

A 10-year-old Girl with a Melon Seed-like Nail on her Index Finger: A Quiz

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A 10-year-old girl presented to the dermatology clinic with a small-size nail on her right index finger. This condition had been present from birth. Her mother had had gestational diabetes during pregnancy, for which she had not taken any medication. The patient's parents were not biologically related, had no history of drug or alcohol abuse, and no history of any condition similar to the patient's presentation. The patient was in good health, with normal growth and development for her age. She denied having experienced any local damage to her finger, and her right index finger was fully functional. She had a habit of nail biting. Physical examination revealed that the nail plate, which resembled a melon seed, was present only in the ulnar third on the right index finger. On the radial two-thirds of the damaged fingernail, the nail plate, matrix, and apparatus were completely absent. The nail plates on the other fingers were a little short (Fig. 1). Otherwise, patella can be reached, and the teeth, hair, and finger joints were all normal.

What is your diagnosis? See next page for answer.

Differential diagnosis 1: Iso-Kikuchi syndrome

Differential diagnosis 2: Nail-patella syndrome

Differential diagnosis 3: Ectodermal dysplasia

Differential diagnosis 4: Nail bed injuries after trauma

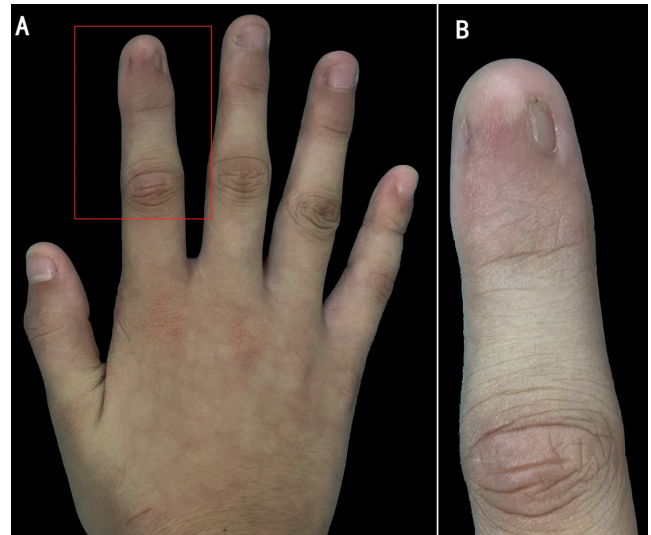


Fig. 1. Physical examination revealed that the nail plate, which looked like a melon seed, was present only on the ulnar third of the right index finger. On the radial two-thirds of the damaged fingernail, the nail plate, matrix, and apparatus were completely absent. The nail plates on the other fingers are a little short.

ANSWERS TO QUIZ

A 10-year-old Girl with a Melon Seed-like Nail on her Index Finger: A Commentary

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Diagnosis: Iso-Kikuchi syndrome

Plain radiographs of the young girl's right hand were obtained. These revealed hypoplastic terminal tuft of the right index finger on the anteroposterior radiograph (Fig. 2A) and a Y-shaped bifurcation at the distal end of the index finger on the lateral radiograph (Fig. 2B). No additional tests, examinations, or pathological biopsies were performed.

Iso-Kikuchi syndrome (IKS) was diagnosed based on the patient's history and clinical characteristics. The patient was not treated, since the condition did not interfere with normal function and neither the patient nor her family wanted cosmetic treatment of her condition.

Iso-Kikuchi syndrome, commonly known as congenital onychodysplasia of the index finger was originally identified by Iso and Kikuchi (1, 2). The incidence rate worldwide is approximately 4.2 cases/100,000 births (3). In China Iso-Kikuchi syndrome is often underdiagnosed; we have described here the second sporadic Chinese case and the first Chinese paediatric case (4)

Several clinical criteria have been proposed: congenital incidence with sporadic or hereditary transmission, unilateral or bilateral involvement of the fingers (mostly the index fingers, although additional fingers and even toes may be affected), variety in nail appearance, and bone deformities (5). Nail abnormalities have a wide variety of forms, including polyonychia, micronychia, anonychia, hemionychocryptosis, misalignment, and lunular deformity. Skeletal abnormalities can be seen on plain radiographs and commonly present as a Y-shaped bifurcation of the distal phalanges; however, they can also appear less frequently as brachydactyly, brachymetatarsi, and missing phalanges (6).

Different pathophysiological theories have been proposed to explain Iso-Kikuchi syndrome, including foetal disorders affecting the palmar digital artery with *in utero* ischaemic changes, *in utero* alterations of the crescent-shaped distal phalanx, and foetal exposure to teratogens, in particular antiepileptic drugs (3, 7).

Surgical procedures might be taken into consideration if they are required for functional reasons or preferred for cosmetic reasons. Understanding this congenital entity, its physical examination, and its radiological appearance is crucial to preventing unnecessary tests and workup. Finally, clinicians should reassure patients that Iso-Kikuchi syndrome is benign and that no related diseases exist.

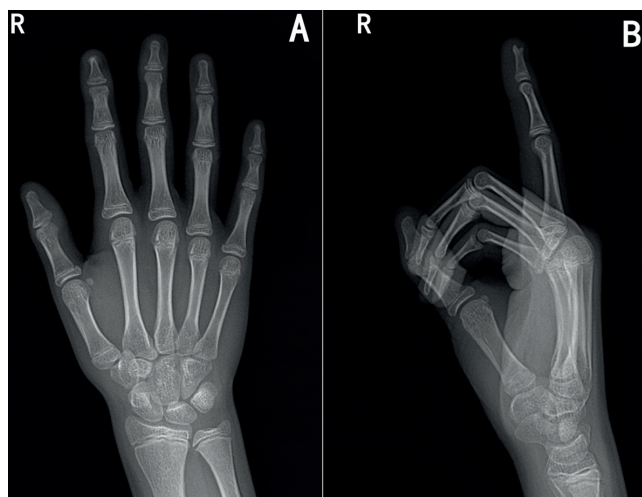


Fig. 2. Plain radiographs of the right hand revealed (A) hypoplastic terminal tuft of the right index finger on the anteroposterior radiograph and (B) a Y-shaped bifurcation at the distal end of the index finger on the lateral radiograph.

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