A 4½ years old female child presented to the department of dermatology with multiple streaks of hyperpigmentation following the lines of Blaschko, all over the body. She was born of a full term vaginal delivery following an uneventful first pregnancy in the 27-year-old mother. The growth and development of the child was normal. Her parents and other family members were normal, and there was no history of consanguinity.

The child remained well for the first 6 weeks of life; but after 6 weeks, the mother noticed dark-colored streaks of linear and whorled pattern developing over the body of the child, involving almost the whole body; the streaks gradually became darker and darker. There was no history of erythema or vesiculobullous lesions. At about 1 year the lesions stabilized in color, and no new lesions developed after that.

Cutaneous examination revealed multiple, brown, hyperpigmented swirls and streaks along the lines of Blaschko, located symmetrically all over the body. The texture of the skin was normal over the streaks. In between the hyperpigmented streaks, the skin was normal. On the abdomen, a whorl type pattern was present; while the legs and arms had linear streaks. On the face, upper eyelids and ears were involved, but palms, soles, eyes, nails, and teeth of the child were normal. Skin biopsy showed diffuse basal cell hyperpigmentation with increase in the number of basal melanocytes. There was no incontinence of pigment, melanophages in dermis or giant melanosomes. The benign nature of the disorder was explained to the patient.

Somatic chromosomal mosaicism may present as isolated pigmentary abnormalities or multiple congenital anomalies with mental retardation.[2-4]

The differential diagnosis includes incontinentia pigmenti, epidermal nevi, and hypomelanosis of Ito. In incontinentia pigmenti,[5] the skin manifestations pass through four successive stages: vesicular, verrucous, whorl- or streak-like hyperpigmentation, and hypopigmented scars. Skin appendages, including hair and teeth, are commonly affected in incontinentia pigmenti with scarring alopecia and peg-shaped teeth. Lack of the above-mentioned features, absence of basal cell degeneration, incontinence of melanin pigment, tissue eosinophilia, and melanophages in the dermis in the skin histopathology ruled out the possibility of incontinentia pigmenti in the present case.

Epidermal nevi are often noticeable during infancy as hyperpigmented streaks along Blaschko's line, which become papillomatous and hyperkeratotic with time. Extensive skin involvement is often associated with skeletal, ocular, and nervous system anomalies. In the absence of histopathologic evidence of hyperkeratosis, acanthosis, elongation of rete ridges, papillomatosis, and occasional evidence of vacuolization, the possibility of epidermolytic hyperkeratosis was not entertained.

The scarcity of the reports prompts the present communication; and to the best of our knowledge, this is the second case reported from India.

The importance of this report lies in the fact that this condition is not only rare but also, there are very few cases of LWH reported in the literature till date.

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Iso-Kikuchi syndrome with absence of ring fingers and metacarpal bone abnormality

Sir,

Iso-Kikuchi syndrome (I-K S), congenital onychodysplasia of the index fingers (COIF), is a rare condition characterized
by various forms of nail dysplasia commonly involving the index fingers. Not infrequently, the neighboring fingers such as the middle fingers and thumbs are also affected.[1] A case of COIF in a 34-year-old female patient is reported here for its rarity.

A 34-year-old female patient, born of a consanguineous marriage, presented with generalized body ache. On examination she was found to have 4 digits bilaterally and absent nail on the right index finger, while the left index finger showed a rudimentary nail. There was micronychia on the middle fingers bilaterally, being more marked on the right side [Figures 1-2]. Lunula was absent in all nails. Palmar creases were normal. She could not flex her right index finger. All toe nails were normal. Her hair and mucous membranes were normal. She had normal dentition. Systemic examination did not reveal any abnormality. An X-ray of both the hands showed normal carpal bones and five metacarpal bones. The phalanges of the ring fingers were absent. The third and fourth metacarpal bones were found articulating with a single large proximal phalanx of the middle finger. Except for an elder brother, who had syndactyly bilaterally, her 3 other siblings were normal. Our patient had 2 children, who were unaffected. The patient gave a history that her mother had consumed an abortifacient while she was in utero.

Ichiro Kikuchi et al.,[2] in 1974, coined the term ‘congenital onychodysplasia of the index fingers’ (COIF) and identified a clinical syndrome consisting of nail dysplasias of the index fingers associated with underlying bone abnormalities. The first case report of this condition was by Kamei. Later, Iso collected a series of patients and defined the clinical syndrome. The term Iso-Kikuchi syndrome was introduced in 1980 by Robert Baran, a French dermatologist in Cannes.[1] Iso-Kikuchi syndrome is rarely reported outside Japan, with an international incidence of 4.2 cases per 100,000 live births.[2]

The nails of COIF include the full spectrum of nail dysplasia, from irregular lunula, malalignment, micronychia (hypoplastic and rudimental), polyonychia (split rudimental), to anonychia, specifically affecting the index fingers. In our patient, there was micronychia of the middle fingers, while micronychia and malalignment of the left second toenail has been described by other authors.[3]

The five criteria characterizing COIF include the following: (i) congenital occurrence; (ii) unilateral or bilateral index finger involvement; (iii) variability in nail appearance; (iv) possible familial involvement; and (v) frequently associated bone abnormalities.[3] Our patient satisfied all the five of the above-mentioned criteria. The configuration of the lunula is supposed to play an important role in shaping the free edge of the nail plate.[4] In our patient, lunula was absent in all nails. Hemi-onychogryphosis of both index fingers and bifurcation of the distal phalanges are the other reported anomalies.[5] Our patient had two metacarpal bones articulating with the proximal phalanx of the middle finger, in addition to the absence of ring finger on both sides. The proximal phalanx of the middle finger was large and could be suggestive of syndactyly. The absence of alopecia and palmoplantar keratoderma and normal dentition ruled out ectodermal dysplasia.

In utero, ischemia of the palmar digital artery and a dysplastic
change in the crescent-shaped cap of the distal phalanx are the two main candidate pathogenetic mechanisms that have been proposed. Exposure to teratogens, especially antiepileptic drugs in utero, in mothers with epoxide hydrolase deficiency is another supposed cause of COIF. This probably could explain our patient’s predicament, as her mother had consumed an abortifacient when our patient was in utero. Due to patient’s noncompliance and lack of facilities, arteriographic studies could not be undertaken. Iso-Kikuchi syndrome has also been associated with discoid lupus erythematous. As there were filiform arteries of the fingers and slow blood circulation on angiographic studies, the authors considered vascular pathogenic mechanisms to be responsible for this syndrome. However, there was no clinical or laboratory evidence of any connective tissue disease in our patient.

Transmission of COIF can be either hereditary as autosomal dominant or sporadic. In our patient, a positive family history involving her brother could be suggestive of some hereditary involvement. Due to unavailability, chromosomal studies could not be undertaken.

Since COIF is only of cosmetic significance and has not interfered with her day-to-day activities, our patient was reassured and advised physiotherapy for the limitation of movements of the fingers.

The present case is being reported for its rarity and the hitherto unreported abnormality of the metacarpal bones, in addition to the absence of one digit (ring finger) on both sides.

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Eumycetoma due to Curvularia lunata
Sir,
Mycetoma is a chronic granulomatous, suppurative, and progressive inflammatory disease that usually involves the subcutaneous tissue and bones after traumatic inoculation of the causative organism. The condition may be caused by true fungi or by higher bacteria and therefore is classified as eumycetoma or actinomycetoma respectively.[1] It is mainly seen in Africa, India, Mexico, and parts of South America. In India actinomycotic mycetoma is prevalent in south India, southeast Rajasthan, and Chandigarh; while eumycetoma, which constitutes one third of the total cases, is mainly reported from north India and central Rajasthan.[2] The common etiological agents of eumycetoma reported from different centers are Madurella mycetomatis, M. grisea, Acremonum spp., Aspergillus spp. and Fusarium spp.[3] We report here a rare instance of eumycetoma caused by Curvularia lunata in a 65-year-old male farmer, who presented to the dermatology outpatient clinic of our hospital in September 2007, with swelling of right foot, multiple nodules, and sinuses discharging black-colored granules. His problem started 6 years back as a single nodular swelling on the plantar surface of the foot following trauma. After a few months, painless multiple nodules developed on both plantar and dorsal surfaces of the foot. Some of the nodules broke down, forming openings discharging black-colored granules. Physical examination of the patient revealed non-tender, gross swelling of the right foot with multiple discharging...