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Pediatric Imaging *Case report*

Phalangeal microgeodic syndrome, COVID-19, and antinuclear antibodies in a child: A case report

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ABSTRACT

Phalangeal microgeodic syndrome (PMS) is a rare rheumatological disease affecting children strongly associated with cold weather. It is considered to be benign and self-limiting, with most cases resolving in warmer months, and no studies have investigated its association to autoantibodies or viral infection. In this peculiar case, a 12-year-old child with the previous COVID-19 infection developed PMS that did not improve in the warm weather and, subsequently, Raynaud's phenomenon and facial rash. With the increasing number of new studies showing the correlation between COVID-19 and autoimmunity, this case report highlights a plausible link between COVID-19, PMS, and autoimmunity in the pediatric population that should be investigated further.

Keywords: Phalangeal microgeodic syndrome, COVID-19, Antinuclear antibodies

INTRODUCTION

Phalangeal microgeodic syndrome (PMS) is a rare and benign rheumatological disease affecting children. It is strongly associated with cold weather and not linked to autoantibodies or viral infection. We present a peculiar case of a child with the previous COVID-19 infection who developed PMS that did not improve in the warm weather and, subsequently, Raynaud's phenomenon and facial rash.

CASE REPORT

A 12-year-old girl presented in the winter season with persistent swelling and pain in the right 1st and 2nd fingers with overlying red rash for a few weeks. There was subsequent involvement of the 3rd and 4th fingers with boggy swelling, and pain over the proximal and distal interphalangeal joints (PIPs and DIPs). She did not have any systemic symptoms. Her background included seborrheic dermatitis, asthma, atopic dermatitis, food allergies, previous Streptococcal pharyngitis, and developmental dysplasia of the hip. At that point, she had not experienced Raynaud's phenomenon. She had COVID-19 6 months previously. Her inflammatory markers, liver and thyroid function tests, were normal. She had an autoimmune screen which showed weak positive antinuclear antibodies (ANA) with speckled pattern. Extractable nuclear antigen, cyclic citrullinated peptide, and double-strand DNA antibodies were negative.

While plain radiograph of both hands was normal, magnetic resonance (MR) of the right hand showed diffuse marrow edema within the middle phalanges of all fingers as well as the distal

phalanx of the thumb and the head of the proximal phalanx of the index finger. There were no joint effusions or evidence of synovitis to suggest an inflammatory arthropathy [Figure 1].

On follow-up 3 months later, the pain had subsided; however, the fingers swelling had worsened. In addition, Raynaud phenomenon triggered by cold as well as eye dryness and photosensitive rash were noted.

DISCUSSION

PMS, first described by Maroteaux in 1970, is a rare disease which presents with frostbite-like symptoms such as spindle-shaped swelling and mild pain in one or more fingers and, more rarely, toes. Moreover, skin redness on the affected fingers might be present. It predominantly affects children and the most common sites are the middle phalanx of index and middle finger. While the etiology is unknown, transient peripheral circulatory impairment due to exposure to cold temperatures is thought to be a major precipitating factor.[1-4]

The cases described in the literature did not present with systemic signs of illness.[2] In line with that, our patient presented in the winter, with swelling, tenderness, and redness of the first four fingers of the right hand.

As PMS develops mainly in children and most often at the metaphyses of the phalanges, it is speculated that fast-growing phalanges are vulnerable to cold injury.[1,5] In 2018, Yang *et al*. analyzed 20 cases of PMS, leading to the conclusion that the disease may develop as a result of bone absorption and destruction in response to vasculopathy caused by chilblain, with impairment of the phalangeal blood circulation due to high pressure in the subcutaneous and subaponeurotic space. The distal phalanges are thought to be spared due to their richer vascularity and absence of the deep fascias.[2] This was also present in our case, where MR findings showed more significant inflammation of the proximal and middle phalanges.

Radiological findings present in the literature include multiple small, round, and well-defined osteolytic areas in the medulla and cortex, described as small lacunae with sclerotic margins and periosteal reaction in the affected phalanges as well as fusiform soft-tissue swelling. Lesions were found to be larger in proximal metaphyses.[1,2] MR images are represented by marrow edema and osteonecrosis with low signal intensity on T1-weighted images and high signal intensity on fat-saturated T2-weighted images. Contrast-enhanced T1-weighted images exhibit marked enhancement in the affected phalanges. Soft-tissue swelling was also observed.[2] Importantly, MR findings show more diffuse skeletal involvement than that visible on the skin and radiographs.[1,3,6] This is consistent with our case, where MR showed diffuse phalangeal and metaphysis involvement, while radiograph images were unremarkable. Our patient had clinical signs affecting only the first four fingers; however, marrow edema was present within all first phalanges of that hand.

PMS has a good prognosis with spontaneous regression within 2–3 months.[1-4] It is therefore important to be aware of this differential diagnosis while assessing young patients presenting with finger swelling and no other systemic complaints and make sure to follow them up in few months. Sometimes, PMS has been misinterpreted as malignant or post-infectious inflammatory condition, including COVID-19, as mentioned in the 2021 European pediatric rheumatology congress.[4] Nevertheless, our patient

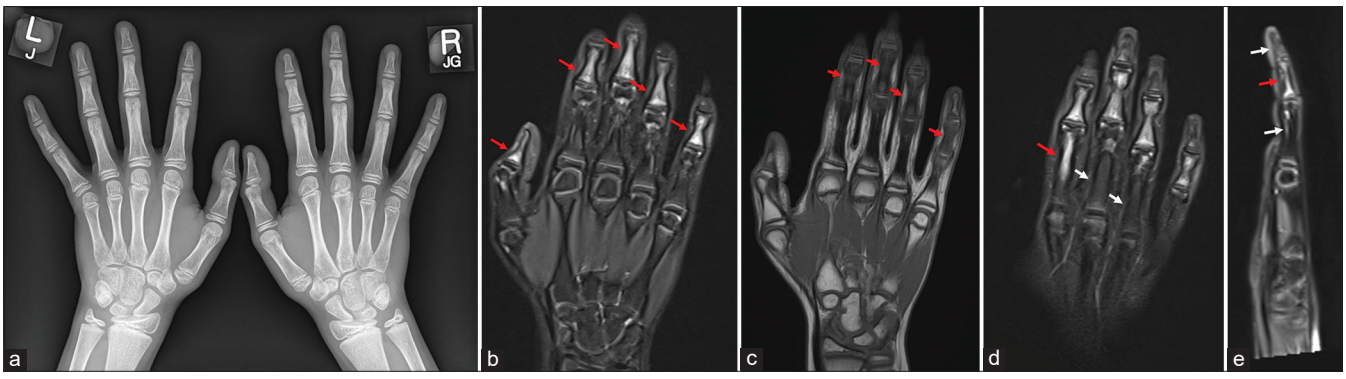

Figure 1: (a) A 12-year-old girl presented with swelling and pain in the right fingers. She previously had COVID-19 6 months ago. A dorsi-palmar radiograph of both hands showed no bony abnormality. There was no periosteal reaction or evidence of bony erosion. (b) T2 fat suppressed coronal images of the right hand shows abnormal signal hyperintensity within the middle phalanges of the fingers and distal phalanx of the thumb consistent with edema (red arrows). (c) T1 weighted sequence of the same hand shows corresponding loss of the fatty marrow signal (red arrows). (d) Coronal T2 fat suppressed sequence of the same hand shows edema of the proximal phalanx of the index finger (red arrow) with sparing of proximal phalages of the other fingers (white arrows) as well as all distal phalanges. (e) Sagittal proton-density image of the right ring finger shows involvement of the middle phalanx (red arrows) with relative sparing of the proximal and distal phalanges.

experienced persistent swelling and further development of Raynaud's, photosensitive rash and eye dryness, hinting in the direction of connective tissue disease.

Finally, this case is different from the ones already present in the literature due to the presence of ANA, the non-resolution of PMS symptoms, and the recent COVID-19 infection. PMS is not linked with specific antibodies; plus, positive ANA is not utilized as a diagnostic element in rheumatological diseases. It is in fact common to detect such antibodies in about one-tenth of healthy children[7] and almost one-third of healthy individuals.[8] In different context, for example in juvenile idiopathic arthritis, the presence of ANA is used as a risk biomarker for the development of uveitis.[7] It is also worth noting that recent studies have shown a link between autoimmunity and infection by COVID-19, with single-center studies, where almost half of hospitalized patients were found to have developed ANA post-COVID-19.[9,10] Therefore, despite PMS not being considered to be a possible COVID-19 complication and the lack of evidence for COVID-19, PMS, and connective tissue disease to be correlated in children, the development of autoimmunity with joint inflammation and PMS as a consequence of COVID-19 should be investigated further.

CONCLUSION

PMS is a rare syndrome which should be one of the differential diagnoses when assessing children with joint pain and swelling. MR is often performed quite early; however, this could be avoided in patients with typical radiological findings as this condition is deemed to be benign and self-limiting. Little is known about the long-term effects of COVID-19 in children; nevertheless, autoimmunity, joint pain, and autoantibodies are known sequelae of this viral infection with increasing studies looking into that. Therefore, research should be conducted to investigate possible connections between COVID-19 and inflammatory conditions, including PMS, in children.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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