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Case Report

Congenital Insensitivity to Pain with Septic Arthritis: A Case Report

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Abstract

Objective: Congenital Insensitivity to pain is a rare inherited disorder that has been associated with multiple musculoskeletal complications (e.g,: septic arthritis, osteomyelitis, joint dislocation, avascular necrosis…etc.). A detailed medical history and routine joint examination and assessment are needed with a high index of suspicion of such complications. There is a high risk of morbidity and mortality in cases that are missed and present with multisystem involvement. Early recognition and intervention are of the essence in such cases.

Clinical presentation and intervention: We report a 9-year-old female child, admitted to the pediatric intensive care unit due to septic shock because of septic arthritis with congenital insensitivity to pain. She remained in a critical state till irrigation and debridement were done for the joint then she showed full recovery.

Conclusion: Patients with insensitivity to pain require detailed examination and close observation of their joints. A simple joint abscess can be complicated to multi-organ dysfunction when it is missed and not identified early on. A scheduled routine joint examination with caregiver education when seeking medical advice is crucial. Children with insensitivity to pain will not manifest early symptoms of their musculoskeletal complications.

Keywords: Congenital Insensitivity to Pain; Septic Arthritis

Introduction

Congenital Insensitivity to pain (CIP) is an extremely rare genetic disorder. Multiple known genes might lead to failure of nociceptor development in CIP, which might help in the diagnosis while others remain cryptogenic [1]. Patients with CIP have a wide range of presentations. Orthopaedic manifestations should be considered and assessed with each presentation, hospital visit, and follow-up, especially in weight-bearing joints; it can start as an infected ulcer but end up with chronic osteomyelitis or septic arthritis requiring amputation to control the infection [2]. Septic arthritis is not an uncommon complication of CIP but due to the rarity of the disease early recognition is missed and the patient end up with unfavourable outcome such as amputation [3]. Here, we present a case of congenital insensitivity to pain with septic arthritis, which required Pediatric critical care admission with septic shock and multiorgan dysfunction due to septic arthritis. Once the definitive management of septic arthritis was carried out, the child showed significant improvement.

Case Report

A 9-year-old female known case of congenital insensitivity to Pain (CIP) presented to the hospital emergency department (ED) on Nov. 01, 2021, with vomiting, diarrhea, and inability to bear weight. She had a right hip dislocation and was using a wheelchair with the assistance of a walker to roam around at home. She was admitted with the suspicion of a case of viral gastroenteritis to the general pediatric ward on the same day of the ED presentation. After 2 days of admission, she was shifted to the High Dependency unit (HDU) due to deterioration of her clinical condition in form of hypotension and tachycardia. She received fluid resuscitation of 70

ml/kg in total for which she temporarily showed improvement but after three hours she started to desaturate with an altered level of consciousness so shifted to Pediatric Intensive Care Unit (PICU). In the PICU, she developed fluid refractory hypotensive shock so inotropic support (Epinephrine and Norepinephrine) was started on the same day of admission and with the desaturation which might be due to capillary leak syndrome into her lungs and the development of a picture of Acute Respiratory Distress Syndrome (ARDS), for that she was intubated. Echocardiography was done for her on PICU admission with the signs and symptoms of poor perfusion, hypotension, and prolonged capillary refill time; myocarditis could not be rolled out. It showed depressed biventricular systolic function with an estimated EF of 43%, so Milrinone was started on top of Epinephrine and Norepinephrine. All bacterial cultures (blood, urine, and respiratory) were negative except for a wound culture in her digits which showed growth of Methicillin-Resistant Staphylococcus aureus (MRSA). The wound was already healing with no redness, pus, or signs of infection. She was already receiving broad-spectrum antibiotics (Meropenem and Linezolid) but was still hemodynamically unstable with inotropic support and multi-organ dysfunction. On Nov. 04, 2021, A suspicion of septic arthritis was raised as there isn't a clear source of infection, so a hip ultrasound was ordered, and the result showed bilateral moderate joint effusion with synovial irregularities, hyperemia as well as internal debris.

Pediatric Intervention Radiologist (IR) did Ultrasound Guided aspiration at the bedside since the child wasn't stable enough to be moved to the operation theater (OT), which drained 12 ml of pus from the right hip and 20 ml of serous fluid aspirated from the left hip in a trial to control the source of infection. After 3 days, the child was still requiring inotropic support, covered by antibiotics with multi-organ dysfunction, a decision was taken to send her for irrigation and debridement of the hip joint to clear the site of infection in the OT. A multidisciplinary team meeting was done with Pediatric Orthopaedic, Pediatric Anaesthesia, and Pediatric Critical Care team, and the child's parents to discuss the risks and benefits of the procedure and the possibility of morbidity and/or mortality. The parents understood the critical situation and an agreement was reached to proceed with the surgery. Pediatric Orthopaedic shifted the patient to OT for irrigation and debridement of the right hip. On aspirating, the right joint drained 50 ml pus, and samples were collected for culture and sensitivity from the capsule and synovial membrane, which later came normal. A total of 9 Litters of normal saline mixed with antibiotics (Cefazolin and gentamycin) was used to irrigate and clear the joint. The next day her hemodynamic became stable and both milrinone and Norepinephrine were weaned off. Epinephrine was weaned 4 days later. Hip MRI was done 3 days post-op, which showed an intramuscular abscess in the right gluteus muscle with pockets of effusion around the femoral head with thick synovial enhancement. She went again for hip irrigation and debridement on Nov. 11th with the insertion of a surgical drain. She showed significant improvement post-op and was extubated 3 days after the second surgery. She continued her stay in PICU till Nov. 24th as she developed withdrawal and Delirium for which she received the proper management and then shifted to the High Dependency unit (HDU).

Currently, the child discharged from the hospital and is at home, back to her baseline.

Discussion

Congenital Insensitivity to Pain (CIP) is a rare genetic disorder. It is characterized by the loss of the basic human defense mechanism, which is nociception. It can be a result of a failure of development of the receptors in the first place or they developed normally but failed to respond to tissue damage. Currently, some gene abnormalities associated with the disease are known and can be detected when there is a high index of suspicion of CIP [1]. CIP can be associated with anhidrosis, temperature instability, or orthopaedic abnormality [4]. The orthopaedic manifestations of CIP can vary widely, it can be as simple as having joint instability or an ulcer in the skin covering that joint to septic arthritis which requires amputation and losing a limb [3,4]. Septic arthritis incidence can range from 4 to 10 per 100,000 healthy children [5] but when it comes to children with CIP where they can get recurrent trauma to their joints without seeking medical advice and that joint became deformed the risk of developing an infection at that site is tremendously high. The usual background of fever and decreased range of motion, tenderness, and swelling might not be there for patients with CIP. Decreased range of motion is a result of pain sensations that limit flexion or extension of the joint, which they lack in the first place [5]. In the E.Bar-On review for orthopaedic manifestations for patients with CIP, it was found that 5 out of the 13 patients they reviewed, developed infected bursitis which later on progressed to chronic osteomyelitis or septic arthritis [2]. Hip joint dislocation is a common complication in patients with CIP [6], which makes it difficult to suspect infection or septic arthritis if the affected joint was the same hip joint except in later stages when the patient manifests systematically. Early recognition when the infection is local at the site of the joint requires a high index of suspicion and delicate examination of joint. Early antibiotics treating septic arthritis before the systemic manifestation would result in a preferred outcome and the hallmark of surgical treatment would involve irrigation of the joint and incision and drainage might be avoided [5]. Unfortunately, due to pain insensitivity, most patients with CIP present late which requires surgical interventions such as our case and a case, which was described by Dr. Zamzuri Z., which required amputation to control the source of infection [3]. Due to the rarity of the condition, there is no consensus about regular assessments, follow-ups, radiology work up, and when to

intervene for joint aspiration in case of suspicion. The main goal in the management of patients with CIP should be parents' education and help them to early detect any abnormalities in their child's musculoskeletal and seek medical advice to avoid complications. In our case, though the diagnosis was reached late, and she developed systemic manifestation of sepsis in form of hypotension and requirement of inotropic support, the moment joint incision and debridement were done, immediately she showed improvement as the source of infection was controlled.

Conclusion

Patients with insensitivity to pain require detailed examination and close observation of their joints. A simple joint abscess can be complicated to multi- organ dysfunction when it is ignored and not identified early on. Regular assessments, follow-ups, and investigations should be constructed for patients with CIP.

Conflicts of Interest: The authors declare that there are no financial or other conflicts of interest to disclose.

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