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FAT EMBOLI SYNDROME IN A CHILD WITH DUCHENNE MUSCULAR DYSTROPHY AFTER MINOR TRAUMA

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□ **Abstract—Background: Fat embolism syndrome is the result of systemic manifestations of fat emboli in the microcirculation. Duchenne muscular dystrophy is a condition that increases the risk of fracture resulting in fat emboli. Case Report: We describe a patient with Duchenne muscular dystrophy who exhibited cardiopulmonary, neurologic, and ophthalmologic sequelae consistent with fat emboli syndrome after minor trauma. Why Should an Emergency Physician Be Aware of This?: Fat embolism syndrome is a rare but important consideration with significant morbidity and risk of mortality in patients with Duchenne muscular dystrophy after even minor trauma. Early recognition and aggressive resuscitation are crucial to positive clinical outcomes. Published by Elsevier Inc.**

□ **Keywords—Duchenne muscular dystrophy; fat embolism syndrome; pediatrics; fat microglobulinemia; tibial plateau fracture**

INTRODUCTION

Duchenne muscular dystrophy (DMD) is an X-linked recessive disease that affects one in 3600–6000 live male births that results in replacement of skeletal muscle tissue with fat due to deletions in the dystrophin gene. This causes a progressive muscle degeneration, weakness, and obesity, which predisposes affected individuals to falls and fractures (1). Loss of independent ambulation begins at age 9–12 years, and patients are usually nonambulatory by age 13–16 years, putting them at higher risk of falls and subsequent fractures (2). Cardiac, pulmonary, and orthopedic complications are most frequent, and without interventions, death from cardiac or pulmonary causes occur by age 20 years (3). Fat embolism syndrome (FES) is a clinical entity that arises from systemic manifestations of fat emboli in the microcirculation most commonly seen after orthopedic trauma (4). Although an alternative biochemical theory exists involving the release of free fatty acids from bone marrow in response to trauma, resulting in hypoxemia and inflammation, the end result is the same—increased capillary permeability and damage to surrounding tissue from inflammatory mediators (4). We report a case of FES in a 15-year-old boy with DMD who sustained a tibial plateau fracture after minor trauma.

CASE REPORT

A 15-year-old boy with DMD presented to the Emergency Department for evaluation of leg pain after a witnessed fall out of his wheelchair onto his left leg. There was no head trauma or loss of consciousness. He had a similar event 1 year previous that resulted

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in a femur fracture requiring internal fixation. In addition to DMD, his past medical history was significant for restrictive lung disease for which he was treated with chronic inhaled steroids. Surgical history was significant only for the previously noted internal fixation of the left femur. He had no known allergies, and his medications included carvedilol, lisinopril, albuterol, and vitamin D.

Initial examination revealed an obese 15-year-old boy in no acute distress. Oral temperature was 36.8°C (98.2°F), heart rate 136 beats/min, respiratory rate 20 breaths/min, oxygen saturation 98% on room air, and blood pressure 137/92 mm Hg. His lungs were bilaterally clear to auscultation without wheezes or rales. He was tachycardic but had a regular rhythm. Abdominal examination was unremarkable. Extremity examination revealed decreased active range of motion and muscle tone consistent with muscular dystrophy. There was mild swelling about the left knee, with minimal ecchymosis. He complained of pain with left knee movement but had full range of motion, as well as transient blurry vision. On initial neurological examination, he was alert and oriented to person and place, and his verbal interactions were at baseline per his parents. Cranial nerves II–XII were intact.

Left knee radiographs showed stable postsurgical changes of the left femur without evidence of acute fracture and a new irregularity of the medial physis of the left tibia (Figure 1). Computed tomography (CT) of the left knee showed a medial tibial plateau fracture with extension into the posterior tibial plateau. Orthopedics was consulted and recommended splinting with outpatient follow-up. During his stay, the child became increasingly tachycardic in the 140–150 beats/min range, above his baseline heart rate of 90–120 beats/min. He was also noted to have a temperature of 38°C (100.4°C) and a new oxygen requirement for saturations in the mid 80s. He was now speaking in only one-word sentences and appeared increasingly lethargic. For these reasons, a chest radiograph (CXR), urinalysis, and blood cultures were ordered in search of an infectious etiology, presuming these changes could be the result of sepsis. A head CT scan was ordered due to altered mental status, and a CT scan of the chest for pulmonary embolism was obtained for his increasing respiratory distress. Urinalysis, CXR, and CT of the head were normal. The CT of the chest did not show a pulmonary embolism, but did reveal diffuse ground glass appearance in both lungs (Figure 2).

The pediatric intensive care team was consulted and the child was admitted for further work-up of his tachycardia, altered mental status, and oxygen requirement. Bilevel positive airway pressure (BiPAP) was initiated due to continued respiratory distress. Azithromycin and ceftriaxone were started as empiric antibiotic coverage for presumed pneumonia. A magnetic resonance imaging study of the brain was obtained, which revealed

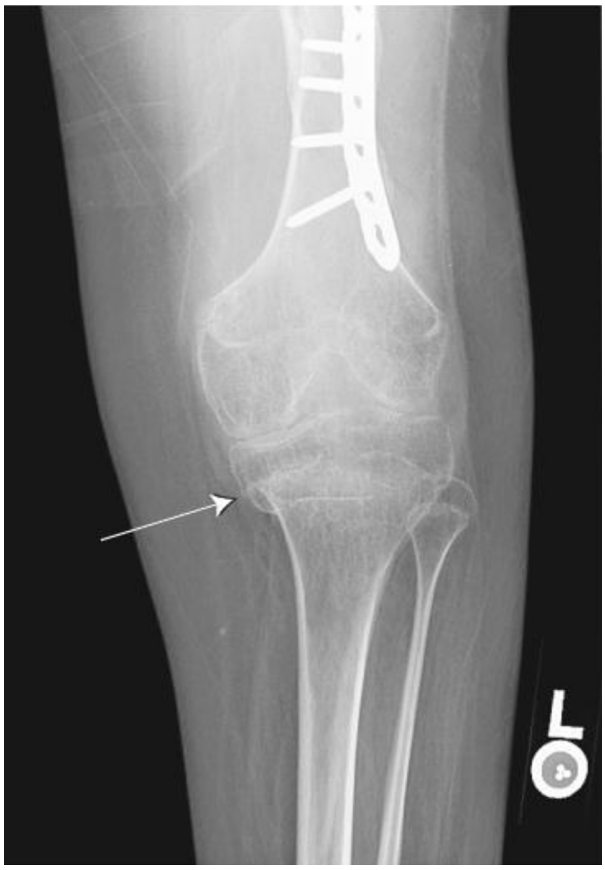

Figure 1. Left anterior-posterior knee x-ray study demonstrating stable postsurgical changes of the left femur without evidence of acute fracture and a new irregularity of the medial physis of the left tibia.

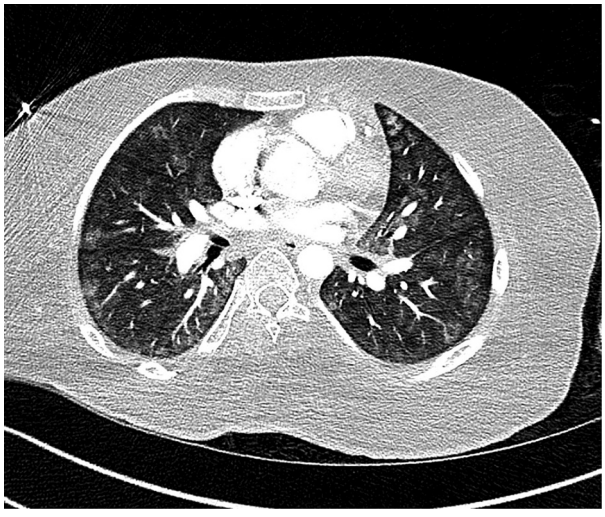

Figure 2. Computed tomography scan of the chest demonstrating ground glass opacities.