STARS FROM THE BONES - AN UNCOMMON PRESENTATION OF FAT EMBOLISM SYNDROME

LOKESH KOUMAR Sivanandam[1], Prakash Gupta[2], Maliha Rahim[3], Vivek Sanker[4], Ashna Pandita[5], and Umang Gupta[6]

[1]SLIMS
[2]University of Miami Miller School of Medicine
[3]Jinnah Sindh Medical University
[4]Noorul Islam Institute of Medical Science and Research Foundation Medicity
[5]University of Nis Faculty of Medicine
[6]Nepalgunj Medical College

March 1, 2023

Abstract

Trauma or surgery to the lower limbs can cause fat from the marrow within the leg bones to enter the bloodstream and form an embolus. However, if there is cerebral involvement without any pulmonary or dermatological manifestations at diagnosis, it could delay identifying cerebral fat embolism (CFE).

Introduction

Fat embolism is an abrupt disruption of the circulatory system commonly caused by orthopedic trauma, which is shown physically by the appearance of globules of fat in the vessels of the circulatory system. Fat embolism syndrome is the systemic presentation of fat emboli in the microcirculation, manifesting with the Gurd's triad of respiratory distress, neurological deficits, and petechial rashes [1,2]. According to the observations, pulmonary symptoms occur first, typically 24 to 72 hours after the trauma, but symptoms have been reported as early as 12 hours, usually followed by neurological symptoms [2]. The exact occurrence of FES varies from 1% to 30%, as there are no universal standards for diagnosis [3].

In 1861, Zenker was the first to conduct a clinical study of FES and discovered fat droplets in the capillaries of the lungs of a railroad worker who had died from a traumatic injury. However, despite Gauss' mechanical explanation of FES in 1924 and Lehman and Moore's biochemical explanation in 1927, the concept of FES remains obscure [4].

Recent studies suggest that FES may have possible connections with neurocognitive dysfunction and the development of deep vein thrombosis. Clinical manifestations of FES vary widely and can include restlessness, dyspnea, delirium, coma, and, in some cases, death. Case studies have also highlighted the rare occurrence of FES in patients not resulting from trauma, particularly during bone marrow removal, c-sections, liposuction, lung transplants, and cosmetic surgeries [2].

There are many ways to prevent FES and other long bone fractures from happening that have been discussed in the literature. Unfortunately, there is no particular treatment for FES, so the main alternatives are to give supportive care, although this is not a universal practice. Damage control orthopedics (DCO) and optimizing reaming approaches have been the principal management solutions for individuals with long bone fractures. Although FES can be very dangerous, it is much less common [5].

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Here we present a case of a 23-year-old male who suffered a left both bone closed fracture after an iron rod fell over his left lower limb. He eventually developed the classic triad of Fat embolism syndrome after being asymptomatic for 5 days following the trauma.

Case Report:

A 24-year-old male presented to the emergency department with shortness of breath and altered sensorium of 3 days duration. No symptoms were seen 5 days back, and no history of expectoration, hemoptysis, chest pain, palpitations, swollen limbs, or fever. On the night of the presentation, the sensorium was worsening in the form of decreased responsiveness, decreased speech output, and eye contact. Medical consultation was requested for persistent unresponsiveness. On examination, there was no history of weakness, seizure syncope, headache, cranial nerve palsies, visual defects, head trauma, or decreased urine output. No history of childhood epilepsy was revealed. Further investigations revealed he had a closed long bone fracture of the tibia and fibula five days back due to an iron rod that fell over the left lower limb. (Fig.1) He presented to the hospital the same day, and above-knee (AK) slab was placed, which the patient removed on his own the next day, and 2 days later, he attended a native treatment for fracture where manipulation and gross alignment were done.

Figure 1 : Left lower limb X-Ray showing closed long bone fracture of the tibia and fibula: AP View

General examination at presentation to Emergency Medical Services revealed mild pallor. He was conscious but not oriented. The pulse rate was 110 beats/min, the respiratory rate was 30 beats/min, blood pressure was 100/60 mmHg, and random blood sugar was 74 mg%. He had tachypnea and mild tachycardia and developed oxygen desaturation (O2 of 88%). The Thoracic Computed tomography (CT) findings showed diffuse bilateral pulmonary infiltrates comprising consolidations and ground-glass opacities. (Fig.2) He was sedated and intubated with subsequently transferred to the intensive care unit.
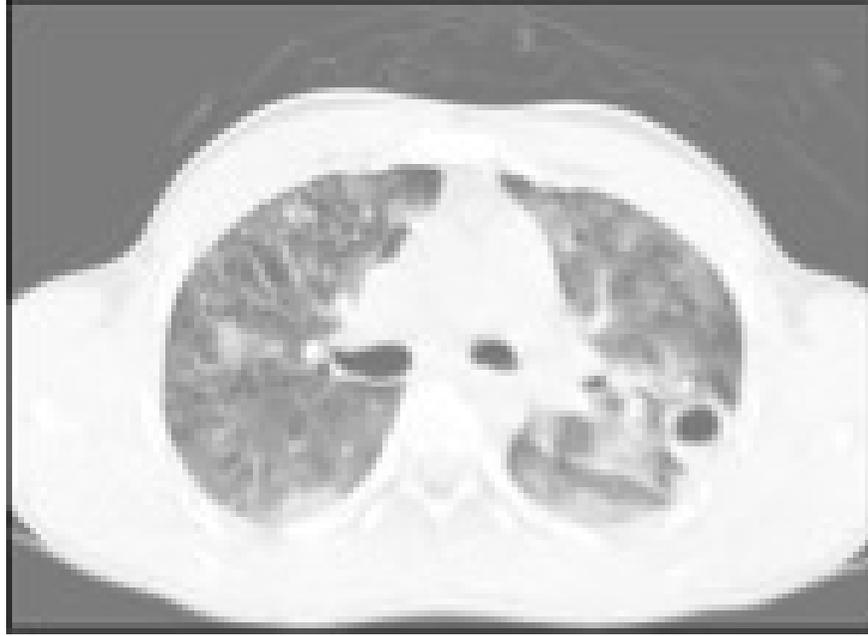
Figure.2: Thoracic computed tomography showing diffuse bilateral pulmonary infiltrates. Ground-glass opacity of uncertain cause is present.

There were no abdominal or pelvic abnormalities upon initial imaging workup, and a plain CT scan of the brain was unremarkable, with no ischemia or hemorrhage. (Fig.3) On the third day of admission, emergency magnetic resonance imaging (MRI) Brain revealed extensive bilateral symmetrical punctate foci of nearly the same size with abnormal signal intensities. These foci affect the cerebral hemispheres with restricted diffusion, decreased signals in susceptibility-weighted imaging (SWI) indicative of microbleeds, and mottled fluid-attenuated inversion recovery (FLAIR) signals. (Fig.4)
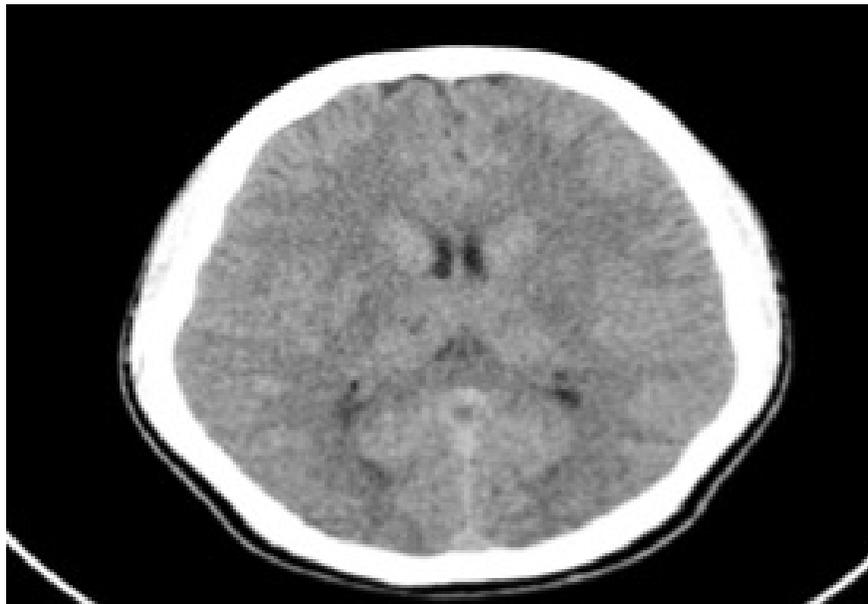

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Figure.3 : The results of the head computed tomography scan showed that there was no obvious bleeding or abnormality in the skull.
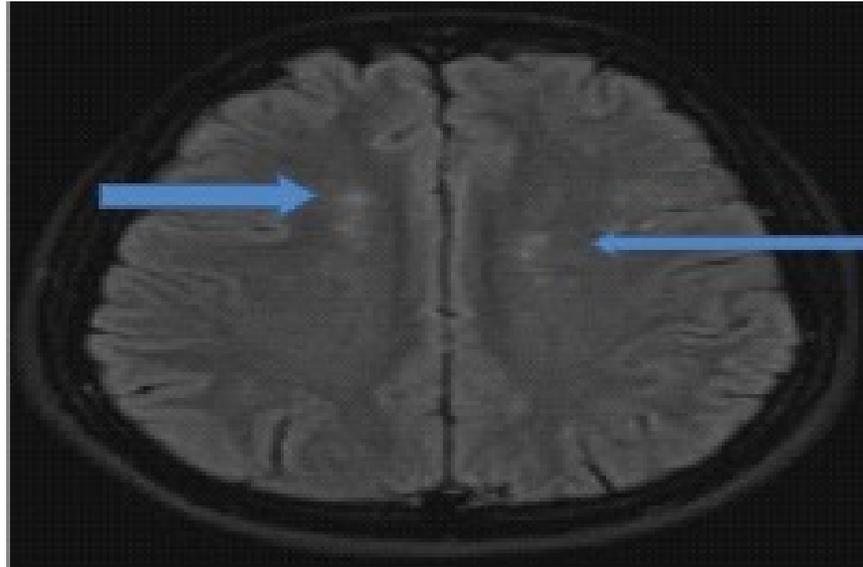

Figure.4: MRI Brain shows extensive bilateral, symmetrical punctuate foci of nearly the same size and abnormal signal intensities giving the appearance of a starfield pattern on T2-weighted images with restricted diffusion.

On the fourth day of admission, bilateral palpebral conjunctival petechial spots also appeared. His pupils were isochoric, and the light reflex was bilaterally positive. There were no meningeal signs. The reflexes and cranial nerves were normal. He could move all 4 limbs. The Glasgow Coma Scale score was 12: E4V2M6. Cardiac echocardiography also revealed no evidence of patent foramen ovale.

Treatment:

One dose of hydrocortisone intravenously on day 1 of admission and subcutaneous heparin was given in view of the pulmonary embolism. His sensorium improved (GCS 15) with supportive measures. Respiratory distress was managed conservatively with oxygen by mask. The patient eventually improved with the same line of management.

Follow-up:

The patient was discharged with a repeat chest X-ray, which was normal, and hypoxia was quick to resolve with a repeat arterial blood gas (ABG) analysis showing normal parameters with follow-up at a multidisciplinary clinic and rehabilitation program.

Discussions:

Fat embolism syndrome (FES) is most frequently associated with trauma and orthopedic procedures and is characterized by major and minor findings, as defined by Gurd [5]. Three major criteria and two minor criteria marked by (*) in the [Table 1], was manifested in our patient making this a strong case of FES [5,6].

Major criteria	Minor criteria
Respiratory insufficiency* Cerebral involvement* Petechial rash	Tachycardia* > 110 bpm Py
2 major criteria or one major and criteria and four minor criteria suggest a diagnosis of FES	2 major criteria or one majo

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Table 1. Gurd's and Wilson criteria for FES

The highest incidence of FES is among closed, long bone fractures of the lower extremities, especially the femur, and pelvis. Severe burns, infection, kidney transplant, liposuction, cardiopulmonary bypass, and transfusions are other possible causes. Males Age: 10–40 years, multiple fractures, and unstable bone fracture movement are the common risk factors. In our case, a closed long bone fracture of the tibia and fibula followed by joint manipulation makes the patient a high-risk candidate for FES. [2,3]

The onset of clinical symptoms can occur within 12 hours, but symptoms mostly present after 24–72 hours. The classic triad of respiratory manifestations (95 %), cerebral symptoms (60 %), and petechiae (33 %) are seen among patients with FES and are seen in our case [7,8]. Cerebral emboli cause neurological signs in up to 86 % of cases and mainly occur after the onset of respiratory distress, as observed in our patient's clinical manifestation. Neurological manifestations of FES can vary from mild cognitive changes to coma, as evidenced by our patient's Glasgow Coma Scale score of E4V2M6 [9]. Petechial rash, as seen in our patient's conjunctiva, is considered pathognomonic of FES and is reportedly present in up to 60 % of patients [10]. Neurological symptoms are typically accompanied by respiratory failure and skin eruptions. Several cases of isolated cerebral fat embolism have also been reported [11,12]. In our case, respiratory system, skin, and eye examinations were abnormal. The main initial symptom was respiratory distress followed by an alteration in consciousness.

Brain CT showed normal findings in most cases. On the other hand, MRI is more sensitive and can show small high-signal-intensity lesions scattered in the cerebral white matter, cerebellum, and brainstem on T2-weighted or diffusion-weighted images [13,14]. In our case, CT Brain was unremarkable, whereas brain MRI showed bilateral symmetrical punctate foci with restricted diffusion "star field" pattern and mottled fluid-attenuated inversion recovery (FLAIR) signals. Also, a Chest CT scan of the patient showed diffuse bilateral pulmonary infiltrates with consolidation and ground glass opacities. A study by Gupta et al. evaluated 1692 patients with long bone and pelvic fractures and found that 12 patients met the diagnosis of FES. Three were diagnosed with CFE, whereas five suffered multiple bone fractures. Neurological status alterations were seen among all the patients with CFE with T2 and FLAIR hyperintense lesions in the bilateral cerebral hemisphere, basal ganglia, thalamus, pons, and cerebellum [15].

Despite changes in sensorium and respiratory distress, the symptoms resolved with supportive care. Although the initial presentation of CFE may be moderate to severe, most case reports on CFE demonstrate that the cerebral deterioration associated with CFE is amendable [16].

Even though the pathophysiology of FES remains poorly known, mechanical and biochemical theories are the two main proposed theories to explain pathology. Gossling et al. [17] described a mechanical theory, which states that a rise in intramedullary pressure after an injury forces marrow to pass into the injured venous sinusoids causing large fat droplets to be released into the venous system. These fat droplets that travel to the lungs and occlude pulmonary capillaries and systemic vasculature may enter the arterial circulation via a patent foramen ovale or directly through the pulmonary capillary bed, causing the characteristic neurological and dermatologic findings of FES. Similarly, Baker et al. [18] described the biochemical theory, which states that the clinical manifestations of FES are attributable to a pro-inflammatory state. The intermediate products, such as glycerol and toxic-free fatty acids, because of local hydrolysis of triglyceride emboli by tissue lipase, may lead to an injury to pneumocytes and pulmonary endothelial cells, causing vasogenic and cytotoxic edema, leading to the development of acute lung injury or respiratory distress syndrome.

Prevention, early detection, and appropriate treatment are critical parameters in FES. Intracranial pressure and cerebral tissue oxygenation monitoring help maintain optimal perfusion, as suggested by Kumar et al. [19]. Traumatic patients have improved outcomes with splinting and fixation of orthopedic fractures in an early phase, and supportive care remains the mainstay of treatment for FES [20, 21].

Adequate supportive treatment and improved care can help the majority of the patients of FES recover completely. The severity of respiratory problems is a close indicator of the risk of death, with overall mortality for this condition 5–15 % [22]. Most case reports on CFE demonstrate that the cerebral deterioration

associated with CFE is amendable [23-25,16].

Conclusions:

In summary, a high index of suspicion is required to diagnose FES, especially in long bone and pelvic fractures, which are clinically not apparent. CFE should be considered when patients have long bone and pelvic fractures, multiple bone fractures, or deteriorated neurological status. Our case highlights the delayed systemic manifestations of FES and early neurological manifestations that mimic many other diseases like stroke and meningitis. Patients without an intracardiac shunt may also suffer CFE. Additionally, the treating physician should be cautious in excluding pulmonary embolism, which presents similarly but has a different management line. CFE patients may recover completely with appropriate treatment.

CONFLICTS OF INTEREST:

None declared.

AUTHOR CONTRIBUTION:

All the authors contributed equally in drafting, editing, revising and finalizing the case report.

ETHICAL APPROVAL:

The ethical approval was not required for the case report as per the country's guidelines.

CONSENT:

Written informed consent was obtained from the patient to publish this report.

DATA AVAILABILITY STATEMENT:

The data that support the findings of this article are available from the corresponding author upon reasonable request.

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