



Contents lists available at ScienceDirect

Chinese Journal of Traumatology

journal homepage: <http://www.elsevier.com/locate/CJTEE>

Case Report

Transient cortical blindness in fat embolism syndrome—a diagnostic enigma

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ARTICLE INFO

Article history:

Received 4 December 2020

Received in revised form

3 February 2021

Accepted 4 February 2021

Available online 9 February 2021

Keywords:

Fat embolism

Cortical blindness

Bone fractures

ABSTRACT

Fat embolism syndrome (FES) is a serious life-threatening manifestation of the fat embolism phenomenon characterized by Bergman's triad of dyspnea, petechiae and mental confusion. While fat embolization into systemic circulation is common, FES occurs in a meagre 0.05%–3% of patients having isolated long bone fractures. Though visual symptoms are commonly attributed to fat embolism retinopathy and is a later occurrence, it may not always be the case. Cortical blindness has been seldom reported in association with FES, and less so as a presenting complaint. Furthermore, no previous literature has described the same in context of an isolated tibia fracture. We report a 20-year-old gentleman with an isolated right tibia shaft fracture who developed sudden onset diminution of vision in both eyes less than 24 h following trauma with no other complaints. Lack of any remarkable ophthalmoscopic findings or other symptoms left us with a diagnostic conundrum. He later went on to develop altered mentation, hypoxia and generalized tonic-clonic seizures with subsequent MRI revealing multiple cerebral fat emboli also involving both occipital lobes. Supportive measures were instituted and his general condition as well as vision gradually improved following which he underwent plate fixation of the fracture under spinal anaesthesia. The perioperative period was uneventful and he was discharged following staple removal. At one month of follow-up, the patient had no residual visual field defects or neurological deficits. Though FES is rare among isolated tibia fractures, this clinical catastrophe may strike in any unsuspected setting thereby warranting a high index of suspicion to ensure early diagnosis and improved patient outcomes.

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Introduction

Fat embolism syndrome (FES) is a life-threatening systemic manifestation of fat embolization into circulation characterized by Bergman's classic triad of dyspnea, petechiae and mental confusion.¹ While fat embolization occurs in almost 67% of patients sustaining trauma, it has been found that only 2%–5% of these patients present with clinical manifestations of FES.^{2,3} The incidence further drops to 0.05%–3% those with isolated long bone fractures.⁴

Respiratory symptoms usually precede cerebral and cutaneous manifestations. Visual symptoms occur in 50% of FES patients and are most commonly due to fat embolism retinopathy,^{2,4} which is a much later occurrence in the disease process. Cortical blindness is rarely reported as the cause of visual symptoms in FES and no

previous literature describes the same in context of an isolated tibia fracture. We report a young male with an isolated tibia shaft fracture who developed cortical blindness as an initial symptom prior to respiratory manifestations.

Case report

A 20-year-old gentleman was brought to the emergency with history of being involved in a road traffic accident and was diagnosed to have an isolated closed mid-shaft tibia fracture of the right leg which was promptly splinted in a long-leg plaster of Paris slab. The patient was hemodynamically stable and a thorough clinical evaluation ruled out any head injury. As per hospital protocol, the patient was admitted to plan for operative intervention and a routine panel of preoperative blood investigations were done, all of which were found to be within normal limits.

The following morning, the patient developed sudden onset diminution of vision in both eyes with only perception of light being present. An emergency ophthalmology consultation was

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Peer review under responsibility of Chinese Medical Association.

sought and the patient underwent fundus examination which ruled out the possibility of fat embolism retinopathy as the retina showed no remarkable findings. He was otherwise alert and able to follow commands and the lack of other symptoms or clinical manifestations (tachypnea, tachycardia, petechiae) did not correspond with the diagnosis of FES.

Within 20 min of the onset of visual symptoms, the patient developed confusion and altered sensorium, with a sudden drop in Glasgow coma scale from 15 to 6 followed by generalized tonic-clonic seizures and bradycardia. He was shifted to the intensive care unit, intubated and given intravenous anti-epileptics, prophylactic antibiotics and subcutaneous enoxaparin. Arterial blood gas analysis at this point showed features of type 1 respiratory failure with a pH of 7.47, PaO₂ of 54 mmHg and PaCO₂ of 30 mmHg. Repeat haemoglobin showed a drop from 143 g/L to 118 g/L which continued to fall serially and stabilized at 102 g/L. Total counts were marginally raised to 12300/cumm and platelet counts dropped to 127000/cumm which eventually normalized. His urine did not contain fat globules, fibrin degradation products were positive (>200 ng/mL) and whole blood D dimer was elevated to 2480 ng/mL.

Pulmonary embolism, cerebral vasospasm and cerebral infarct were among the differential diagnoses; however, by temporal association of trauma the possibility of cerebral fat embolism could not be discounted. An urgent neurology consult was sought and a diffusion weighted MRI of the brain was done once the patient was stabilized. The MRI revealed few scattered focal areas of T2/FLAIR hyperintensity involving subcortical white matter of bilateral frontal, parietal and occipital lobes and in bilateral cerebellar hemispheres showing restricted diffusion suggestive of acute infarcts. The images showed the typical “starfield” appearance described by Parizel et al.⁵ suggestive of cerebral fat emboli (Fig. 1). Concomitant MR angiography showed no evidence of stenosis, occlusion, aneurysm or arteriovenous malformations in the vessels of the brain.

Serial chest radiographs were grossly normal and electrocardiographs showed J-point elevations that were later justified by two-dimensional echocardiogram findings of a prolapsing anterior mitral valve leaflet. Features of pulmonary embolism were absent

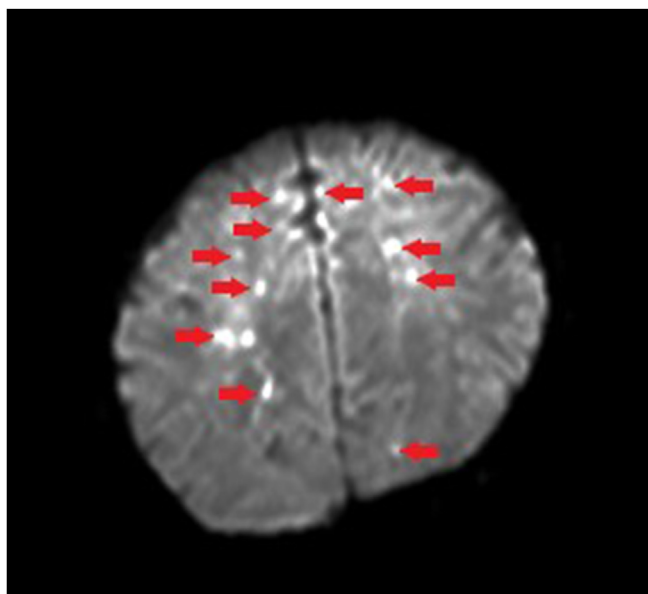


Fig. 1. Diffusion weighted MRI brain showing scattered foci of diffusion restriction involving subcortical white matter in bilateral cerebral hemispheres with typical “starfield” appearance.

on electrocardiograph and echocardiogram. His electroencephalogram was fairly normal barring the presence of background slowing which was attributed to administration of sedatives.

Subsequently, the patient developed oliguria with normal renal profile that resolved with appropriate fluid management. His sensorium improved with supportive measures and he was gradually weaned off the ventilator a week later. Hypoxia was quick to resolve with a repeat arterial blood gas analysis showing normal parameters.

By the end of a week from the onset of symptoms, his visual acuity improved to 6/18 in both eyes and perimetry showed non-specific neurofield defects with normal fundus examination. He had no other residual neurological deficits, and was deemed medically fit for surgery and underwent open reduction and plate fixation of the right tibia using a locking compression plate under spinal anaesthesia (Fig. 2). During the perioperative period, he maintained stable vitals, did not have recurrence of symptoms and was discharged following staple removal. At one month of follow-up, he had no visual field defects or neurological deficits. He had resumed his daily activities and was compliant with rehabilitative efforts aimed at appropriate mobilization of the operated limb.

Discussion

FES is a lethal entity that contributes to an estimated 5%–15% deaths in addition to morbidity, delayed treatment, prolonged hospital stay and need for intensive care facilities.⁶ While treatment is predominantly supportive in nature, the condition warrants early institution of the same to prevent further deterioration of the patient.

In 1974, Gurd and Wilson⁷ described diagnostic criteria for FES with major and minor parameters to aid in identification of the condition and institution of supportive measures. While there are exceptions to every rule, this criterion persists to be the standard against which a clinical diagnosis is made. Lindeque's criteria accounts only for respiratory symptoms, and Schonfeld's scoring system in our patient was <5 on a total of 16 thereby rendering it insignificant.² The modified Gurd and Wilson's criteria is a simplification of the original one that incorporates MRI findings and omits features such as fat macroglobulaemia, hyperbilirubinemia, elevated erythrocyte sedimentation rate (ESR) and fat globules in urine or sputum which are not pathognomonic and are rarely tested in an emergency setting.⁸

Blindness in FES is often attributed to the presence of microvascular changes in the retina on fundoscopic examination characterized by fluffy white exudates, fine streaks of haemorrhage and macular edema.⁷ History of preceding trauma is common and fundus examination may demonstrate fat embolism retinopathy.⁹ These retinal changes are considered one among the minor diagnostic criteria for FES which may be bilateral and present with visual field defects with subsequent complete recovery of vision.

Most often, patients with FES present with respiratory symptoms first, followed by cerebral manifestations and petechial rashes are the last to appear. Respiratory symptoms ordinarily occur in as many as 95% of patients with FES¹⁰; however, in our patient hypoxia was a late finding established on arterial blood gas analysis without tachypnea. Meena et al.¹⁰ described a similar instance wherein their patient presented with cortical blindness and motor aphasia with no appreciable changes on fundus examination after sustaining an isolated segmental fracture of the femur. Their patient went on to develop breathlessness with sudden drop in saturation, tachycardia and ultimately was able to satisfy Gurd and Wilson's diagnostic criteria. The authors drew attention to the atypical chronology of events wherein neurological symptoms had preceded respiratory distress.

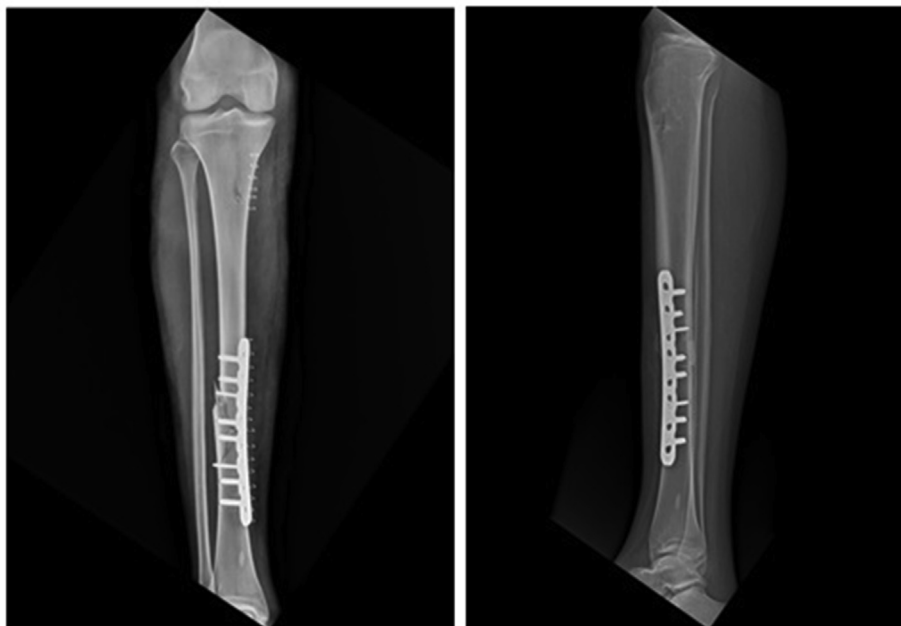


Fig. 2. Postoperative radiograph.

Patients presenting with non-specific visual field defects and normal retinal findings in the context of trauma to a long bone may be considered to have FES on the basis of temporal association of symptoms with history of trauma.¹¹ Lim et al.¹¹ described a similar case wherein a polytrauma patient complained of isolated complete loss of vision in both eyes with no retinal findings. Subsequent MRI revealed infarcts in bilateral occipital lobes along the posterior cerebral artery distribution and the patient gradually had complete recovery of vision. The authors were unable to ascertain whether or not the aetiology of cortical blindness was cerebral fat embolism, and the clinical scenario did not satisfy Gurd and Wilson's criteria. However, the complete recovery of neurologic dysfunction paired with history of preceding trauma were the basis upon which temporal association could be established.

Shahrulazua et al.¹² described a polytrauma patient who developed cortical blindness along with headache, nausea and vomiting following manipulation of a femur fracture. Their patient subsequently developed respiratory distress, deterioration of mental status and petechiae thereby satisfying all major, and additionally, four minor criteria for fat embolism syndrome. While temporal association of symptoms could be established, the chronology of cerebral events preceding respiratory distress was considered atypical.

In our patient, lab parameters showed a significant fall in haemoglobin, with platelet counts that persisted to stay above 100,000/cumm. Fibrin degradation products and whole blood D-dimer levels were deemed insignificant due to history of preceding trauma with tissue injury.^{13,14} Our patient failed to satisfy Lindeque's criteria or Schonfield's scoring system for diagnosis of FES.² While we were ultimately able to make a diagnosis based on Gurd and Wilson's criteria and modified Gurd's criteria after obtaining arterial blood gas reports, the lack of symptoms of respiratory distress and the chronology of clinical manifestations were misleading.

Additionally, the cases described by Shahrulazua et al. and Lim et al. described patients who had sustained fractures to multiple bones, while Meena et al. described a case of cortical blindness associated with an isolated fracture of the shaft of a femur. Isolated

long bone fractures, as in our case, tend to exhibit FES in a meagre 0.05%–3% of patients.⁴ This compounded with minimal displacement of fragments, make it an unsuspected clinical setting for the same.

To sum up, fat embolism syndrome has a variety of clinical presentations which may not adhere to the classic Bergman's triad¹ nor follow any typical chronology of events.⁷ Though respiratory distress is often the most common presenting symptom in FES and visual symptoms are commonly attributed to the presence of fat embolism retinopathy,⁹ it may not always be the case. The diverse symptomatology in combination with the tendency to stereotype the disease may lead to a diagnostic conundrum, delayed treatment and further deterioration. FES is a life-threatening complication of long bone fractures that warrants high index of clinical suspicion to ensure early diagnosis and prompt institution of treatment to improve chances of survival.

Funding

Nil.

Ethical statement

The patient involved gave his written informed consent authorizing use and disclosure of his protected health information and ethical approval was obtained from Father Muller Institutional Ethics Committee.

Acknowledgements

We would like to acknowledge Dr. Lenon J D'Souza for his technical support with regard to radiological findings.

Declaration of competing interest

The authors declare no conflict of interest relevant to this article.

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