Painless Swelling of Proximal Interphalangeal Joints in an Adolescent Child With Preserved Function and Normal Acute Phase Reactants

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A previously healthy 12-year-old male patient presented with one-year history of bilateral (left more than right) swelling of the proximal interphalangeal (PIP) joints (Figure 1). The patient denied any tenderness with movement, nocturnal pain, or morning stiffness. The only findings on physical examination were swelling of the PIPs of the fingers. Laboratory investigations including complete blood count with differentials, erythrocyte sedimentation rate, rheumatoid factor, and antinuclear antibodies were normal. Hand radiographs revealed nonspecific swelling of the volar soft tissues abutting the distal metaphysis of the proximal phalanges of the second to fifth fingers (Figure 2). The patient was diagnosed with Thiemann’s disease (Online Mendelian Inheritance in Man: 165700).

Figure 1. Swelling of patient’s digits.

Figure 2. Hand X-ray showing nonspecific edema in volar soft tissues of proximal phalanges of second to fifth fingers.
First described in 1909, there has been 32 cases of Thiemann’s disease reported in the literature characterized by clinical and radiological hand and foot epiphyseal abnormalities. Handa et al. proposed that Thiemann’s disease appears before the age of 25 with PIP joint swelling and absence of elevated acute phase reactants. Radiologically, Thiemann’s disease is characterized by fragmentation and broadening of the basal phalangeal epiphyses, which can be followed by joint space narrowing, premature physeal fusion, and phalangeal shortening. Clinicians should be aware of Thiemann’s disease whenever an adolescent presents with painless swelling of the digits. An investigation involving radiograph and laboratory tests is crucial to eliminate possible differentials.

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