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Case Report

Fat Embolism Syndrome: Our Experience

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Abstract:

Fat embolism syndrome is a serious consequence of fat emboli producing a distinct pattern of clinical symptoms and signs. It is most commonly associated with fractures of long bones and the pelvis. Patients usually present with signs and symptoms of multiorgan dysfunction, particularly involving the triad of lungs, brain, and skin. A combination of clinical criteria and MRI brain will enable early and accurate diagnosis of FES. Prevention, early diagnosis, and adequate symptomatic treatment are the mainstay of management. We present a case who became drowsy, after a few hours of a traumatic fracture and developed pulmonary insufficiency, cutaneous petechiae. MRI findings were consistent with cerebral fat embolism. Treatment included intensive care, artificial respiration, and symptomatic therapy. The patient had recovered completely.

Key Word: *fat embolism, clinical criteria, imaging studies, treatment.*

Introduction

The fat embolism syndrome (FES) is a rare clinical condition which needs a high index of suspicion to ensure diagnosis. Zenker first described FES at autopsy in 1862. Von Bergmann diagnosed clinically FES for the first time in 1873. It is usually asymptomatic but may be potentially lethal, complication of long bone fractures.

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Case Report

A 28-year-old male suffered traumatic closed fracture of the right femur that was initially treated conservatively. Fifteen hours after the incidence he was admitted to our hospital for early fixation. During admission he was fully conscious, with no evidence of head injury. Respiratory and hemodynamic status was stable. Twenty three hours after the occurrence, the patient developed sudden shortness of breath with lowering of consciousness. He became confused, unresponsive to verbal stimuli with spontaneous eye opening. The Glasgow Coma Scale was 11/15 (eye opening- 4; motor response- 5; and verbal response- 2). No focal neurologic deficit was observed. He was tachypnic, tachycardic, and cyanosed with SpO_2 81% on room air. ABG analysis showed hypoxemia, with PaO_2 53 mm Hg and $PaCO_2$ 34 mm Hg, with 6L/min oxygen through a re-breather face-mask. He was shifted to the Intensive Care Unit. Full blood count showed anemia with fall of hematocrit, and normal platelet count. Pulmonary embolism was excluded by CT pulmonary angiogram; but there was bilateral pleural effusion with collapse-consolidation. Cerebral CT was normal. Supportive treatment was started with NIV and prophylactic low molecular weight heparin.

On 2nd day after admission he developed a petechial rash on anterior chest and in the axillary regions. On 3rd day his conditions further deteriorated with development of seizure and decorticate posture. He was intubated and put on artificial respiration. Magnetic resonance imaging was performed and showed small hyperintense lesions on T2-weighted images located at subcortical and deep white matter of both cerebral hemispheres including splenium of corpus callosum (figure-1). On diffusion weighted

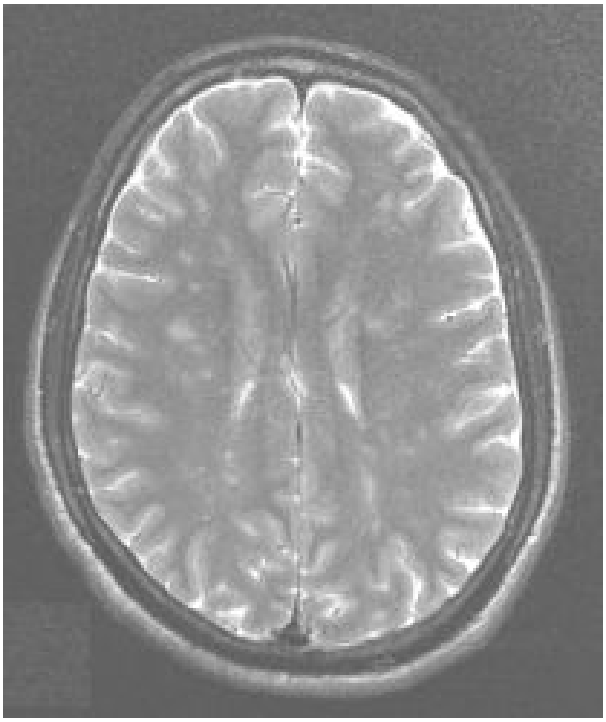

Fig.-1: *T2-weighted Magnetic resonance imaging showed small hyperintense lesions located at subcortical and deep white matter of both cerebral hemispheres.*

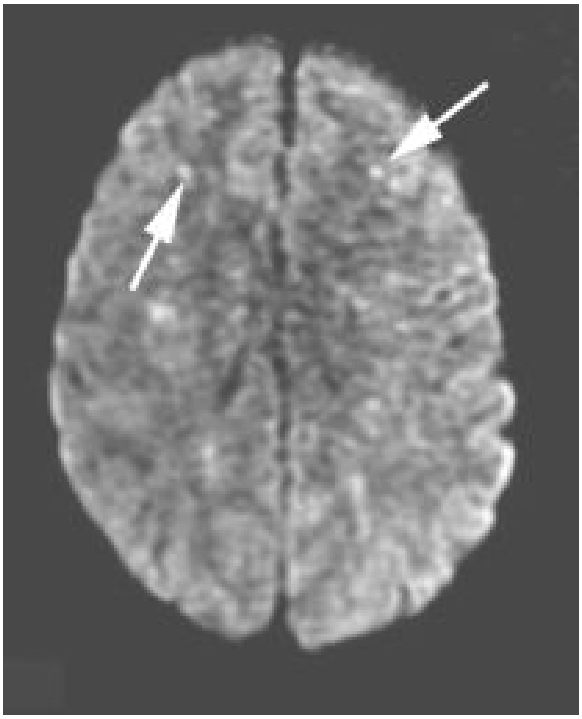

Fig.-2: *Figure 2 Diffusion weighted images (DWI) showed increased signal intensity signifying restricted diffusion producing a "starfield" appearance (arrow).*

images (DWI), these lesions were seen as increased signal intensity dots on a dark background signifying restricted diffusion producing a "starfield" appearance (figure-2). The MRI findings were consistent with the cerebral fat embolism syndrome.

Patient was managed with symptomatic & supportive care. He developed ventilator associated pneumonia with sepsis; and was treated accordingly. He was successfully deventilated on day [11]. His consciousness was gradually improving. He underwent internal fixation of fracture femur under spinal anesthesia on day 14. He was transferred to cabin on day 16, and discharged from the hospital on day 21 in excellent clinical condition. Follow up visit after 2 months he was found to have normal respiratory and neurological function.

Discussion

The fat embolism syndrome (FES) is a rare clinical condition in which circulating fat emboli or fat macroglobules lead to multisystem dysfunction.[1] It is usually asymptomatic, but a few patients will develop signs and symptoms of multiorgan dysfunction, particularly involving the triad of lungs, brain, and skin.[2] The incidence of FES ranges from < 1 to 29%.[1] The incidence of fat embolism syndrome after long bone fractures is 0.9% to 2.2%.[3] The pathogenesis of FES is not clearly understood; mechanical and biochemical causes are described.[4] Fat emboli may occur either by direct entry of depot fat globules from disrupted adipose tissue or bone marrow into the bloodstream in areas of trauma (mechanical) or via production of toxic intermediaries of fat present in the plasma (biochemical). It is feasible that both mechanisms are involved, with embolized fat from traumatized tissues undergoing subsequent biochemical degradation. Intraluminal fat globules smaller than 7 mm in diameter can pass through the pulmonary arteriolar network directly to the brain causing a blockage of capillary blood vessels.[5]

FES typically presents 24 –72 hours after the initial injury. Patients usually present with a classic triad of respiratory changes, neurological abnormalities, and petechial rash. Pulmonary dysfunction is the earliest to manifest and is seen in 75% of patients; it progress to respiratory failure requiring mechanical ventilation in 10% of the cases.[1] The manifestations include tachypnea, dyspnea, and cyanosis. Neurolological changes are seen in 86% of patients with FES.[4] Neurological manifestations are highly variable and nonspecific: headache, lethargy, irritability, delirium, stupor, decorticate posturing, seizure, or coma. Many cases occur as subclinical events and remain undiagnosed.[6] Almost all neurological deûcits are transient and fully reversible. Petechial rash is considered pathognomonic,

although it is present in only 20 – 50% of cases;[7] distributed in the conjunctiva, oral mucous membrane, and skin folds of the upper body, especially the neck and axilla.[8] The rash appears within the ûrst 36 h and is self-limiting, disappearing completely within 7 days. The particular distribution of the rash is related to the fact that the fat particles float in the aortic arch like oil in water and thus get embolized to the nondependent areas of the body.[9] Several other signs are nonspecific, like tachycardia and pyrexia. Renal changes may include lipuria, oliguria, or anuria and hepatic damage may manifest as jaundice. The retina may show exudates, edema, hemorrhage, or intravascular fat globules.[10]

Table-I

Gurd's and Wilson's criteria

Major criteria
Petechial rash
Respiratory insufficiency
Cerebral involvement
Minor criteria
Tachycardia
Fever
Retinal changes
Jaundice
Renal signs
Thrombocytopenia
Anemia
High ESR
Fat macroglobinemia

Table-II

Lindeque's criteria

Sustained pO2 < 8 kpa
Sustained pCO2 > 7.3 kpa
Sustained respiratory rate >35/min, in spite of sedation
Increased work of breathing, dyspnea, tachycardia, anxiety

FES is commonly diagnosed on the basis of the clinical findings along with bio-chemical changes. The most commonly used criteria was proposed by Gurd and Wilson[11] (Table 1); For the diagnosis of FES, at least one major and four minor criteria must be met. Other reliable schemes include Lindeque's criteria based on respiratory feature alone[12] (table 2); and Schonfeld's criteria, a fat embolism index as a semi-quantitative measure to diagnose FES[13] (table 3); a score > 5 is required to diagnose FES.

Table-III

Schonfeld's criteria

	Score
Petechiae	5
X-ray chest diffuse infiltrates	4
Hypoxemia	3
Fever	1
Tachycardia	1
Tachypnea	1
Confusion	1

There are many non-specific biochemical findings; anemia, thrombocytopenia, increased erythrocyte sedimentation rate (ESR), decreased hematocrit, and hypofibrinogenemia.[1] Cytological examination of urine, blood, and sputum with Sudan or Oil Red O staining may detect fat globules that are either free or in macrophages.[4] This test is not sensitive and its absence does not rule out fat embolism.[1] Blood gases will show hypoxia and hypocapnia.

A number of radiological ûndings have been described but none is diagnostic of fat embolism syndrome.[4] The chest X-ray is often normal initially; may reveal increasing diffuse bilateral pulmonary infiltrates, fleck-like pulmonary shadows ('snow storm' appearance), increased pulmonary markings, and dilatation of the right side of the heart.[1] CT pulmonary angiogram findings may be normal. Parenchymal changes consistent with lung contusion, acute lung injury, or adult respiratory distress syndrome (ARDS).[14] Cerebral CT scans are usually negative. MRI is more sensitive and consistently shows multiple small, scattered, nonconfluent hyperintense intracerebral lesions on T2-weighted scans.[6] Signal abnormalities occur in both gray and white matter. Their number correlates with the Glasgow Coma Scale.[15] DW-MRI may enhance the sensitivity and specificity of the neuroradiological diagnosis by the presence of the starfield pattern of scattered hyperintense bright spots on a dark background.[6] Areas of increased signal intensity on T2-weighted scans presumably reflect vasogenic edema, which develops at a later stage, whereas DW-MRI reveals the cytotoxic edema, which develops immediately.

There is no speciûc therapy for FES. Supportive care is the mainstay of therapy for clinically apparent fat embolism syndrome; maintenance of adequate oxygenation and ventilation, stable hemodynamic, blood products if needed,

hydration, prophylaxis of deep venous thrombosis and stress-ulcer, and nutrition. The Mortality from FES is 5 – 15%, but most patients will recover fully.[16, 17]

Continuous pulse oximetry monitoring in high-risk patients may help in detecting desaturation early, allowing early institution of oxygen (and possibly steroid) therapy; it would thus be possible to decrease the chances of hypoxic insult and the systemic complications of FES.[18] Immobilization and early fixation of long-bone fracture is important to prevent or to decrease the severity of FES.[19] Preoperative use of methylprednisolone may prevent the occurrence of FES.[13]

A high index of suspicion is needed to diagnose FES. Any non– head-injured trauma patient who is initially lucid and subsequently develops acute mental status deterioration should undergo immediate evaluation for possible cerebral fat embolism.[20] A combination of clinical criteria and MRI brain will enable early and accurate diagnosis of FES. Preferably DW-MRI of the brain may be the first choice to diagnosis or to rule out cerebral fat embolism. Prevention, early diagnosis, and adequate symptomatic treatment are the mainstay of management.

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