

## Case report

### **Two rare cases of adult onset phalangeal microgeodic syndrome with magnetic resonance imaging-proven bone edema transiently occurring in winter**

Ayako Nishino<sup>1</sup>, Shin-ya Kawashiri<sup>1,2</sup>, Yoshikazu Nakashima<sup>1</sup>, and Atsushi Kawakami<sup>1</sup>

<sup>1</sup>Unit of Translational Medicine, Department of Immunology and Rheumatology, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan

<sup>2</sup>Department of Public Health, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan

*Address for correspondence and reprint requests:*

Shin-ya Kawashiri, M.D., Ph.D.

Department of Public Health

Nagasaki University Graduate School of Biomedical Sciences,

1-7-1 Sakamoto, Nagasaki 852-8501, Japan

Phone: + 81-95-819-7067

Fax: +81-95-819-7069

E-mail: shin-ya@hotmail.co.jp

**Abstract**

We report two rare cases of adult onset phalangeal microgeodic syndrome (PMS), which commonly develops in children. Both cases were Japanese women, with Case 1 60 years old and Case 2 58 years old. They developed swelling and stiffness in their fingers in winter, and their symptoms disappeared without any treatment in summer. Magnetic resonance imaging (MRI) of bilateral hands showed diffuse bone edema of the middle and proximal phalanges in winter, but the abnormal findings also disappeared in summer. The specific abnormalities observed on MRI were very useful for the diagnosis of PMS. Interestingly, they appeared in winter and disappeared in summer in parallel with the symptoms.

**Key words**

Phalangeal microgeodic syndrome, Magnetic resonance imaging, Bone edema

**List of abbreviations used**

ACPA: anti-cyclic citrullinated peptide antibodies

CRP: C-reactive protein

ESR: erythrocyte sedimentation rate

MMP-3: matrix metalloproteinase-3

MRI: magnetic resonance imaging

PMS: phalangeal microgeodic syndrome

RF: rheumatoid factor

STIR: short inversion time inversion recovery

## **Introduction**

Phalangeal microgeodic syndrome (PMS) is a rare condition affecting the fingers, mostly in children [1]. Patients with PMS show frostbite-like symptoms, such as swelling, redness, and pain in several fingers [2-5]. Case reports on PMS in adults are rare. Recently, the observation of diffuse bone marrow edema on magnetic resonance imaging (MRI) was reported to be helpful in making a diagnosis of PMS [1, 2].

We herein describe two rare cases of adult-onset PMS. The typical abnormalities seen on MRI developed in winter and disappeared in summer in parallel with changes in the symptoms.

## **Case report**

### **Case 1**

A 60-year-old Japanese woman visited our hospital because of swelling and stiffness in her fingers in February 2011 (winter). On physical examination, she had swelling and tenderness in the index and middle fingers of bilateral hands. Laboratory data showed that C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), and matrix metalloproteinase-3 (MMP-3) were within normal limits. Both rheumatoid factor (RF) and anti-cyclic citrullinated peptide antibody (ACPA) were negative. Plain radiography (**Figure 1A**) of bilateral hands was normal. Ultrasonography of bilateral hands did not detect articular synovitis and tenosynovitis. MRI-short inversion time inversion recovery (STIR) showed diffuse high signal intensity in the bone marrow of the middle and proximal phalanges of bilateral hands, which suggested bone edema (**Figure 1B**). In the following summer, her symptoms and the above MRI findings disappeared without any treatment (**Figure 1C**). She was diagnosed as PMS based on the specific clinical course and the specific findings of MRI.

### **Case 2**

A 58-year-old Japanese woman developed stiffness and tenderness in her fingers in February 2010 (winter). She visited our hospital in the following summer, but her symptoms had improved. Laboratory data showed CRP, ESR, MMP-3, RF, and ACPA to be negative. Plain radiography (**Figure 2A**), ultrasonography, and MRI (**Figure 2B**) of bilateral hands were normal. In the following winter, her symptoms flared up again. Although she had swelling and tenderness in the index and middle fingers of bilateral hands on physical examination, laboratory data showed that acute phase reactants were normal. MRI-STIR showed diffuse high signal intensity in the bone marrow of the middle and proximal phalanges of bilateral hands (**Figure 2C**). She was diagnosed as PMS based on the specific clinical course and the specific findings of MRI. Although her symptoms disappeared again the next summer without any treatment and relapsed again the following winter.

## **Discussion**

Maroteaux described five infants who had so-called PMS as having frostbite-like symptoms of the phalanx [3]. The PMS cases previously reported were almost all in Japan [1, 2, 4-7]. The pathogenesis of PMS is considered to be a transient disturbance of the peripheral circulation caused by cold temperatures, because it frequently occurs in winter. The most common lesions are the middle phalanges of the index and middle fingers [7]. Most cases of PMS occur in children, and only one adult case has been previously reported [4]. The radiographic features of PMS typically show mild osteosclerosis with cortical irregularity and multiple small radiolucent spots of osteolysis in the diaphysis [1, 3-5, 7]. In the recent reports, MRI was found to be helpful for the diagnosis of PMS [1, 2]. In PMS, the bone marrow of the phalanges shows diffuse low signal intensity on T1-weighted images and high signal intensity on STIR images [1, 2], which indicates bone edema. The symptoms regress within six months without any treatment in most patients. The prognosis of PMS is generally excellent, but recurrent cases have been reported [6].

We herein described two rare cases of adult-onset PMS. The specific abnormalities observed on MRI were very useful for the diagnosis of PMS in case differential diagnosis is necessary forward inflammation peripheral arthritis including rheumatoid arthritis and osteomyelitis. Interestingly, the abnormalities observed on MRI appeared in winter and disappeared in summer in parallel with the symptoms. The latter patient was a recurrent case. Radiographic abnormalities may appear with a relapse of the condition.

## References

- [1] Yamamoto T, Kurosaka M, Mizuno K, Fujii M. Phalangeal microgeodic syndrome: MR appearance. *Skeletal Radiol* 2001;30:170-2.
- [2] Fujita A, Sugimoto H, Kikkawa I, Hyodoh K, Furuse M, Hoshino Y. Phalangeal microgeodic syndrome: findings on MR imaging. *AJR Am J Roentgenol* 1999;173:711-2.
- [3] Maroteaux P. A microgeodic disease of unknown etiology affecting the finger phalanges in infants: report of five cases. *Ann Radiol (Paris)* 1970;13:229-36.
- [4] Sato K, Sugiura H, Aoki M. Transient phalangeal osteolysis (microgeodic disease). Report of a case involving the foot. *J Bone Joint Surg Am* 1995;77:1888-90.
- [5] Yamamoto T, Marui T, Akisue T, Nakatani T, Kurosaka M. Phalangeal microgeodic syndrome resulting in rapid digital shortening. *Clin Orthop Relat Res* 2004;424:191-3.
- [6] Hikosaka K, Koyama A, Yabe H, Mori K, Nakagawa K, Inoue K, Mizutani M. Pathological fracture in microgeodic disease, a report of six cases. *J. Japanese Soc. Surg. Hand* 1985;2:404-7.
- [7] Inoue G, Miura T. Microgeodic disease affecting the hands and feet of children. *J Pediatr Orthop* 1991;11:59-63.

## **Acknowledgements**

n.p.



**Conflict of interest**

The authors have no conflicts of interest to report.

### **Figure Legends**

**Figure 1** The plain radiography and MRI findings in Case 1.

Although plain radiography (A) of bilateral hands was normal, MRI-short inversion time inversion recovery (STIR) (B) showed diffuse high signal intensity in the bone marrow of the middle and proximal phalanges. The following summer, the MRI-STIR abnormalities disappeared (C).

**Figure 2** The plain radiography and MRI findings in Case 2.

Plain radiography (A) and MRI (B) of bilateral hands were normal at the patient's first visit in summer. An MRI-STIR image of bilateral hands showed diffuse high signal intensity in the bone marrow of the middle and proximal phalanges in winter (C).

Figure 1

A



B



C

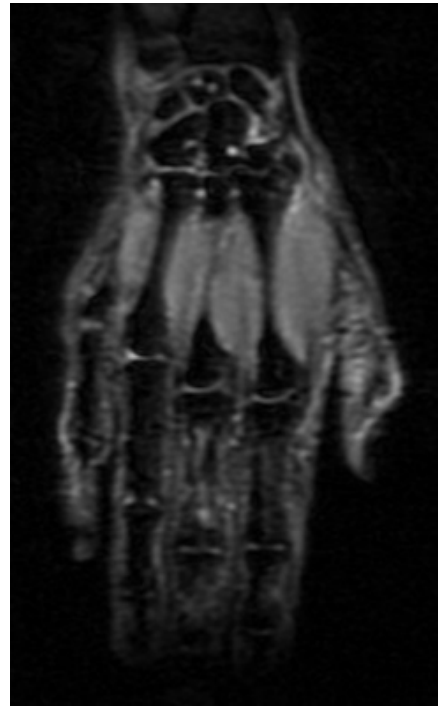
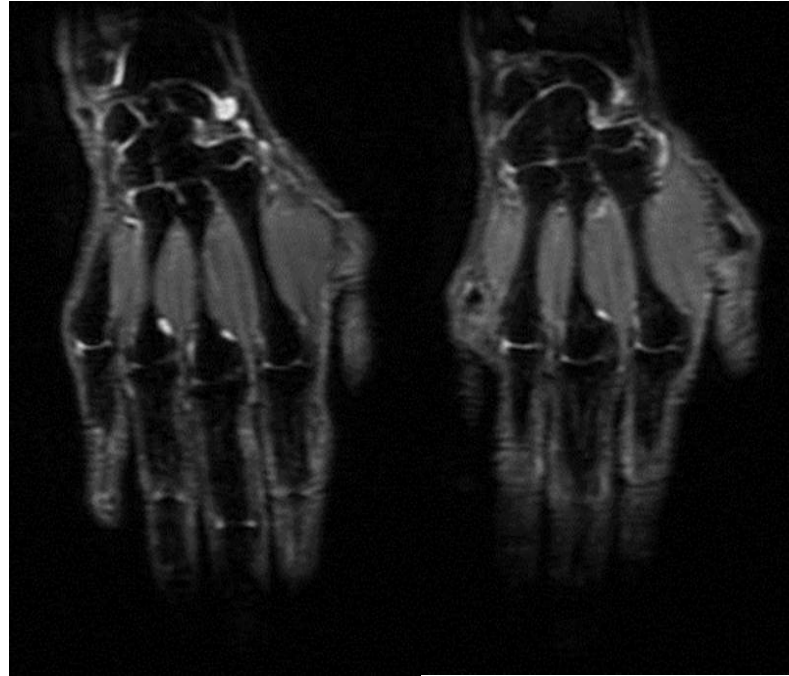


Figure 2

A



B



C

