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Cerebral Fat Embolism Syndrome-case Report

Mehmet Berk Eyinç[1], Dilek Atik[2], Beril Cevizoğlu[1], Boran Polat[2], Habib Ali Yalama[2], Ömer Faruk Demirayak[1]

[1]Karaman Training and Research Hospital, Clinic of Emergency Department, Karaman, Türkiye
[2]Karamanoğlu Mehmetbey University Faculty of Medicine, Department of Emergency Medicine, Karaman, Türkiye

Abstract

Fat embolism syndrome (FES) is an uncommon complication, typically presenting with respiratory insufficiency following orthopedic trauma. The condition is referred to as cerebral FES (CFES) when neurological symptoms dominate, which is an even rarer form of FES. We present a case of a 21-year-old male who was brought to the emergency department with lower extremity fractures following a traffic accident. The patient experienced a sudden decline in consciousness and developed confusion during his follow-up. An echocardiogram performed in the emergency room revealed hyperechogenic structures in heart chambers, and subsequent magnetic resonance imaging scan showed multiple infarct areas, leading to a diagnosis of CFES. The diagnosis of CFES is challenging due to the non-specific nature of the symptoms and the frequent normality of computed tomography scans. High clinical suspicion is crucial in patients with orthopedic injuries who experience sudden neurological decline.

Keywords: Fat embolism, orthopedic trauma, cerebral fat embolism syndrome

Introduction

Fat embolism syndrome (FES) is a rare complication, typically presenting with respiratory insufficiency following orthopedic trauma (1). Diagnostic criteria for FES were first established by Gurd in 1970 and later modified by Wilson, becoming widely used. He proposed that the diagnosis of FES be established by the simultaneous identification of at least two major clinical findings or one major and four minor clinical findings. The major criteria encompass respiratory insufficiency, neurological disturbances, and petechial rash. Minor criteria include the detection of fat globules in urine or sputum, renal dysfunction, jaundice, retinal changes, pyrexia, tachycardia, unexplained alterations in platelet (PLT) count or hematocrit, and the presence of fat macroglobulinemia (2,3). Other classifications include Lindeque's classification, which focuses on respiratory symptoms, and Schonfeld's classification, which develops a scoring system. The Schonfeld classification is organized so that Petechiae are 5 points, Chest X-ray changes (diffuse alveolar infiltrates) are 4 points, hypoxemia is 3 points, fever (>38 °C), tachycardia (>120 bpm), tachypnea (>30 bpm), and confusion are each given one point (2,4). Regardless of the classification system, the classic presentation includes a triad of respiratory distress, petechial rash, and neurological changes (3). When neurological symptoms dominate, the condition is referred to as cerebral FES (CFES) (5). Early recognition is crucial, especially in orthopedic trauma patients showing sudden neurological decline. Diagnosing CFES is challenging due to the lack of universal criteria, relying instead on clinical assessment and the patient's medical history (6).

Case Report

A 21-year-old male was brought to the emergency medicine department complaining of back and leg pain after a traffic accident. There is no history of drug use other than psychiatric medication. Upon arrival at the emergency depeartment, his vital signs were stabile and glaskow coma scale is scrored 15. conscious, oriented-cooperative, pupillar isochoric, infrared +/+. Abdominal and respiratory examinations are normal. No obvious pathology was observed in the cranial nerve examination. Multiple bone fractures were present in the right lower extremity; muscle strength was not clearly assessed; no obvious muscle strength loss was observed in other extremities. The patient's imaging shows right femur, fibula and tibial shaft fractures (Figure 1), lung

Corresponding Author: Boran Polat MD, Karamanoğlu Mehmetbey University Faculty of Medicine, Department of Emergency Medicine, Karaman, Türkiye
E-mail: borannpolat@gmail.com **ORCID ID:** orcid.org/0009-0006-5436-8849
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contusion, and C6 corpus fracture. No pathological findings were observed in abdominal imaging. No pathological findings were present in the laboratory results obtained during admission to the hospital. Toxicology results were normal. Laboratory data showed a drop in hemoglobin (HGB) during its follow-up at emergency department (HGB: 15.1 g/dL, first HGB: 16.6 g/dL). Also, during his intensive care unit stay, he was reported to have a petechial rash, mostly on the anterior surface of his trunk. There was a decrease in PLT count (PLT: 85K/uL) and a drop in HGB (HGB: 10.1 g/dL, after orthopedic surgery HGB: 14.3 g/dL, reference range: 12-18 g/dL).

The patient, who did not need urgent surgery by the thoracic and neurosurgery departments, had a long-leg splint applied to his right lower extremity and was monitored according to the operation plan, with a Philadelphia neck collar attached. During the follow-up, the patient's vital signs remained stable. Approximately 11-12 hours after admission, he suddenly developed confusion and agitation followed by a rapid decline in the level of consciousness. Thereupon, an emergency bedside ultrasound was performed. Upon the detection of thrombus-like structures at Echo (Figure 2), computed tomography (CT) and diffusion magnetic resonance imaging (MRI) were performed. According to diffusion MRI (Figure 3), focal areas were observed in the localization of both basal ganglia, some of which restrict linear moderate diffusion, and these areas were in the millimeter dimension. In addition, a focal area restricting diffusion in millimeter dimensions was observed in the right half of the pons. Focal area restricting linear moderate diffusion at the

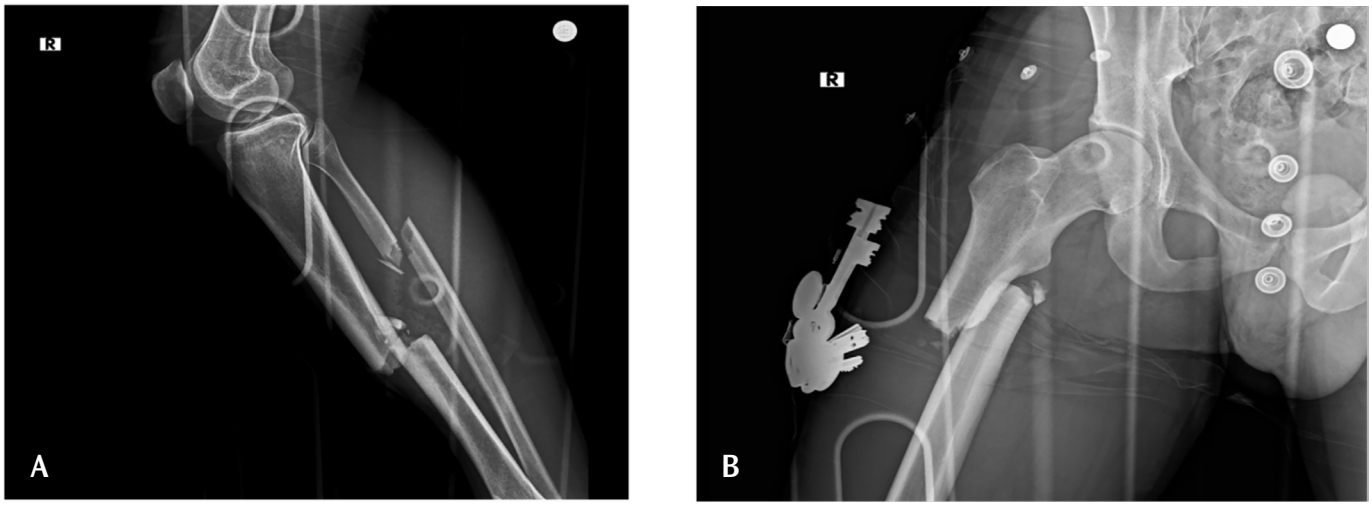

Figure 1. (A) Femur shaft fracture, (B) Fibula and tibial shaft fractures

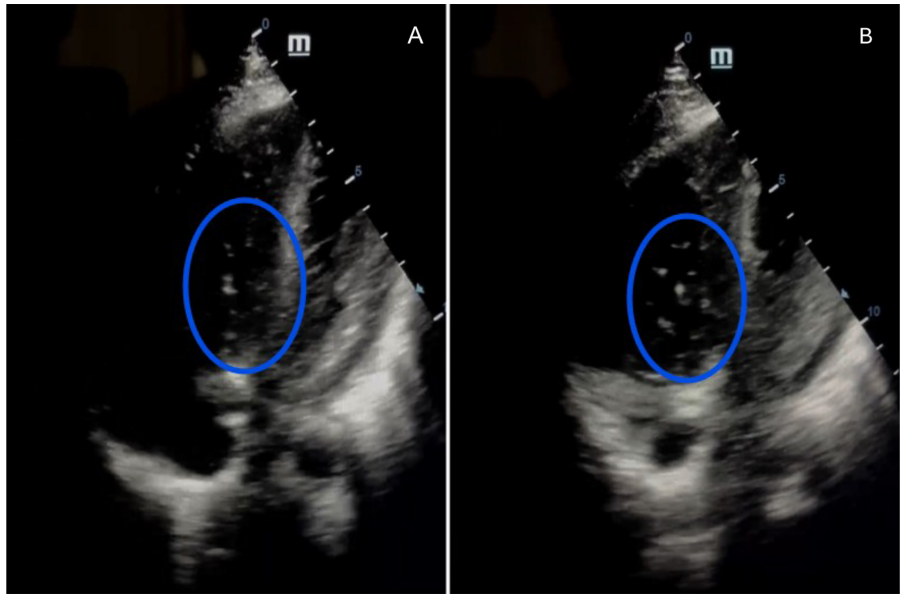

Figure 2. (A, B) Thrombus-like structures highlighted by blue circles on echocardiography

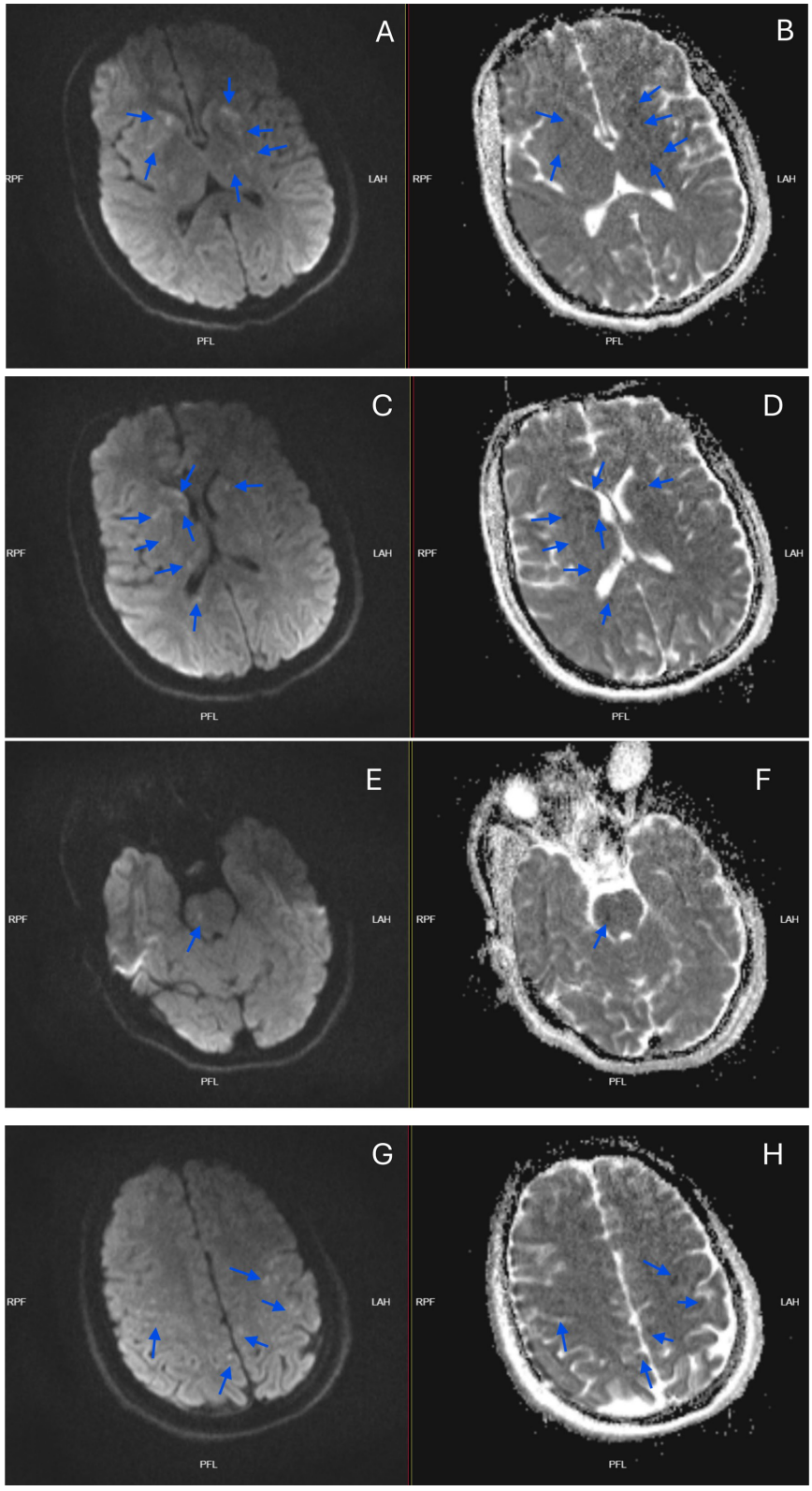

Figure 3. Multiple infarct areas on MRI (starfield pattern). Axial diffusion-weighted imaging (DWI) and corresponding apparent diffusion coefficient images (A–H), demonstrating areas of diffusion restriction indicated by blue arrows

MRI: Magnetic resonance imaging, RPF: Right posterior frontal, PFL: Posterior fronto-lateral, LAH: Left anterior hemisphere

cortical level was observed at the centrum semiovale level, and focal areas restricting diffusion in millimeter size in the frontal and parietal lobes on the left were also observed. MRI findings are consistent with a starfield pattern suggestive of cerebral fat embolism. There is no hemorrhage on CT. Additionally, the patient developed desaturation and underwent a pulmonary CT angiography to rule out pulmonary pathologies, but pulmonary embolism was detected. According to the report, a smear-like effusion was observed in both hemithoraxes, and nonspecific density increases were observed in the dependent segments. With supportive treatment; intensive care follow-up was planned with a preliminary diagnosis of fat embolism.

Discussion

FES is an uncommon disorder primarily characterized by respiratory insufficiency, often arising as a complication following orthopedic trauma (1). Typically presenting within 1-2 days following trauma involving long-bone fractures or orthopedic surgery, it manifests as a petechial rash, progressive respiratory insufficiency, and declining mental status (7). Only a small subset of patients, comprising 0.9% to 2.2%, subsequently develops FES, which is classically characterized by a triad of respiratory symptoms, petechial rash, and alterations in mental status (6). The diagnosis of FES is predominantly clinical; however, the Gurd and Wilson criteria, as well as the Schonfeld fat embolism index, remain two of the most widely utilized and validated diagnostic frameworks (4). In our case, the patient fulfilled 3 major criteria of the Gurd and Wilson Criteria for FES: respiratory distress and petechial rash and neurological symptoms as altered mental status, along with minor criterias, drop in HGB (from 16.6 to 10.1 g/dL), thrombocytopenia (85 K/uL) and according to the Schonfeld fat embolism index, the patient met the following criteria: petechial rash (5 points), and confusion(1 point).

The term "CFES" is used to describe cases where neurological symptoms are the predominant features (5). CFES is a rare variant of FES characterized by isolated neurologic symptoms including ischemic and hemorrhagic strokes, seizures, convulsive and non-convulsive status epilepticus, autonomic impairment, acute encephalopathy, and coma (1,6). Literature reviews and case reports have shown that CFES is common in men with bilateral femur fractures in their early 30s (8).

This diagnosis should be considered in trauma patients with long bone fractures who were found to have confusion or focal neurologic deficits. CFE should also be considered in patients with a history of remote trauma, as in this case, who present with new-onset neurologic symptoms (8).

The hypothesized mechanism is that fat globules enter arterial circulation either through an intracardiac shunt such as a patent foramen ovale, or, more commonly, micro globules filter directly through lung capillaries to reach arterial circulation (6).

MRI utilizing either diffusion-weighted imaging or susceptibility-weighted imaging sequences offers the greatest sensitivity for confirming the diagnosis (9,10). While there are no pathognomonic imaging findings, the "starfield pattern" observed on MRI is the most prevalent and recognized manifestation of CFES (11). This imaging finding is also consistent with the MRI findings in our case, further supporting the possibility of CFES in our patient.

Also, in the study by Maghrebi et al. (12), transthoracic echocardiography showed the "snowstorm" appearance of numerous hyperechoic particles in the inferior vena cava. Bedside echocardiographic evaluation in our patient demonstrated hyperechoic structures suggestive of thrombus formation. There are no definitive or specific treatments of FES; therefore, management is entirely supportive (9).

Considering the multifactorial etiology of FES, early operative fixation of long bone fractures is argued to reduce the incidence of FES, although it cannot be prevented in all patients (13,14).

In our case, we discussed a patient who was admitted to the hospital as a result of a traffic accident and who developed a change in consciousness during follow-up. Based on the clinical, laboratory, and imaging findings, differential diagnoses, such as diffuse axonal injury, disseminated intravascular coagulation, and intraparenchymal, subdural, or epidural hemorrhages, should be considered in trauma patients presenting with altered mental status. However, cerebral fat embolism was considered the more likely diagnosis in our case.

Conclusion

The diagnosis of CFES is challenging due to the non-specific nature of the symptoms and the frequent normal results of CT scans. High suspicion should be maintained in patients with orthopedic injuries who exhibit sudden neurological decline. There are no universal diagnostic criteria for CFES, making the diagnosis dependent on the patient's medical history and clinical manifestations (6).

Ethic

Informed Consent: Written informed consent was obtained from the patient for publication of this case and related images.

Footnotes

Author Contributions

Surgical and Medical Practices: D.A., B.P., H.A.Y., Ö.F.D., Concept: D.A., B.P., Design: D.A., H.A.Y., Data Collection or Processing: M.B.E., B.C., Analysis or Interpretation: M.B.E., B.C., Literature Search: M.B.E., Writing: M.B.E., B.P.

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