Phalangeal microgeodic syndrome: A case series in five adults

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ABSTRACT

Phalangeal Microgeodic Syndrome (PMS) is rare, characterised by cold related discolouration, pain and swelling of the digits. A handful of case reports exist mainly in a paediatric population in Japan. We describe five cases of PMS. The underlying aetiology remains uncertain, however, a possible correlation with infection or autoimmune dysregulation may suggest a common pathway which is yet to be identified. However, many cases seem to resolve spontaneously.

Keywords: Phalangeal Microgeodic Syndrome, Raynaud's phenomenon, paediatrics, aetiology

Introduction

Phalangeal microgeodic syndrome (PMS) is rare, characterised by cold related discolouration, pain and swelling of the digits. A handful of case reports exist mainly in a paediatric population in Japan.[1]

PMS appears to be distinct from Raynaud's phenomenon (RP). In both conditions, MR imaging demonstrates abnormal bone marrow signal.[2,3] In primary RP, signal abnormality starts at the distal phalangeal tuft, progressing proximally.[4] However, signal abnormality in PMS preferentially affects the middle and proximal phalanges.[5]

PMS is largely described as an isolated condition, although a single case of PMS in a patient with existing systemic lupus erythematosus (SLE) has been published.[6]

Case reports

We describe five cases of PMS in this paper. Written informed consent was obtained for all cases.

Case 1

A 24-year-old Caucasian female developed dusky red discolouration of the distal fingers with episodic tenderness and swelling during the Winter of 2020. Cardiolipin IgG antibody was just raised at 10GPLU (normal range: 0.0-9.9), but normalised on repeat testing. MRI scan demonstrated bone marrow oedema in the middle phalanx of both middle fingers and left middle finger distal phalanx on the right with superficial soft tissue oedema. By July 2021 the symptoms had improved spontaneously. (See Figure 1)

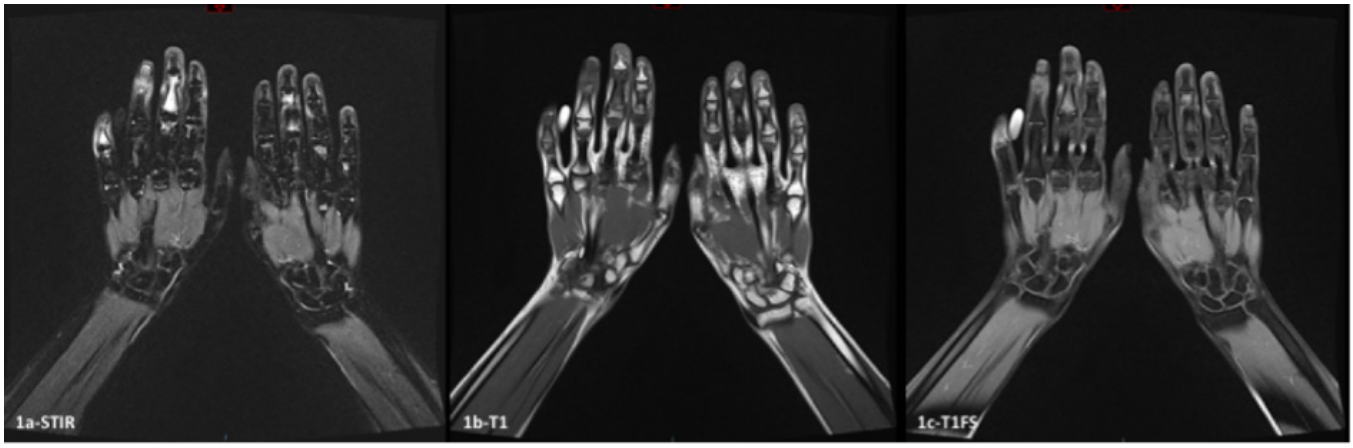

Figure 1a-c: MR Image both hands (1a Cor STIR, 1b Cor T1w 1c Cor T1FS post Gadolinium)
There is high signal in middle phalanx of both middle fingers and left little finger with similar changes in the distal phalanx of the left little finger. Corresponding decreased signal in the T1w sequence and mild enhancement on the T1FS. Surrounding soft tissue oedema also present.

Case 2

A 79-year-old Caucasian female presented with new onset pain, redness and swelling centred around the proximal interphalangeal (PIP) joints of both hands. Symptoms started two days after receiving the first dose of Pfizer/BioNTech RNA vaccine against Covid-19 in January 2021. Rheumatoid factor (RF) was marginally raised at 17 IU/ml (normal: <14IU/ml). MRI scan of both hands demonstrated bone marrow oedema in the proximal, middle and distal phalanges of all the fingers. Symptoms resolved after six weeks, with no significant adverse reaction to the second dose of Covid-19 vaccine.

Case 3

A 60-year-old Caucasian female presented in January 2021 with recurrent pain and swelling in the fingers during the winter for three years, associated with new onset biphasic RP, but no other features of connective tissue disease (CTD). Immunological tests were negative. Musculoskeletal ultrasound excluded synovitis. MRI showed bone marrow oedema affecting the proximal phalanges of both index fingers, proximal and middle phalanges of the left middle and index fingers and all phalanges of the little finger on the right. A trial of amlodipine had no impact on symptoms and was discontinued.

Case 4

A 40-year-old Korean female with SLE (histologically proven cutaneous involvement, ANA positive, La antibody positive 30U/ml, no history of RP) on maintenance hydroxychloroquine and methotrexate developed acute pain, swelling, and redness of the right distal little finger in December 2020. MRI of the right hand in January 2021 showed bone marrow oedema in the right index to small digits with no synovitis. A course of prednisolone was prescribed and methotrexate dose increased (from 7.5mg to 12.5mg weekly), which had no impact on symptoms. 60mg intravenous (IV) pamidronate was given in February 2021. In March, symptoms were only modestly better so modified release nifedipine 10mg once daily was issued. By May 2021, the right-hand symptoms had improved but new symptoms had developed on the left.

Repeat MRI June 2021 showed improvement in appearances. A further 60mg IV pamidronate was given in November 2021, followed by a significant improvement in the hand symptoms four weeks later.

Case 5

A 36-year-old Caucasian female developed pain and purple discolouration of the toes in November 2020. In January 2021, she had similar symptoms in the fingers of both hands. Musculoskeletal ultrasound excluded synovitis. MRI scan of the right hand confirmed patchy bone marrow oedema in all the phalanges excluding the thumb. By July symptoms had improved spontaneously. Just prior to symptom onset, the patient had a confirmed exposure to Covid-19 and had to self-isolate although no infection was confirmed.

Discussion

In these authors' experience, it would appear the incidence of PMS is increasing, which may relate to the especially cold winter of 2020/21. The patient with co-existing SLE appeared to have more refractory disease with persistent symptoms despite increasing seasonal temperatures. Management is conservative with avoidance of cold exposure being paramount, similar to Raynaud's phenomenon.

One patient developed symptoms two days after receiving the first dose of Pfizer/BioNTech RNA vaccine against Covid-19. One patient had to self-isolate following a Covid-19 contact and developed symptoms of PMS shortly thereafter, although there was no proven infection in the affected individual. Chilblain-like acral lesions have been associated with Covid-19 infection, although evidence is conflicting.[7,8,9] One case report associates chilblains and the Moderna vaccine.[10] Any association between Covid-19 infection or vaccine and PMS is purely hypothetical and would require more study to identify any true relationship. The underlying aetiology remains uncertain, however, a possible correlation with infection or autoimmune dysregulation may suggest a common pathway which is yet to be identified.

Conclusion

PMS is a rare condition with an uncertain aetiology although there are pointers to a possible association with infection and / immunological disturbance. Many cases seem to resolve spontaneously.

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