SAS Journal of Medicine ISSN 2454-5112
SAS J. Med., Volume-3; Issue-7 (Jul, 2017); p-177-181
Available online at http://sassociety.com/sasjm/

Case Report

Fatal fulminant fat embolism syndrome: a case report

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Abstract: Fat embolism syndrome (FES) is a multi-organ disorder with potentially serious consequences; it is commonly seen following polytrauma including several long bone fractures. The major clinical features of FES include hypoxia, pulmonary dysfunction, mental status changes, petechiae, tachycardia, fever, thrombocytopenia, and anemia. We report a dramatic and fatal case of a young 23-year-old woman that went on to develop FES six hours after early intramedullary nail fixation of femur and tibia fractures, despite early diagnosis and aggressive supportive therapy.
Keywords: fat embolism syndrome, femur shaft fracture, intramedullary nailing, fulminant, petechiae.

INTRODUCTION

First clinically described by von Bergmann in 1873[1], the fat embolism syndrome (FES) is still somewhat enigmatic. The complex pathogenesis of FES seems to involve both mechanical (fracture, soft tissue injury) and biochemical (activation of plasma lipase, phospholipase A2) factors leading to destabilization of circulating fat and influx of fat to the lungs [2,3]. Progressive respiratory insufficiency, petechial rash and altered mental status, gradually developing in 12h-72 h after injury, are the main clinical manifestations of FES [3]. All of those manifestations may be masked by associated injuries, especially in multiple-injured patients [4], making therefore the diagnosis and the treatment a real challenge. The aim of this work is to remind the ever present risk of FES by presenting fatal and fulminant case of the FES in a young patient with closed fractures of femur and tibia.

CASE REPORT

A 23-year-old woman weighing 66 kg was admitted to emergency room, 30 min after being involved in a car accident (pedestrian hit by car). She has no remarkable past medical history. The initial clinical examination found a well oriented patient, capable of conversation with a Glasgow Coma Scale (GCS) of 15, and haemodynamically stable (initial BP 120 / 60 mmHg, heart rate: 120 beats/min). Closed fractures of the left femoral shaft and right tibia shaft were found. Except for a superficial wound on the forehead, no signs of head injury were detected. Assessment of the abdomen, thorax and pelvis (including chest and pelvis X-ray) did not reveal any signs of injury.

Fractures were temporary immobilized by traction for femoral fracture and left tibia splinting, and the patient was transferred to the orthopedic surgery department of Ibn Sina hospital where, 6 hours after the trauma, she underwent intramedullary nailing surgery of both femur and tibia under general anesthesia. Both femur and tibia were sequentially reamed using flexible reamers to 10.5mm, and then 9mm diameter antegrade nails with proximal and distal locking screws were placed (Figure 1) During surgery, patient was stable, blood loss was estimated to be around 250ml, and total operating time was around three hours.

Immediate postoperative period was with no incident, then 6 hours postoperatively the patient presented polypnea with SpO2 of 94% at ambient air, a low-grade fever (38,3°), tachycardia at 105 beats/min and a neurological impairment with a GCS of 10. Early auscultation, cerebral CT and chest X-ray were all normal.

The patient was moved to intensive care unit (ICU) for further care and investigations where she was intubated because of the respiratory deterioration. Arterial blood gas analysis revealed a pH at 7,3, a PaO2 of 100 mmHg (FIO2 1.0) and a PaCO2 of 45 mmHg, HCO3 : 23. Transthoracic heart and pulmonary ultrasound exams were performed and eliminated pneumothorax and showed neither evidences of an intracardiac shunt nor any direct signs of pulmonary embolism. 7 hours after her admission in ICU, the patient presented patent conjunctival and chest petechiae that spontaneously disappeared later. All

other laboratory results were unremarkable except a thrombocytopenia of 105000/mm.

The patient's clinical condition quickly deteriorated; on neurological level the GCS went to 8 and on respiratory level the Pao2/Fio2 went to 80 in the third day. Later chest X-ray showed lung infiltrates with interstitial syndrome (Figure 2), cerebral CT revealed cerebral edema (Figure 3) and chest CT angiography showed bilateral condensation images. Although the clinical pulmonary infection scores (CPIS) was at 5, the patient was put under empiric antibiotic therapy (Tienam, Amiklin) with ventral ventilation sessions.

At the fifth day, Haemodynamic and respiratory failure developed associated with acute renal failure (creatinine clearance at 33) for which the patient was hemodialysed. Thrombocytopenia at 50000/mm, anemia at 6,9g/dl, prothrombin time at 50% and signs of lower limbs hypoperfusion also developed.

Despite maximal supportive care including transfusion of 4 units of packed red blood cells and 3 units of fresh frozen plasma, ventilatory and inotropic support, the patient died in the sixth day after her injury, in a sever setting of acute respiratory distress syndrome (ARDS) associated with disseminated intravascular coagulation (DIC).

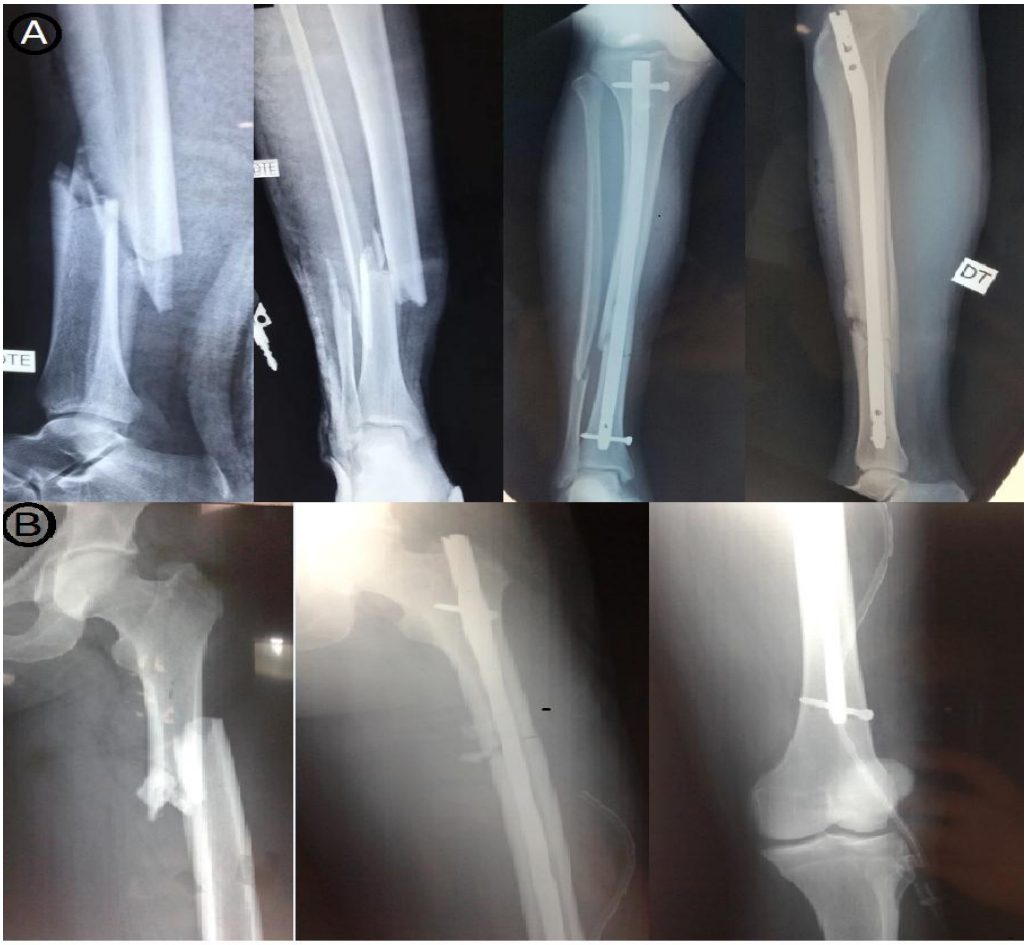

Fig-1: preoperative and post-operative X-Rays of the right tibia (A) and left femur (B). Both fractures were fixed with anterograde intramedullary nails and locking screws

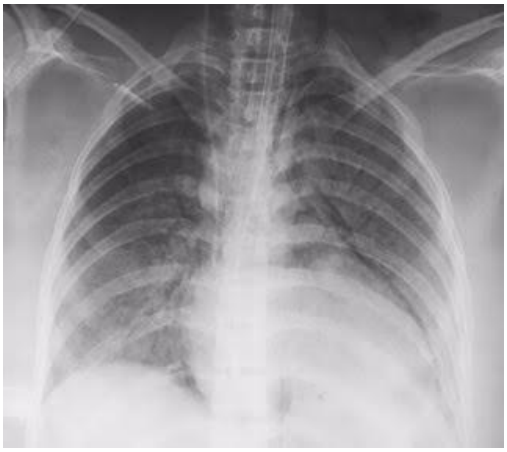

Fig-2: Chest X-Ray showing diffuse bilateral pulmonary infiltrates 10 hours postoperatively

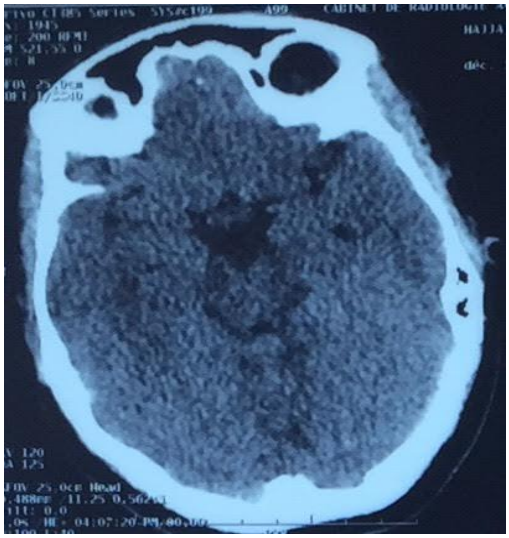

Fig-3: Cerebral CT showing hypodense diffuse white matter changes related to cerebral edema

DISCUSSION

Fat embolism syndrome (FES) is a serious complication most commonly observed following polytrauma including several long bone fractures [4,5]. In patients with a single long bone fracture, the frequency of FES has been reported to be around 3%, whereas it reaches almost 30% in the case of multiple long bone and ⁄or pelvic fractures [5]. The complex pathogenesis of FES seems to involve both mechanical (fracture, soft tissue injury) and biochemical (activation of plasma lipase, phospholipase A2) factors leading to destabilization of circulating fat and influx of fat to the lungs causing ventilation-perfusion mismatching and subsequent acute respiratory distress syndrome (ARDS) [2, 3].

The diagnosis of FES is primarily clinical [6], Its most common presentation is hypoxia (96 percent), which often occurs before pulmonary symptoms develop [7,8]. Cyanosis, tachypnea, dyspnea, and hypoxemia are the main clinical manifestations of pulmonary dysfunction that occurs in 75 percent of FES patients [6]. Hypoxemia has been previously associated with subclinical FES and it is common after long bone fracture [6]. Although authors indicated that the incidence of critical hypoxemia is similar between trauma patient with and without FES [8,9], they recommended that subclinical hypoxia should be monitored closely with continuous pulse oximetry monitoring for earlier detection. Initial pulmonary dysfunction may progress to respiratory failure in 10 percent of patients [6]. Other presenting symptoms include cognitive status changes (59 percent), petechiae, fever, tachycardia, thrombocytopenia, and anemia [6,7]. Our patient was diagnosed with FES after fulfilling Gurd and Wilson's two major and five minor criteria [6.] These clinical signs included diffuse tachypnea with lung infiltrates, petechial rash, tachycardia, pyrexia, sudden anemia, and thrombocytopenia.

This patient was diagnosed with cerebral fat emboli syndrome (CFES) after postoperative cognitive impairment (deterioration of GCS). The incidence of CFES is 0.9 to 2.2 percent [10]. Encephalopathy is the hallmark for diagnosing cerebral embolism syndrome in the setting of pulmonary symptoms [11]. Cerebral signs include headache, irritability, stupor, convulsions, and

coma. Less common findings include apraxia, hemiplegia, scotoma, anisocoria, and conjugate eye deviation [11,12].

Although non-specific, Chest X-rays can show diffusely increased pulmonary images (snow-storm appearance) and right heart dilatation [13,14]. A high-resolution chest CT scan will show bilateral or centrilobular opacities [15].A transthoracic ultrasound can show evidence of an intracardiac shunt, which may predispose patients to develop CFES [16,17]. Our patient developed CFES although no evidence of intracardiac shunt was found, this could theoretically be explained by an increase in pulmonary arteriovenous anastomosis that occurs during periods of exercise and hypoxia [14,18] potentially creating a way for fat emboli to be systemically released.

A bronchoalveolar lavage may assist with the diagnosis of FES by showing neutral lipid concentration [19]. Cerebral CT is usually normal the first one to two days post-injury, it can show hypodense white matter lesions that typically resolve with residual subdural effusion and cerebral atrophy [20]. Brain MRI imaging is the most sensitive imaging technique for diagnosing cerebral fat embolism, and shows multiple hyperintense nodular or punctate foci on T2 sequences as early as four hours after the onset of cerebral fat embolism [21,22].

Prevention and treatment of fat emboli syndrome is based on early fracture management, supportive care, and treatment of shock [6,22]. Albumin has shown its efficacy for volume resuscitation by retaining blood volume and binding fatty acids to decrease lung injury [23].The benefits of using methylprednisone in the prevention and treatment of FES is controversial [6,24]. Although, A meta-analysis of seven double-blind randomized studies and 389 patients with isolated tibia and femur fractures showed that corticosteroids reduced the risk of FES by 78 percent and hypoxia by 61 percent [6,24], prophylactic corticosteroids were not administered to our patient, due to limited clinical evidence in the setting of associated femur and tibia fractures and unknown long-term effects [25-27].

In regards to fracture fixation, it has been shown that there was a higher incidence of FES in patients that received delayed definitive intramedullary fixation [28], especially after ten hours in patients with isolated femur fractures [29]. Based on mechanical pathogenesis theory, numerous techniques and devices have been developed in an attempt to reduce intramedullary pressures such as slow insertion of hollow nails, distal venting, narrower reamers, and reamer irrigator aspirator (RIA) system [22,30,31]. Muller in his study concluded that most of the pressure build-up was related to the diameter of the flexible driver, with significant pressure decreases going from a 9mm to a 7mm diameter driver. In another comparison study, Volgas compared the standard sequential reaming technique with the reamer irrigator-aspirator (RIA) system [31], and concluded that the RIA system reduces intramedullary pressures, but its significant expense and bulkiness limits its widespread use in the orthopaedic trauma surgery. Overall, the modern commonly used reamer systems have allowed reducing the risk of systemic extravasation of medullary fat and subsequent development of FES.

CONCLUSION

Fat embolism syndrome is a relatively rare clinical entity that is most commonly seen in high-risk orthopedic injury, it reflects a multisystem pathology with a possible early onset after trauma and rapid development of fulminant clinical consequences. In this case, there was a dramatic development of FES with cerebral manifestations after definitive closed tibia and femur fracture stabilization. The patient unfortunately went on to cognitive impairment and respiratory failure and subsequent death despite early diagnosis and aggressive supportive therapy.

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