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Distal Femoral Physiolysis in a Myelomeningocele Patient - Early and Accurate Diagnosis

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Abstract

Background: Neurogenic physiolysis is the spontaneous widening, inflammation, and fracture of long bone physes in children with spinal cord defects. This condition is also referred to as epiphysiolysis and is often missed by parents and physicians alike due to the lack of typical fracture symptoms like pain and deformity.

Case Presentation: We present the case of a 5-year-old lumbar-level myelomeningocele patient who presented to the emergency department with edema, warmth, and erythema of the knee joint, and was later found to have elevated inflammatory markers concerning for infection. After a thorough workup helped to rule out infection and other life-threatening illness, the patient was successfully treated for physiolysis of the distal femur with cast immobilization.

Conclusions: General pediatricians and orthopaedic specialists should remain vigilant in recognizing physiolysis in this patient population. Missed or delayed diagnosis may lead to iatrogenic harm and can have negative long-term effects on patient's physical development and independence.

Abbreviations

MMC - Myelomeningocele, HKAFO - Hip-knee-ankle-foot-orthosis, PT - Physical therapy, ESR - Erythrocyte sedimentation rate, CRP - C-reactive protein, AP - Anterior-posterior, MRI - Magnetic resonance imaging

Introduction

Children with congenital spinal cord defects like myelomeningocele (MMC) are often times paraplegic and insensate in the lower extremities. Low bone mineral density and lack of pain feedback put them at high risk of atraumatic fractures in the lower extremities, seen in 10-30% of patients with myelomeningocele [1,2]. Spontaneous isolated long bone physeal fracture or physeal widening in spina bifida patients, referred to as physiolysis or epiphysiolysis, is thought to occur secondary to a combination of repetitive microtrauma, musculoligamentous imbalance, and osteopenia [3,4]. The proximal tibial and distal femoral physes are most often affected. The most common presenting symptom is a painless, warm, and swollen joint, oftentimes first noticed by a parent [2,3,5]. These patients, and their parents, often do not recall any precipitating event. The lack of pain and history of trauma not only delays diagnosis but often leads to the misdiagnosis of infection, possibly subjecting patients to iatrogenic harm secondary to invasive diagnostic tests, as well as continued repetitive microtrauma due to the delayed diagnosis [3]. The physeal widening seen on plain films of patients with physiolysis is similar to that seen in patients with scurvy, rickets, syphilis, and osteomyelitis [5,6]. Thus, early and accurate diagnosis is important.

We present the case of a child with lower lumbar spina bifida who presented with an atraumatic warm and swollen knee, found to have distal femoral physiolysis. Due to her lack of traumatic history, compounded with joint swelling and redness, the initial workup was for infection. This case demonstrates the importance of recognizing the unique presentation physiolysis in patients with spina bifida and avoiding a delayed or missed diagnosis.

Case Presentation

A 5-year-old female with a history of lower lumbar level myelomeningocele had been followed in our multidisciplinary spina bifida clinic since age 5 months. Her managed conditions included hydrocephalus, bowel and bladder incontinence, and bilateral lower extremity paralysis. She had been using a hip-knee-ankle-foot-orthosis (HKAFO) since age 2 and was able to use a stander for short periods during physical therapy (PT) but was otherwise wheelchair-bound. Due to the COVID-19 pandemic, her PT sessions had been cancelled for several months prior to presentation. The patient's mother brought her into the pediatric emergency department over the weekend with a chief complaint of left knee swelling noticed that morning. There was no recent history of trauma or travel. On exam, the left knee was moderately swollen with a large effusion, erythematous, and warm (Figure 1).

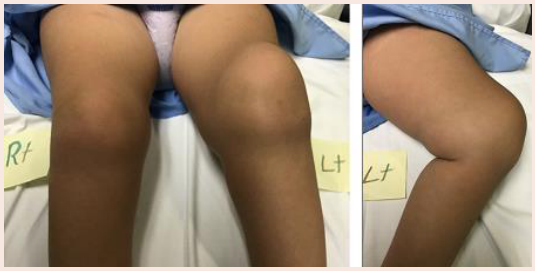

Figure 1: Physical examination of the patient's lower extremities obtained in the emergency department upon initial presentation with left knee swelling and warmth. There was a notable effusion and resting knee flexion best seen on the lateral view.

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There was no sensation below the proximal thighs and no motor function below the waste on bilateral lower extremities, as per baseline. The knee was grossly stable on exam with full range of motion. Plain X-Ray films of the left knee demonstrated distal femoral physeal widening and periosteal reaction (Figure 2).

Vital signs and labs at that time were within normal limits. The patient was placed into a long leg splint by the orthopaedic resident on call and scheduled for outpatient follow up. Three days later, the patient presented to the multidisciplinary spina bifida clinic for follow up. She again was noted to have a painless large effusion of the left knee. She was converted to a long leg cast and new x-rays were obtained. There was noted to be no significant change in alignment and osteolysis of the distal femoral physis with periosteal reaction. Salter-Harris I fracture of the distal femur was suspected. Magnetic resonance imaging (MRI) of the left knee with and without contrast was obtained 2 weeks from the time of presentation to further delineate the process. MRI showed inflammation throughout the distal femur and synovitis of the knee joint (Figure 3).

Erythrocyte sedimentation rate at that time was elevated at 53 and C-reactive protein was borderline at 3, raising some concern for infection. Knee arthrocentesis was performed and resulted in a normal cell count and negative cultures. At this point, we had comfortably ruled out the possibility of osteomyelitis or septic arthritis, and the diagnosis was confirmed as left distal femoral physiolysis. The patient was managed in well-padded cast immobilization of the left lower extremity for 2 months and then was transitioned to a custom HKAFO for another month. X-rays obtained 4 months from the time of onset demonstrated robust callus formation at the distal femoral metaphysis and early reconstitution of the physis (Figure 4).

The patient was then cleared for return to routine physical therapy without orthosis. At latest follow-up, 8 months after onset, the patient was now 6 years old and was doing very well without any recurrent left knee symptoms. Her function had returned to baseline. Final X-rays demonstrated complete resolution of osteolysis, further consolidation of callus, and maintenance of anatomic alignment (Figure 5).

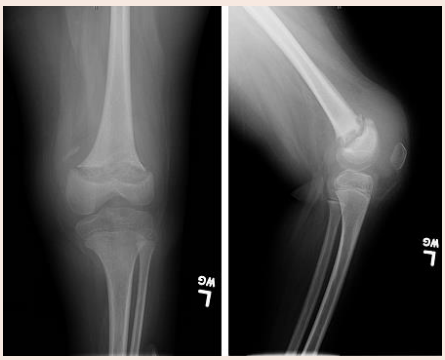

Figure 2: AP and Lateral radiographs of the left knee obtained in the emergency department upon the patient's initial presentation with left knee swelling and erythema. There is widening of the distal femoral physis and some early periosteal reaction seen on the lateral view. Anatomic alignment is maintained.

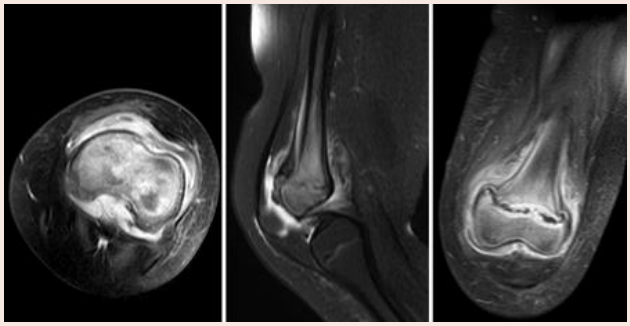

Figure 3: MRI of the left knee obtained 2 weeks from the time of initial patient presentation. T2 fat-saturated contrast-enhanced sections are shown in the axial (left), sagittal (middle), and coronal (right) planes. There is increased edema signal and contrast enhancement in the distal femur surrounding the physis. There is also a moderate effusion and synovitis in the knee joint. There is no obvious neoplastic or infectious nidus.

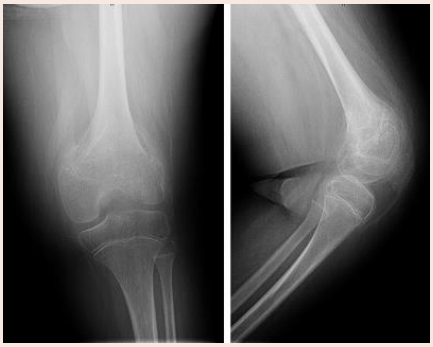

Figure 4: AP and Lateral radiographs of the left knee obtained 4 months from the time of the patient's initial presentation. There is robust callus formation in the periosteum and the area of prior osteolysis. There is early reconstitution of the distal femoral physis. Anatomic alignment is maintained.

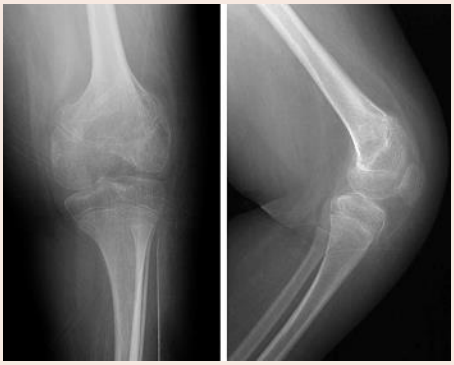

Figure 5: AP and Lateral radiographs of the left knee obtained at last visit 8 months from onset of symptoms. Shows further consolidation and resolution of the osteolysis. Anatomic alignment is maintained.

Discussion & Conclusion

The early recognition of physeal fractures in pediatric patients with MMC spina bifida is crucial in improving patient outcomes and avoiding long term consequences affecting their mobility and independence [2,3,7]. The delayed recognition of injury by parents in insensate patients can be further compounded by a physician's missed

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or delayed diagnosis. This delayed diagnosis can turn relatively minor injuries into unstable fractures due to repetitive microtrauma, which can ultimately lead to complications such as premature physeal arrest, angular deformity, or non-union [3,8]. Chronic stress on a fractured physis can lead to physiolysis with widening and delayed healing, which should be treated with immobilization of the extremity as soon as possible [2,3]. Initial workup with plain radiographs is the key in making the correct diagnosis, characterized by physeal widening and periosteal reaction without angular or translational deformity. However, these radiographic findings can often resemble osteomyelitis or sarcoma, and should be closely monitored with serial exams [2,7,9].

Furthermore, accurately diagnosing physiolysis in patients with spina bifida is challenging due to the presentation of systemic symptoms like fever, leukocytosis, and elevated inflammatory markers (ESR, CRP), which often lead to the mistaken diagnosis of cellulitis, osteomyelitis, or septic arthritis [2,3,10,11]. These symptoms and lab findings typically resolve with immobilization and close observation and should not always indicate advanced imaging or invasive procedures like arthrocentesis. However, if patient follow-up is limited, early and thorough diagnostic workup may be indicated to rule out more severe, life-threatening disease. MRI with and without gadolinium contrast can help identify the characteristic physeal widening of physiolysis with islands of calcification, irregularity of the zone of provisional calcification, and enhancement of the adjacent epiphysis and metaphysic [8].

General pediatricians and orthopaedic specialists should remain vigilant in recognizing the unique presentation of physiolysis in pediatric patients with spina bifida. Missed or delayed diagnosis may lead to iatrogenic harm and can have negative long-term effects on patient's physical development and independence.

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