

# Congenital Insensitivity to Pain with Recurrent Septic Arthritis of the Left Knee

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## ABSTRACT

A 15-year-old Malay male with congenital insensitivity to pain presented with recurrent septic arthritis of the left knee complicated by osteomyelitis. Repeated arthrotomy and wound debridement was done, however, the condition was not resolved. Amputation was suggested as the best method of treatment to eradicate the infection.

**KEYWORDS:** Congenital insensitivity to pain, Osteomyelitis

## INTRODUCTION

Congenital insensitivity to pain is a rare inherited disorder with multiple musculoskeletal problems such as infections, fractures, growth disturbances, avascular necrosis, Charcot arthropathy, joint dislocation and heterotopic ossifications. We reported a case of congenital insensitivity to pain with recurrent septic arthritis and osteomyelitis of the left knee.

## CASE REPORT

A 15-year-old Malay male known to have congenital insensitivity to pain (diagnosed clinically in 2003), presented to us with recurrent episodes of septic arthritis of the left knee. In 2004, he had 4 episodes of the same problem where arthrotomy of the left knee was done followed by administration of antibiotics. He presented again in early 2005 with swelling of the left knee which was progressively increasing in size. There was no history of night sweats, chronic cough or contact with tuberculosis patients. He had no constitutional symptoms or any other medical problems and he was wheelchair bound.

Clinically he was afebrile. His left knee was swollen and deformed. There was a midline surgical scar due to multiple arthrotomy with small wound at the centre. Multiple scars were seen at the medial and lateral aspect of his left knee (Figure 1).



**Figure 1:** Left knee swelling with a midline surgical scar and multiple healed scars around the knee joint.

It was warm and boggy on palpation. His right knee, right upper limb and left upper limb were deformed due to previous fractures.

His white cell count was  $6.0 \times 10^3$  / ul. Erythrocyte sedimentation rate (ESR) was 112 mm/hour. Radiograph of the left knee showed soft tissue swelling around the knee, destruction of the knee joint with an evidence of osteomyelitic changes of the distal femur and proximal tibia (Figure 2).



**Figure 2:** Radiograph of the left knee showing destruction of the knee joint with an evidence of osteomyelitic changes of the distal femur and proximal tibia.

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Magnetic resonance imaging of the left knee was performed to access the severity of the intramedullary osteomyelitis in the left femur and tibia. There was thickened synovium and surrounding muscles in the distal third of the left femur and proximal tibia. Osteomyelitic changes of the distal femur and proximal left tibia fibula was also seen. Radiological diagnosis was Charcot joint with septic arthritis of the left knee.

Emergency arthrotomy and debridement of the left knee was performed and intraoperatively five hundred milliliters of pus was evacuated. There was a generalized synovial thickening and sequestra noted in the left knee. Synovial tissue and bone were sent for histopathological examination.

Post-operatively he looked cheerful, histopathological examination of the specimens was consistent with septic arthritis of the left knee. There was no evidence of tuberculosis. The synovial fluid culture and sensitivity examination did not obtained any growth of organism. He was treated with intravenous cloxacillin empirically.

In view of the multiple episodes of septic arthritis of the left knee complicated by osteomyelitis and as the patient was not ambulating, left above knee amputation was suggested but the patient and parents did not agree.

## DISCUSSION

Congenital insensitivity to pain was first described in 1932 by Dearborn.<sup>1</sup> It is a rare inherited disorder which may affect various tracts in the peripheral nervous system and the autonomic nervous system is the worst affected. Presentation in childhood is commonly at the time of tooth eruption, with biting of tongue or lips. Diagnosis can be made by clinical presentation, electromyography (EMG) to exclude other peripheral neuropathies, and skin biopsy to demonstrate absence of epidermal and sweat gland innervation.<sup>2</sup> Genetic studies in a Malaysian family have shown mutations on exon 16 (V709L and G718S) to be responsible for the disease.<sup>2</sup>

The principal orthopaedic manifestations are neuropathic joints, osteomyelitis and recurrent fractures of the limbs.<sup>3</sup> Patients are at high risk of injury due to loss of pain sensation which is a protective mechanism of the body.<sup>4</sup> The importance of orthopaedic management lies in the prevention of musculoskeletal complications by educating the patients and family members regarding the risks in relation to the environment in the absence of pain constraints and the high incidence of self-abusive behavior.<sup>5</sup>

Our patient presented at the late age with recurrent swelling of the left knee. He was afebrile with a normal white cell count and elevated ESR but no aspiration and cultures was made preoperatively. Clinically he was diagnosed to have septic arthritis of the left

knee complicated by osteomyelitis. Schulman et al. reported that osteomyelitis with rapid bony destruction was found in almost 50% of patients.<sup>6</sup> Ideally aspiration and cultures should be done before undertaking any surgical debridement to prevent unnecessary surgery and damage to the growth plate because sometimes it is difficult to differentiate between fractures and infections in congenital insensitivity to pain.<sup>7</sup>

Once the diagnosis of infection has been made, debridement should be done to control the infection. His recurrent episodes of knee infection could be related to inadequate debridement of the left knee joint. However in view of his recurrent problems with severe destruction of the knee joint radiologically and as he was wheelchair bound, an above knee amputation would be the best way to control the infection.

## CONCLUSION

Management of a patient with congenital insensitivity to pain complicated by infection is challenging, correct medical and surgical treatments are mandatory in order to achieve a good outcome.

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