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Thiemann's Disease: a Case Report

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Thiemann's disease is a form of idiopathic avascular necrosis of the immature epiphyses of the phalanges of the fingers and toes. Few cases of Thiemann's disease have been reported because the disease is rare and difficult to diagnose. To the best of our knowledge, magnetic resonance imaging (MRI) findings of Thiemann's disease have not been reported. Here, we report a case of Thiemann's disease diagnosed by typical clinical symptoms and characteristic MRI findings before radiologic bony abnormalities were apparent.

Keywords: Thiemann's disease; Phalanges; Magnetic resonance imaging

Case Report

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INTRODUCTION

Thiemann's disease is considered an avascular necrosis of the epiphyses of the phalanges of the fingers and toes (1, 2). The definite etiology of Thiemann's disease has not been documented, however, as the majority of cases develop within families, a genetic factor is considered to be a main contributor. Most cases of Thiemann's disease show an autosomal dominant pattern of inheritance with strong penetrance (3, 4). However, few sporadic cases have been reported and they have more commonly been reported in males (5).

The most common clinical manifestation is progressive swelling of the involved proximal interphalangeal (PIP) or the distal interphalangeal (DIP) joint of the hand with or without pain. The middle finger is most frequently involved (3, 4). Thiemann's disease develops insidiously without any apparent cause, although sometimes the disease is initiated by trauma or exposure to cold (5). In most of the cases, the disease develops in the second decade of life. On pathologic evaluation of the disease, Dessecker reported varying degrees of aseptic necrosis with normal vessels and an absence of inflammation in the epiphysis of the PIP joint of the middle finger (6).

The diagnosis of Thiemann's disease has been dependent on radiologic abnormalities such as flattening, fragmentation, increased radiographic opacity, broadening of the phalangeal epiphysis of the hand in compatible clinical manifestations, swelling of PIP joints of the fingers, onset before 25 years and negative acute inflammatory laboratory tests (7). In some cases, joint space narrowing and thickening of the phalangeal base with phalangeal shortening simulating Heberden and Bouchard nodes has also been noted in the late phase though while the patient was at a young age (3, 8).

Recently, early diagnosis of avascular necrosis not shown radiography has been possible using magnetic resonance imaging (MRI). To the best of our knowledge, this is the first reported case of Thiemann's disease in Korea. We present a case of Thiemann's disease diagnosed by the typical clinical symptoms and characteristic MRI findings

before any classical radiologic bony abnormalities.

CASE REPORT

A 10-year-old boy with no previous medical history complained of swelling and mild pain in the right fifth finger. This symptom developed after a minor extension trauma, approximately 5 months earlier.

Physical examination showed fusiform swelling in the PIP joint of the right fifth finger with mild tenderness. No restriction of motion or skin color change was noted in this area. There was no involvement of other joints on both hands on physical examination. Laboratory tests including white blood cell (WBC) count, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP) were unremarkable. Simple radiography showed soft tissue swelling around the right fifth PIP joint, but no fracture line or other abnormal bone lesion was noted (Fig. 1). Diagnostic radiology was consulted to rule out ligamentous injury or inflammatory diseases, such as low-grade infection or juvenile idiopathic arthritis. MRI showed diffuse low signal intensity lesion with loss of normal fat marrow signal confined to the proximal epiphysis of the right fifth middle phalanx on the T1-weighted image (Fig. 2a, b). There was no definite linear fracture line on T1-weighted image. A linear high signal intensity lesion confined to the bone marrow space in the proximal epiphysis of the right fifth middle phalanx on the fat-suppressed T2-weighted image (Fig. 2c) suggested osteonecrosis of phalangeal epiphysis, Thiemann's disease.

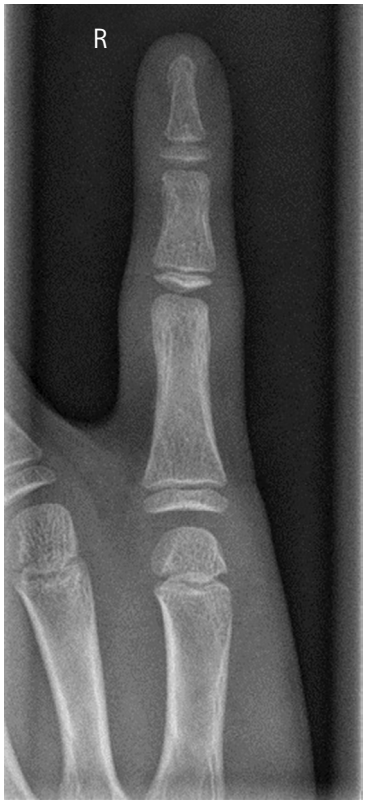
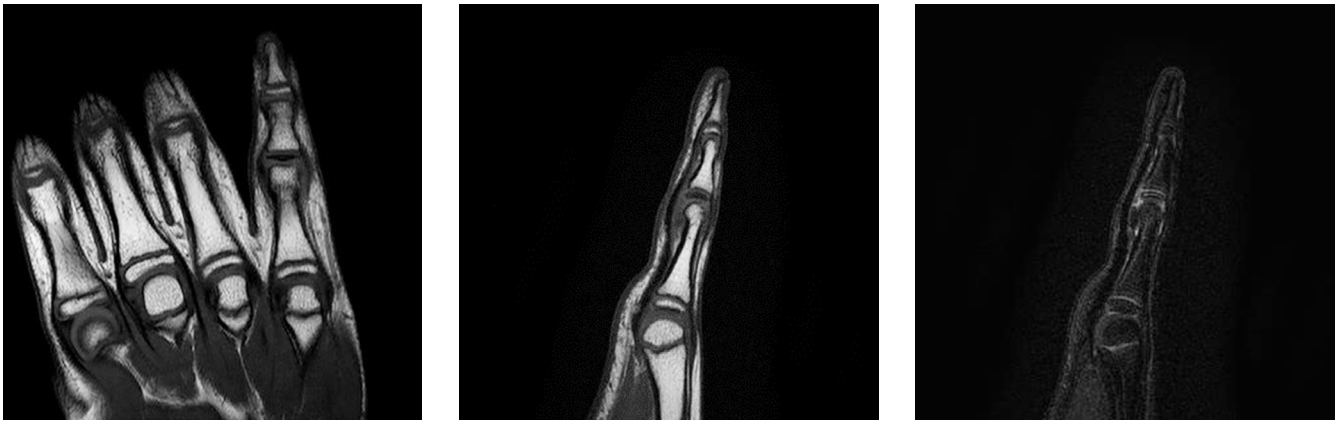


Fig. 1. Simple radiograph of right fifth finger shows no fracture line or other abnormal bone lesion.



a b c

Fig. 2. MRI of right hand (a) on coronal T1-weighted image shows diffuse low-signal intensity lesion with loss of normal fat marrow signal confined to the proximal epiphysis of the right fifth middle phalanx. There was no definite fracture line. (b) The sagittal T1-weighted image also shows diffuse dark signal intensity lesion in same region. (c) The sagittal fat-suppressed (c) The sagittal fat-suppressed T2-weighted image shows abnormal linear high-signal intensity lesion in proximal epiphysis of the right fifth middle phalanx. There was no significant joint effusion in proximal interphalangeal and distal interphalangeal joints of right fifth finger.

There was no significant joint effusion in the PIP joint of right fifth finger.

This patient eventually had resolution of symptoms with conservative management. No deformity of the involved joint with no pain, swelling or limitation of motion was reported on a telephone follow-up after four years.

DISCUSSION

In patients with swelling or pain in the PIP or DIP joint of the finger, traumatic injury or infectious lesion should always be the first diagnostic considerations because of the common occurrences of these causes. A different diagnosis of Thiemann's disease includes infarction of the epiphysis secondary to blood abnormality and metabolic disorders (3, 4). Along with clinical manifestations and laboratory data, imaging findings are helpful to rule out other common causes of finger pain including traumatic, metabolic, infectious or inflammatory diseases. Among the various imaging modalities, MRI is well-known as the best diagnostic modality for accurate diagnosis of various musculoskeletal diseases, due to its ability to provide superior soft tissue contrast. In the present case, the patient could be correctly diagnosed with Thiemann's disease, a rare disorder, by combining the clinical, laboratory and characteristic MRI findings of epiphyseal involvement without other imaging abnormalities in this growing child. Although the MRI showed abnormal bone marrow signal alteration in the epiphysis of involved PIP joint, there was no bony abnormality on radiography, suggesting that the disease was in an early stage. Because there was no familial history of swelling or pain in the phalangeal joint of the hand, this case is considered a sporadic case of Thiemann's disease.

Most reported cases showed deformity of the involved joint on radiography at the time of diagnosis. Some cases show improvement of radiologic finding of the joint on follow-up study (9). This case shows the typical MRI findings of Thiemann's disease before any classical radiologic bony abnormalities appear.

With the typical MRI finding of Thiemann's disease, we were able to exclude other diseases and correctly diagnose the Thiemann's in an early stage. Early diagnosis may improve the prognosis by initiating proper conservative treatment to prevent the deformity of the joint.

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