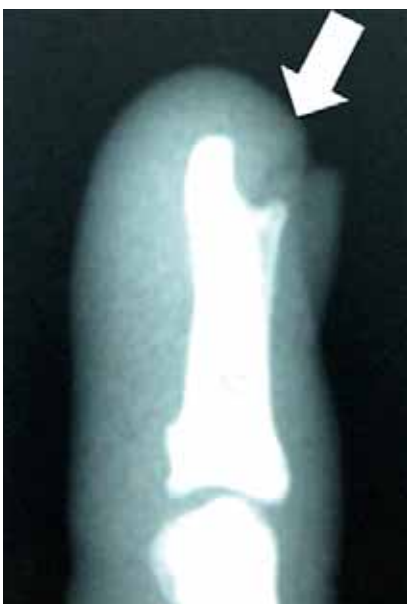

Figure 3: Lateral radiological image of the index finger. Y-shaped image of the distal phalanx

DISCUSSION

Congenital onychodysplasia of the index fingers (COIF), also known as Iso-Kikuchi syndrome, was first described by these researchers in 1969 and 1974, respectively. [1,2] COIF is a rare condition characterized by nail dysplasia involving the index fingers (IF). It affects both sexes, and shows no known predilection for race or ethnicity. The clinical characteristics are, in order of frequency, micronychia (located on the ulnar side), polyonychia (central anonychia and two small incomplete nails on the radial side), anonychia, irregular lunula, malalignment and hemi-onychogryphosis. [3-5]

We have found some case reports in countries like Brazil, Argentina, France, England, Italy, Belgium and India. [5, 6]

Some studies have described theories on the pathogenesis of COIF, attributing it to the use of drugs during pregnancy or to ischemic events located on the radial side of the IF due to direct pressure of the thumbs on the IF of the fetus, which would cause reduction in the diameter of the radial artery of the IF and its consequent abnormal adhesion. However, these theories are controversial.[6] Some studies have also reported associations with several mesodermal malformations and with the autosomal dominant inheritance with variable expression.[7] As the distal phalanx of the IF is a derivative of the mesoderm, it can be understood that a dysplasia of the processus angularis and changes in the membranous ossification at the phalanx would explain associated changes such as brachydactyly, brachymesophalangia, syndactyly, joint malformations, ear abnormalities and inguinal hernia. [6,7]

The antero-posterior radiological image shows hypoplasia and narrowing of the distal phalanx, whereas the lateral radiological image reveals a “Y”-shaped bone projection to the dorsal side. [2,8]

Baran and Stroud suggested the following criteria for the diagnosis of the syndrome: [9]

1 -. Congenital occurrence.
2 -. Unilateral or bilateral index finger involvement.
3 -. Variability in nail appearance.
4 -. Possible hereditary involvement.
5 -. Frequently associated bony abnormalities.

Treatment is limited. Parents should be informed of the need for genetic counseling and possible screening for other potential abnormalities. The hemi-onychogryphosis can be totally or partially corrected with the use of nail orthoses or with surgery. It is necessary to recognize this disease, perform differential diagnosis with ectodermal dysplasias and promote future research lines.❑

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