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Review Article

Orange–Brown Chromonychia, a Novel Finding in Kawasaki Disease: 10 Years Since the First Publication

Abstract

Red/orange–brown transverse nail bed lines in the acute phase of Kawasaki disease (KD) was a new clinical finding reported initially in 2010. Since then, it has been universally and consistently described in the acute phase of KD and may act as an accessory clinical clue in diagnosis. This article tries to review the clinical implication and the various reports citing this finding over the last 10 years.

Keywords: *Chromonychia, Kawasaki disease, nail changes*

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"Eyes are a window to the brain." The nails though somewhat inconspicuous to the clinician, occasionally may act as reflectors of the inner maladies. Although first described by Lindsley.[1] in 1992 as unusual transverse red nail bed lines in four children with Kawasaki Disease (KD), there was no further report for the next 18 years till 2010 when "Transverse Orange–brown chromonychia in Kawasaki Disease"[2] was described in two children in *International Journal of Dermatology* as a case report from the Pediatric Rheumatology Unit of Institute of Child Health, Kolkata. The first major publication on this new entity was in 2012 in "Rheumatology International" from the same unit. "Orange–brown chromonychia, a novel finding in Kawasaki disease"[3] described forty children with KD admitted over 2 years, the nail color change being noted in 29 (72.5%). Since then, it has been described globally and has received twenty citations till date.

Chromonychia literally means colored nails. It is a term used to describe an abnormality in color of the nail plates and/or subungual tissue. Color changes in the nails have been described following the use of antineoplastic drugs (adriamycin, cyclophosphamide, vincristine, or in polychemotherapy),[4] thermal injury, contact exposure to elemental iron, angiotensin receptor blockage, use of nail hardener, and hyperbilirubinemia. Pseudomonas infection has been reported to cause green chromonychia,[5] and AIDS may also cause a change in color significant being with CD4 counts below 200/cu.mm.[6] Connective tissue diseases such as lupus have also been described in this context. However, KD remains the only vasculitic illness causing this characteristic reddish/orange–brown change in color now being described universally[7] with increasing frequency.

The colored lines start appearing usually around the 5th–8th day of illness,[3] although they may be noticed as early as the 4th day of fever mandating it as an early sign in the diagnosis. Once it appears, the coloration stays unchanged over the next 7–10 days. Better appreciated in the fingers [Figures 1-3] than the toes [Figure 4], they start disappearing by the end of the 2nd week. Thus, on appearing, they are present throughout the acute phase of illness as opposed to most of the classical clinical signs in KD, which are known for their markedly temporary appearance.

The exact pathogenic mechanism remains unclear. Capillaroscopy has revealed alteration in nailfold capillaries as a consequence of vasculitis, and it has been proposed that the coloration is due to dense band of microscopic splinter hemorrhages, followed by residual pigmentation as demonstrated by dermoscopy.[8]

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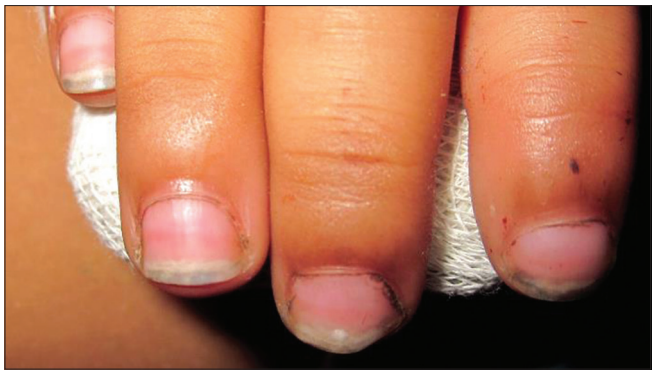

Figure 1: Reddish pink chromonychia of the fingers

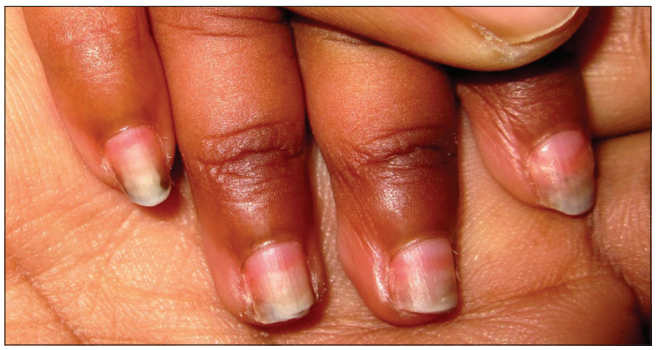

Figure 3: Chromonychia of fingers of some duration

Extremity changes form one of the striking diagnostic features in KD. The erythema of the palms and soles together with edema of the hands and feet which may occasionally be painful often occur in the acute phase. The other classical finding is the presence of periungual desquamation in the fingers and toes, which start around the end of the 2nd week. Although a consistent sign,but because of the late appearance in the subacute phase its clinical utility lies in making a retrospective diagnosis. Another well described albeit a bit rare sign are the transverse grooves (Beau's lines) on the nails which may appear as late as 1–2 months after the disease onset. Beau's lines are better felt than visually appreciated.

Few case reports describe some inconsistent nail changes following KD. Onycholysis has been reported in some cases. Ciastko reported onychomadesis (spontaneous separation of the nail from the matrix) of all twenty nails in an 8-year-old boy with KD, which started 1 week after periungual desquamation.[9] Spontaneously resolving pincer nail deformity (transverse curling of the nail along its longitudinal axis)[10] in an infant with KD and leukonychia partialis[11] (abnormally white proximal portion of the nail) have also been reported. However, all these nail abnormalities are nonspecific, maybe associated with other systemic triggers, and generally resolve spontaneously within 1–2 months.

Since the 2012 publication in rheumatology international, orange/red chromonychia has consistently been described globally. Majority are case reports from countries as

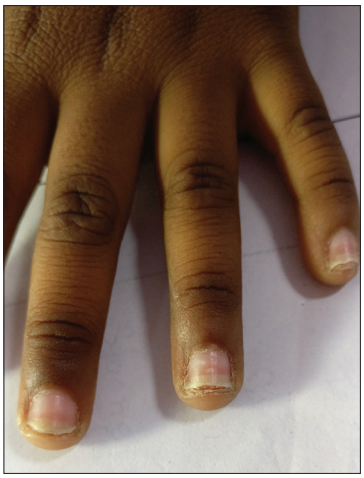

Figure 2: Chromonychia of fingers

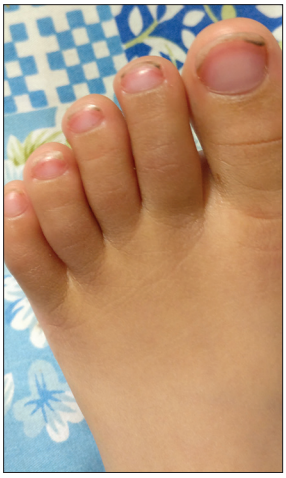

Figure 4: Chromonychia of toes

diverse as the USA, Korea, Australia, Japan, Mexico, and India.[12-14] Authors have described this sign as a novel association and suggested it be considered as an additional clinical sign and a reference[15] in the diagnosis of KD, based on its appearance in the acute phase.

In the recently concluded 12th International KD Symposium at Yokohama, Japan, Pal *et al*.[16] concluded that orange–brown chromonychia is a common clinical finding in KD being present in 63% of the 176 patients admitted between 2012 and 2017 and also the most specific and consistent nail change present in acute KD. They also tried evaluating it for any correlation with coronary artery disease, but no such correlation could be established.

There is an ongoing global search for a specific and consistent biomarker for the early and unambiguous diagnosis of KD. Till that quest is successful, it will be prudent to include this universally described common persistent acute-phase clinical sign as an accessory criteria to aid in diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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