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Case Report

Raynaud's phenomenon manifesting as progressive abnormal MRI bone marrow signal in the toes

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ABSTRACT

Objective: The purpose of this report is to discuss the presentation and the progressive magnetic resonance imaging (MRI) findings in a single patient with clinically-diagnosed Raynaud's phenomenon (RP).

Conclusion: RP can present as non-specific toe pain and manifest as progressive abnormal MRI bone marrow signal in the toes. In addition to patient presentation and clinical assessment, this information could contribute to earlier diagnosis and treatment of RP and other coexisting rheumatologic disorders.

Introduction

Raynaud's phenomenon (RP) is an episodic and recurrent disorder that involves microvascular vasospasm of digital arteries and subcutaneous arterioles in response to cold temperatures or emotional stress [1]. Clinically, patients often describe acute well-demarcated skin color changes of the fingers and toes in a triphasic pattern of pallor, cyanosis, and rubor. These color changes reflect stages of vasospasm including hypoperfusion, hypoxia, and reperfusion, respectively. Associated sensory symptoms are paresthesia, pain, and burning sensations. While the fingers and toes are most commonly affected, RP may also involve the ears, nose, nipples, and tongue [1–3].

Raynaud's disease was initially used to describe these abnormal vascular presentations until it was proposed that multiple etiologic factors could be responsible, thus introducing the concept of RP [4]. While results can vary according to sex, climate, and work exposures, most population-based

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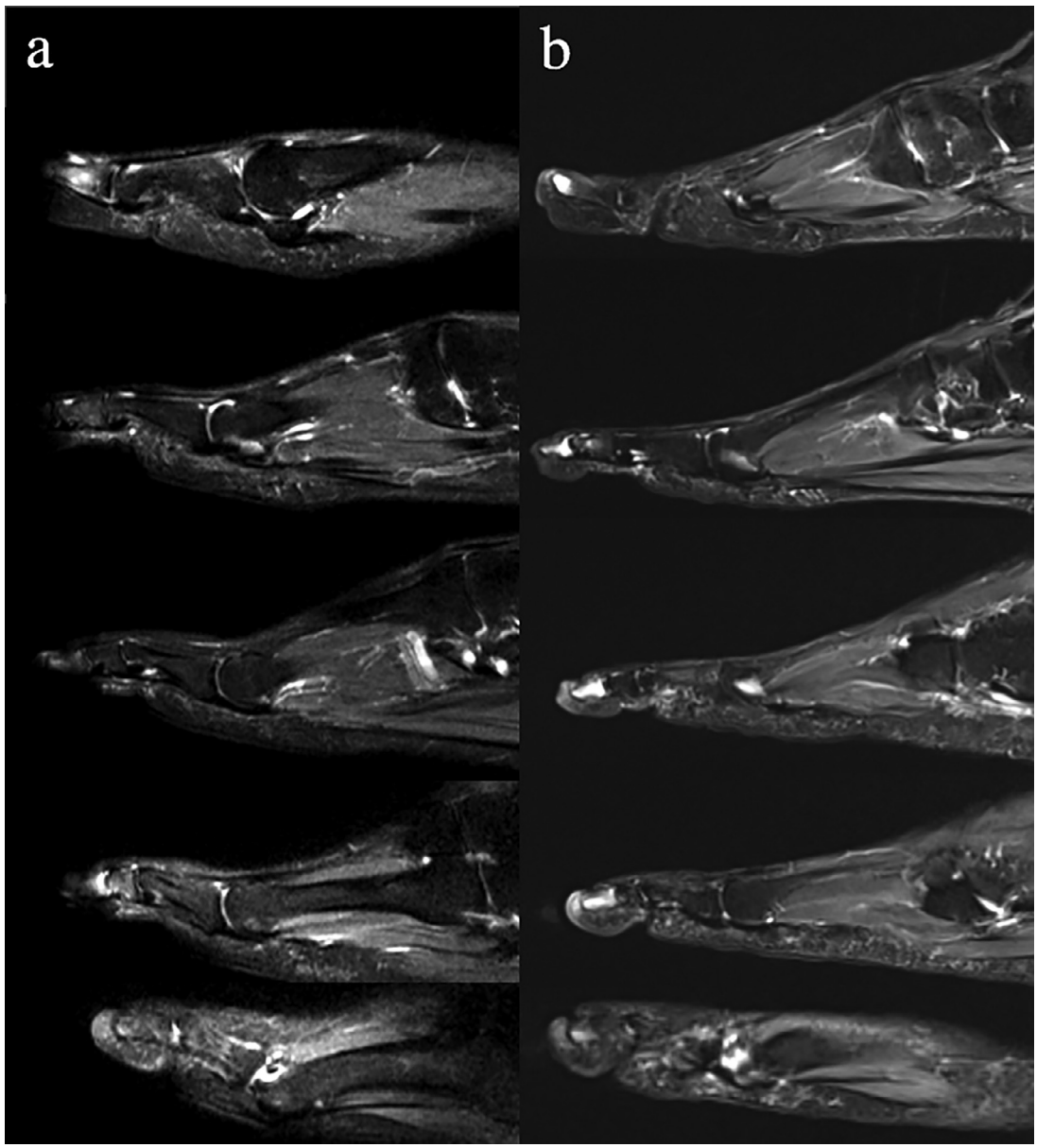

Fig. 1 – Initial (a) and 2.5-year follow-up (b) MRI STIR images of the right foot demonstrate progressive distal-to-proximal hyperintense signal abnormalities in the distal phalanges. Toes are arranged from first (top) to fifth (bottom).

surveys estimate a RP prevalence of 3%-5% [5–7]. Other associations with RP include connective tissue disease in 2%-6% and rheumatologic disorders in 10%-20% [3,8].

Given that RP is predominantly a clinical diagnosis, there is limited literature discussion of the imaging findings [9]. Additionally, there is no literature that provides follow-up imaging of the progression or resolution of RP involving the pedal distal phalanges. While radiologic modalities such as conventional angiography, magnetic resonance angiography, Doppler imaging, infrared thermography, and digital blood flow scintigraphy have supported the diagnosis, phalangeal bone marrow edema on magnetic resonance imaging (MRI) has also been observed in a small subset of patients [9–15]. As most cases lack radiologic correlation to the clinical diagnosis of RP, the purpose of this case report is to describe the progressive abnormal pedal distal phalanges MRI findings in a patient with known RP.

Case report

A 43-year-old Caucasian female with a significant past medical history of RP predominantly affecting the hands presented to an orthopedic surgery foot and ankle specialty clinic for chronic right great toe pain for 3 years after being stepped on. She had also been experiencing medial plantar pain with ambulation and constant generalized toe and medial forefoot pain at rest. Her symptoms were somewhat relieved by

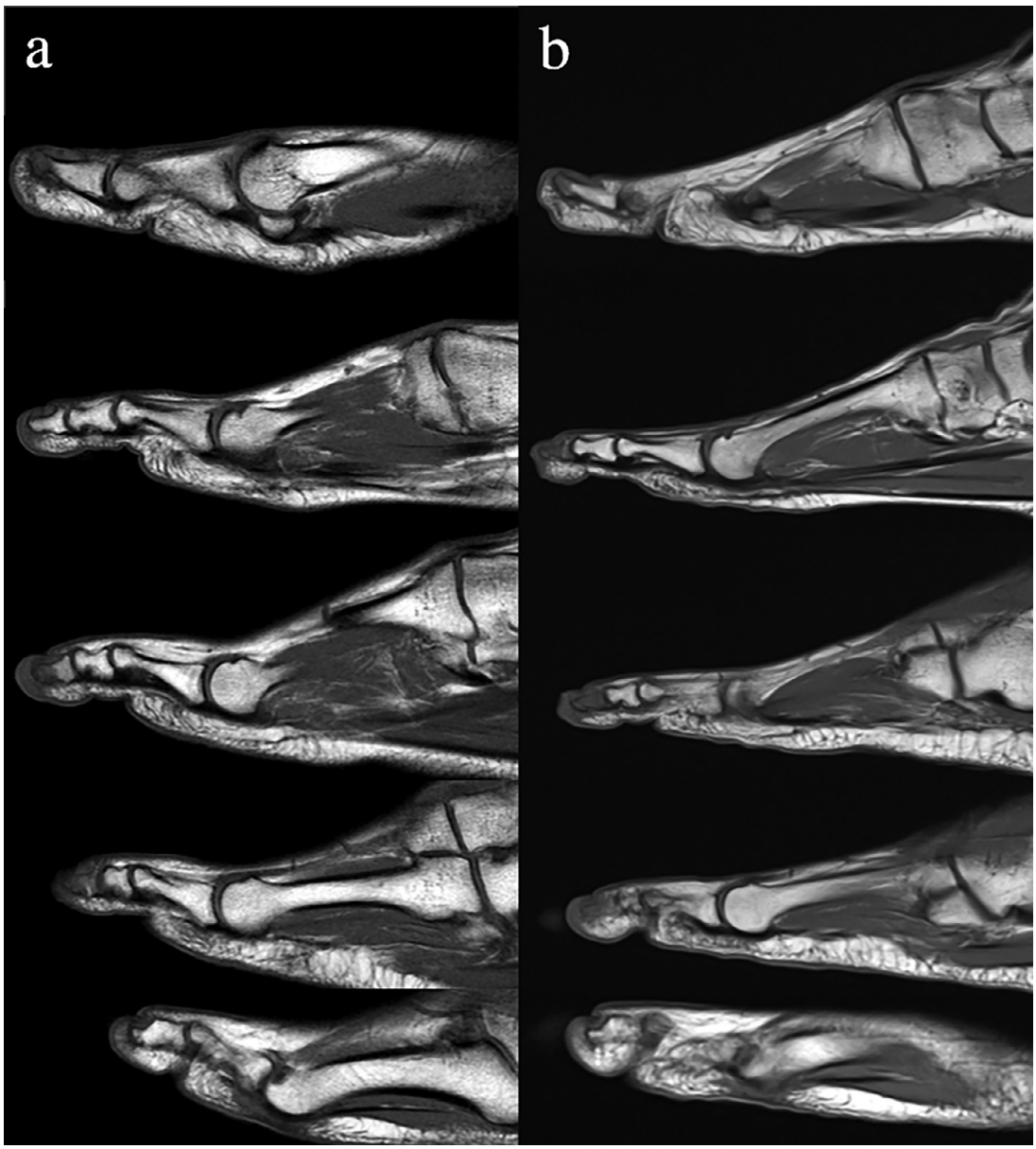

Fig. 2 – Initial (a) and 2.5-year follow-up (b) T1-weighted MRI images of the right foot demonstrate progressive distal-to-proximal hypointense signal abnormalities of the distal phalanges. Toes are arranged from first (top) to fifth (bottom).

physical therapy and acupuncture, but cortisone treatments provided no improvement. The patient refused neuropathic pain medication options. Physical examination of the right foot was significant for mild hallux valgus and tenderness to palpation of the medial first proximal phalanx.

Initial imaging demonstrated negative standing right foot x-rays and hyperintense short-T1 inversion recovery (Fig. 1a) and hypointense T1 (Fig. 2a) signal abnormalities within the distal first, third, and fourth phalanges on a noncontrast right foot MRI. Follow-up noncontrast right foot MRI 2.5 years later was significant for progressive diffuse signal abnormalities of the first through fifth distal phalanges (Figs. 1b and 2b). There was also a 4 mm ganglion cyst noted medial to the right first metatarsophalangeal joint.

At the next follow-up clinic appointment, imaging findings were formally discussed. The patient specifically expressed continued localized pain greatest at the right great toe, and goals were established to continue conservative management through an exercise program. If the pain persisted, the patient was recommended to schedule a diagnostic injection of the right plantar medial halluceal nerve branch.

Discussion

RP is a highly-localized condition that affects arterial inflow to specific distal regions of skin. These sites are distinct from

other skin areas in that they contain high-density arteriovenous anastomoses which function as thermoregulatory structures. Cold temperatures can induce a sympathetic response that causes reflex cutaneous vasoconstriction, and this response can be amplified in people with RP [16]. While this pathogenesis is not completely understood, authors have proposed causes associated with overproduction of endothelin-1, enhanced platelet aggregation, a calcitonin gene-related peptide deficiency, altered blood viscosity, and augmented response alpha-2 sympathetic receptors [17].

Smitaman et al. described identical MRI findings where an observational cohort of 6 patients with RP demonstrated pedal phalangeal bone marrow edema [9]. Signal abnormalities in their study were most evident in the first through fourth toes and displayed a progressive distal-to-proximal distribution between patients. In our case, initial imaging demonstrated short-T1 inversion recovery hyperintense/T1 hypointense signal abnormalities predominantly within the distal first, third, and fourth pedal phalanges. Follow-up noncontrast right foot MRI 2.5 years later demonstrated progressive abnormal MRI bone marrow signal in the toes. In addition to the observational cohort study findings, the distribution pattern on follow-up imaging suggests a chronic progression pattern and supports the theory that terminal perfusion zones regulated by arteriovenous anastomoses may be more prone to vasospasm [18].

It is not conclusive whether these alterations in signal intensity are potentially reversible. These MRI findings also do not establish a disease severity. Distal phalangeal bone marrow edema-like changes can be nonspecific and are also demonstrated in a wide variety of diseases including fractures, crush injuries, insect stings or bites, osteomyelitis, serous bone marrow transformation pathologies, and seronegative spondyloarthropathies [19–26]. Other limitations of this case include presentation confirmation in correlation to known disease and lack of histopathologic correlation. The patient had a known history of RP predominantly affecting the hands but was presenting for medial plantar pain with ambulation and constant generalized toe and medial forefoot pain at rest. Pain was most pronounced at the right great toe following trauma, which corresponded to a 4 mm ganglion cyst medial to the right first metatarsophalangeal joint. Additionally, this likely represents an incidental presentation of RP as the etiology for the patient's bone marrow edema demonstrates a suspected chronic progression pattern.

Our report provides the first follow-up imaging case of progressive RP manifesting as abnormal MRI bone marrow signal in the toes. In addition to history of presentation and clinical assessment, this information could contribute to earlier diagnosis and treatment of RP and associated rheumatologic disorders. Future case reports and studies could collectively provide greater insight into condition progression or resolution in the acute and chronic stages.

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