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A Rare Finding of Onychoheterotopia (Ectopic Nail) in a Case of Systemic Sclerosis

Kailash Bhatia[1], Rajesh Kataria[1], Zainab Hussain Safderi[1] and Ravindra Kumar[2]

[1]Department of Skin and VD, Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh, India. [2]Central Research Laboratory, Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh, India.

ABSTRACT: Systemic sclerosis and ectopic nail are two rare disorders. Here, we are describing a unique case in which both the disorders are presented simultaneously.

KEYWORDS: systemic sclerosis, onychoheterotopia

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CORRESPONDENCE: drbhatiakailash@yahoo.co.in

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Introduction

Systemic sclerosis is a multisystem disorder characterized by overproduction and accumulation of collagen and other extracellular matrix proteins resulting in skin induration and thickening and fibrosis. Chronic inflammatory infiltration of internal organs, microvascular damage and dysfunction, and immune dysfunction are other complications.[1] Systemic involvement may occur in the form of pulmonary vasculopathy, interstitial fibrosis, myocardiopathy, arrhythmia, conduction abnormality, acute renal crisis, lower esophageal incompetence, etc.[2]

Ectopic nail is another extremely rare condition related to acquired or congenital anomalies. It is characterized by development of nail-like tissue in a location other than the nail bed.[3]

Here, we are presenting a case of incidental findings of ectopic nail in a patient with systemic sclerosis.

Case

A 45 year-old woman presented to us with a complaint of dyspnea and difficulty in swallowing since last six months. The patient was apparently normal three years before when she started having pain and burning over her digits of the left hand. The patient noticed the color changes of white, blue, and red on exposure to cold water, cold temperature accompanied by pain and numbness. Similar changes started occurring in the right hand also since four to six months. Following this, the patient noticed swelling of both hands, which gradually subsidized over the months and now has stiffness and contractures of a finger joint. There was thickening of skin since two years, which was first noticed over the hands and then over the face and trunk. She had no history of hypertension, diabetes mellitus, asthma, and tuberculosis. There was no digital ulcers, calcinosis, renal, cardiac, and esophageal involvement.

Clinically, she was pallor and had a mask like facial appearance with skin adherent to underlying tissue (Fig. 1A). She had claw hand deformity on both the hands with decreased flexibility of joints and tendons and inability to flattened hand. Skin thickening, sclerodactyly, thinning of nose with beak like appearance, radial furrows around the mouth, thinning of upper lip, microstomia, and mild telangiectasia were other complications that were observed clinically in the patient.

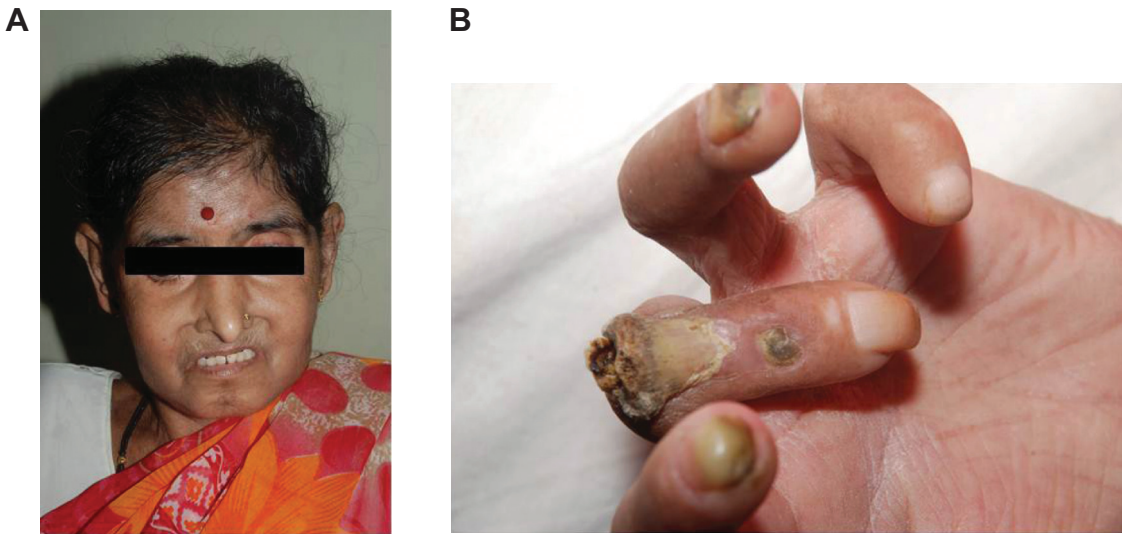

Figure 1. Clinical picture of patient (**A**) and picture of the left hand showing ectopic nail (**B**).

Laboratory examination revealed Hb = 9.3 gm/dL, TLC = 13900/μL with 92% of neutrophil count. Anti-nuclear antibody was found negative. Lung spirometry test showed the moderate restrictive airways pattern. X-ray chest showed the thickened horizontal fissure and reticulonodular shadow on right mid and bilateral lower zone (Fig. 2A). All these observation made us to diagnose the patient as a case of systemic sclerosis.

We found a hard keratotic translucent and inflexible mass at level of PIP joint on left middle finger, appearing as nail with horizontal growth pattern, growing parallel to underlying proximal phalanx, and measured about 6 mm × 5 mm (Fig. 1b). Circumferential ridges were absent, which excludes the diagnosis of cutaneous horn. X-ray examination of hand shows resorption of tufts of terminal phalanx of right thumb, second and fourth fingers. Flexion deformity of pip joint of finger was also seen (Fig. 2B). Histopathological examination of hard keratotic mass at the level of PIP joint on left middle finger revealed it as ectopic nails.

We advised the surgical management of ectopic nail but the patient did not give her consent for the surgery. The patient was discharged after seven days with symptomatic treatment.

Discussion

Systemic sclerosis is a rare chronic connective tissue disease of unknown etiology that can affect many organs and systems. Morbidity and mortality depend on the organ or system involved. Its prevalence varies greatly according to geographic location and disease definition, with estimated prevalence between 3 and 24 per 100,000 inhabitants, higher in North America and Australia compared to Europe and Japan.[4] There is no literature about the prevalence of systemic sclerosis in India.

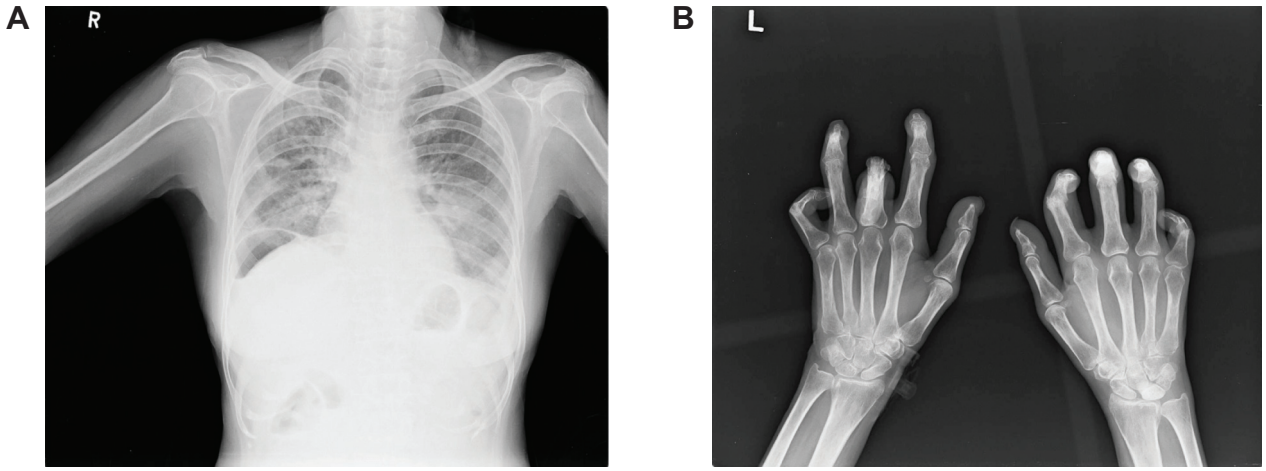

Figure 2. X-ray of the chest (**A**) and hand (**B**).

Ectopic nail is an extremely rare condition and defined as tissue noted in a different location other than in the normal nail bed. It is an extremely rare condition with less than 50 reported cases in the literature.[5] It has been broadly classified as congenital and acquired, the majority of the reported cases being of the congenital type. The pathogenesis of both types remains unclear.[6] Various hypotheses have been made to explain this anomaly such as presence of ectopic germ cells, teratoma, dyskeratotic skin growing into a nail, visible portion of the nail plate of occult polydactyly, inherited syndromes like Pierre Robin syndrome, or congenital palmar nail syndrome.[7] Sometimes ectopic nail may develop as posttraumatic complication.[8] In our case there was no history of trauma.

Our case is a very rare case presenting with two rare entities that are ectopic nail and systemic sclerosis simultaneously.

Author Contributions

R Kumar wrote the first draft of manuscript. R Kataria and ZHS jointly developed the structure and arguments for the paper. KB made critical revision and approved the final version. All authors reviewed and approved the manuscript.

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