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Photo Vignette

Congenital onychodysplasia of the index finger presenting as a congenital bifid nail

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Abstract

Congenital onychodysplasia of the index finger (COIF) is a rare disorder characterized by various nail dystrophies, often with underlying bony deformity. We report a case of a COIF presenting as a congenital bifid nail deformity with underlying bone deformity.

Case synopsis

A healthy 15 year-old girl presented with a lifelong history of a nail abnormality of the left index finger. The patient denied any previous trauma to the nail unit. There was no family history of similar nail changes. On examination, the left index finger had two separate nail matrices and symmetric nail plates, each with corresponding lateral nail folds, separated by a fibrous band (Figure 1). The remaining nails on both hands and feet were normal in appearance. No other dental or muscular deformities were noted. The patient had full range of motion in the digit. Radiographic examination of the left fingers demonstrated a deformity of the distal tuft of the distal phalanx of the index finger with a “Y-shaped configuration” on the lateral view (Figure 2).

Figure 1. Left index finger with two separate nail matrices and nail plates separated with a fibrous band

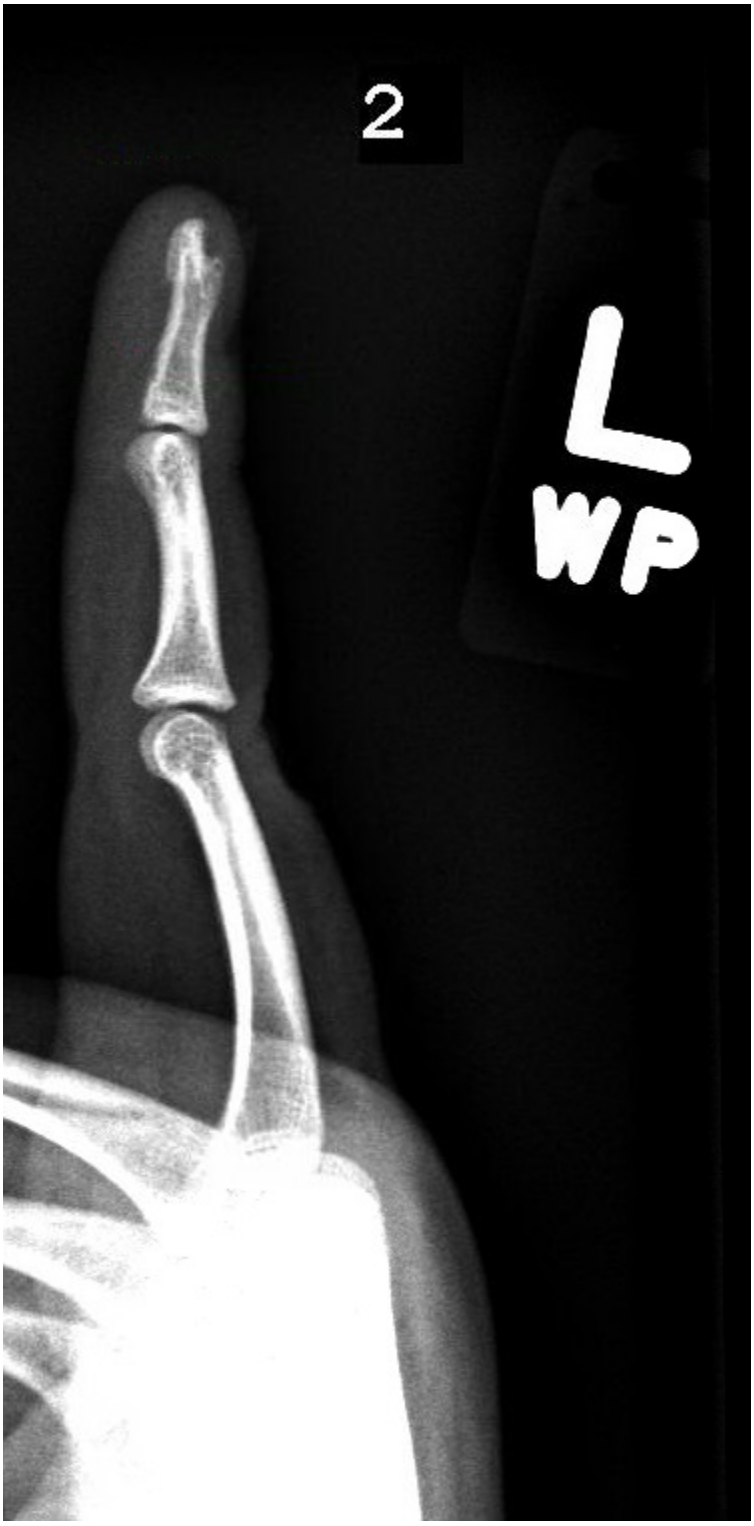

Figure 2. There is deformity of the distal tuft of the distal phalanx of the index finger with a "Y shaped configuration" on lateral view. This corresponds to the congenital onychodystrophy seen clinically. No other abnormality is seen.

Discussion

Congenital onychodysplasia of the index finger (COIF) is a rare disorder characterized by various nail dystrophies, often with underlying bony deformity. The first case was described in 1969 by Iso, who proposed five diagnostic criteria: congenital occurrence, bilateral involvement of the index fingers, variability in nail appearance (micronychia, polyonchia, anonychia), nonfamilial, and no associated underlying bone of joint abnormalities [1]. After the first description in the literature, reports were published presenting bony abnormalities, unilateral involvement, and a familial occurrence. In 1984 Baran and Stroud revised the criteria as follows: congenital, unilateral or bilateral, variability in nail appearance, and possible hereditary transmission [2]. A

full spectrum of nail dystrophies has been reported in association of COIF [2,3]. In cases in which unilateral involvement is present deformities tend to be left-sided, and the deformities more pronounced on the radial rather than ulnar aspect of the digit. Our patient's deformity was present on the left hand, but the nail dystrophy was a symmetric bifid nail [3]. A bifid nail refers to the presence of two nail units on the same finger, delineated by intervening soft tissue. The most common underlying bony defects include narrowing, crescent shaped cap, and a Y-shaped deformity of the distal phalanx. Theories proposed to explain the pathogenesis revolve around defects in the underlying ossification of the phalanx, perhaps with a vascular infarct, that then produces secondary deformities of the overlying ectoderm [4].

Conservative management includes close clipping to prevent irritation or pain. Surgical procedures are generally not recommended owing to the minimal functional impairment caused by COIF [4]. After careful consideration taking into account the potentially poor cosmetic outcome of the procedure, surgical excision was declined. It was decided that the optimal treatment for this patient would be application of an adhesive artificial nail to both nail segments, covering the intervening fibrous band and providing an acceptable cosmetic and functional outcome.

References

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