Case Report

Neuroimaging findings in a case of cerebral fat embolism syndrome with delayed recovery

Divya Sethi, Shveta Kajal, Anupriya Saxena

Abstract

A young male with multiple lower limb fractures admitted to our Intensive Care Unit was diagnosed with cerebral fat embolism syndrome (FES) based on clinical features and initial magnetic resonance imaging (MRI) which showed multiple hyperintensities on T2-weighted imaging, involving bilateral cerebral and cerebellar hemispheres, predominantly in the watershed territory. The serial MRI done at 3 weeks showed more prominent and larger sized lesions which were in line with the patient's initial low Glasgow Coma Score and indicated severe cerebral insult. The patient responded well to supportive intensive care therapy; his neurological recovery though slow was consistent as he could return to his full functional status after 6 months. The follow-up MRI showed resolution of the most of earlier lesions. This indicates potentially good outcomes even in severe cases of cerebral FES with appropriate medical care.

Keywords: Cerebral, fat embolism syndrome, intensive care, magnetic resonance imaging, neurological

Access this article online

Website: www.ijccm.org

DOI: 10.4103/0972-5229.169350

Quick Response Code:

Introduction

Fat embolism syndrome (FES) is a serious complication of fracture of the long bones. Though the exact incidence of the syndrome is difficult to estimate as many subclinical forms remain unrecognized, the reported incidence varies from 0.5% to 30% in orthopedic trauma with higher rates in multiple long bone fractures.[1-3] The mortality rates range from 5% to 15%; however with early diagnosis and careful management, a great majority of patients recover.

Patients may present with pulmonary, neurological, or dermatological symptoms 24–72 h after initial injury. Cerebral involvement is seen in up to 80% of cases of FES and generally worsens its prognosis. Often respiratory symptoms precede neurological ones, but there are case reports of isolated cerebral involvement. Neurological findings vary from drowsiness to acute confusional state and seizures; decorticate posturing and focal deficits including hemiplegia, aphasia, apraxia, and visual field defects are also seen.[4]

We report a case of a young male who presented with symptoms of severe cerebral FES after a road traffic accident. After treatment, the patient had slow but complete neurological recovery. We also review recent literature on cerebral FES and discuss the role of neuroimaging studies in diagnosing it and predicting its prognosis.

Case Report

A young biker was brought to our hospital's emergency room with multiple fractures in the lower limbs (shaft of the right tibia, inter-trochanter of the right femur,

From:
Department of Anesthesia and Critical Care, Employees' State Insurance Postgraduate Institute of Medical Sciences and Research, New Delhi, India

Correspondence:
Dr. Divya Sethi, A-2B/118-B, Paschim Vihar, New Delhi - 110 063, India.
E-mail: divyasth@gmail.com

How to cite this article: Sethi D, Kajal S, Saxena A. Neuroimaging findings in a case of cerebral fat embolism syndrome with delayed recovery. Indian J Crit Care Med 2015;19:674-7.

shaft of the left femur) after a high-speed collision with a truck on the highway. Initial treatment including intravenous (i.v.) fluids, splinting of fractured limbs, and analgesics was given, and computed tomography scan of the head was done which ruled out head injury. After being stable for initial 8 h, the patient had sudden deterioration in consciousness with labored breathing necessitating intubation and controlled ventilation; he was then transferred to Intensive Care Unit (ICU).

On examination in ICU, the patient had a low Glasgow Coma Score (GCS) with decerebrate posturing and no eye opening on painful stimuli ($E_1M_2V_{ET}$). The pupillary responses were sluggish; he also had tachycardia and tachypnea. The magnetic resonance imaging (MRI) scan showed multiple hyperintensities on T2-weighted imaging (T2WI) in bilateral cerebral and cerebellar hemispheres, predominantly in the watershed territory confirming cerebral FES [Figure 1]. Ventilatory support was continued, i.v. pantoprazole, subcutaneous enoxaparin, midazolam infusion, and antibiotic cover were given. The lower limbs were immobilized with traction weights. In the initial few days, the patient showed hemodynamic instability which was treated with central venous pressure guided fluid therapy, and inotropes were given. Later, in the 1st week, he had seizures for which i.v. phenytoin was started.

At the end of 3 weeks, the patient showed improvement in GCS with eye-opening and withdrawal response of extremities to painful stimulus ($E_2M_4V_{ET}$). A repeat MRI showed more prominent and confluent lesions [Figure 2].

At the end of 4 weeks, tracheostomy was done, and the patient was weaned off the ventilator. Neurological recovery was slow but persistent. At 6 weeks, he had spontaneous eye opening and obeyed verbal commands ($E_4M_6V_{ET}$), but there was motor weakness in all limbs as he could not do movements against gravity (motor power = 2). There was improvement in mental functions as he could recognize his parents, fix visual gaze, and respond with facial gestures to verbal questions.

The patient was then transferred to ward; a repeat roentograms of his lower limbs showed spontaneous reunion of fractured long bones. After 3 months, tracheostomy was decannulated, and he could vocalize. At the end of 4 months, with active physiotherapy, he regained his motor power and was able to move against gravity and resistance (motor power = 4). He was then discharged from the hospital. On follow-up at 6 months, he had complete recovery of motor power, and a repeat MRI scan showed resolution of majority of earlier lesions [Figure 3].

Discussion

The neurological abnormality in cerebral FES occurs from two mechanisms, the occlusive effect of embolized fat globules in the cerebral arterioles and the cytotoxic effect of released free fatty acids causing increase in capillary permeability. These lead to cerebral microinfarcts with edema and microbleed; these lesions are particularly distributed over the watershed territory

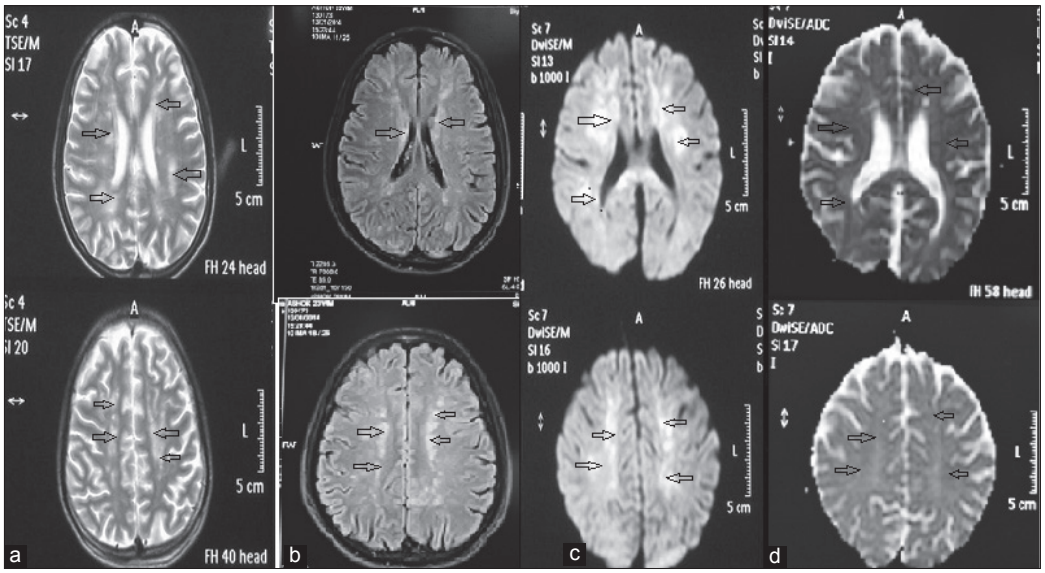

Figure 1: Initial magnetic resonance imaging (day 2). (a) T2-weighted images showing multiple small focal areas of hyperintensities in the deep and periventricular white matter of bilateral cerebral and cerebellar hemispheres, in bilateral corona radiata and basal ganglia. (b) Fluid-attenuated inversion recovery sequence showing hyperintensities in above region. (c) Diffusion-weighted imaging images showing restricted diffusion in the above region. (d) Apparent diffusion coefficient map showing mild reversal in the above region

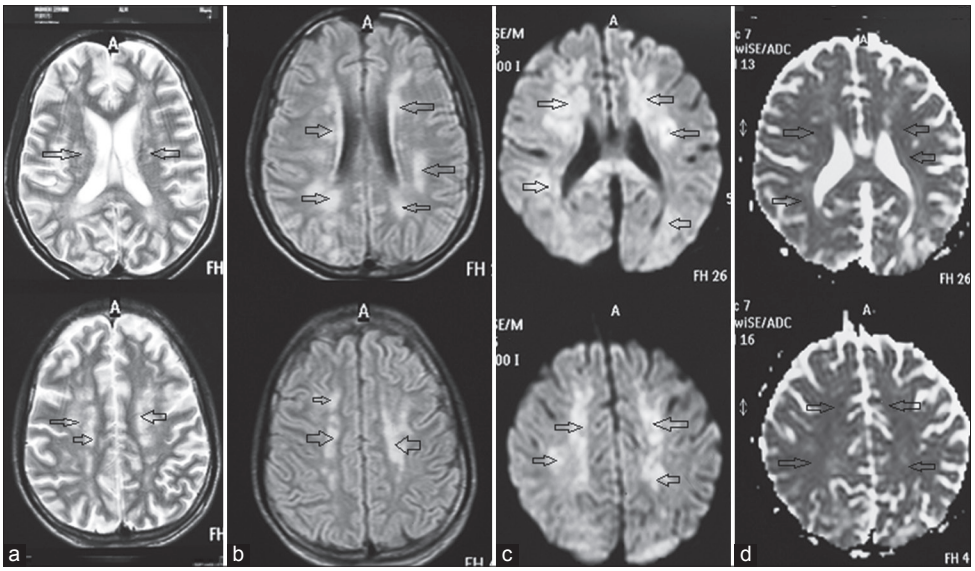

Figure 2: Magnetic resonance imaging at 3 weeks. (a) T2-weighted images showing small focal and confluent areas of hyperintensities seen in the deep and periventricular white matter of bilateral cerebral and cerebellar hemispheres, in bilateral corona radiata and basal ganglia. As compared to initial magnetic resonance imaging, the changes are more prominent in this scan with greater signal alteration. (b) Fluid-attenuated inversion recovery sequence showing hyperintensities in above region. (c) Diffusion-weighted imaging images showing restricted diffusion in the above areas and the appearance of "starfield pattern." (d) Apparent diffusion coefficient map showing mild reversal in the above region

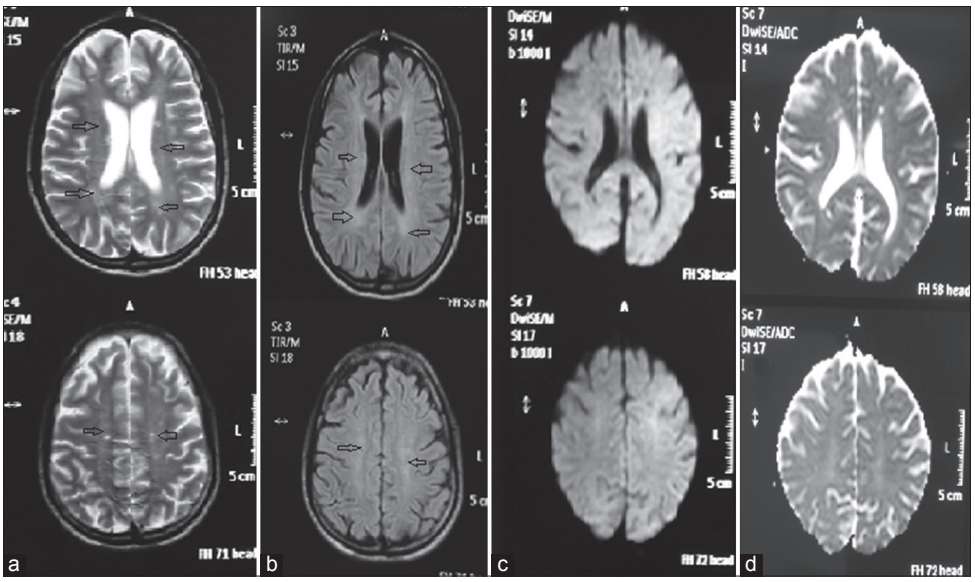

Figure 3: Follow-up magnetic resonance imaging at 6 months. (a) T2-weighted images show the resolution of lesions seen in previous magnetic resonance imaging with only mild confluent hyperintensities in perivascular regions. (b) Fluid-attenuated inversion recovery images show the resolution of lesions in previous magnetic resonance imaging. (c) Diffusion-weighted imaging images are normal and do not show restricted diffusion. (d) Normal apparent diffusion coefficient map images

of the brain. In most cases, the acute microinfarcts and cytotoxic edema are reversible as the emboli are of very small size. However, in some cases of severe cerebral FES, sequelae may occur from persistent ischemia leading to chronic infarcts and neurological deficits.[5]

Though diagnosis of cerebral FES is a clinical one, neuroimaging with MRI is a sensitive and useful tool for confirming it and predicting outcome. The lesions are seen as early as 4 h after neurological deterioration. On T2WI, multiple, diffusely scattered foci of hyperintensities are seen in the deep white matter, basal ganglia, brain stem and cerebellum. These represent microinfarcts in a penumbra of micro-hemorrhages and cerebral edema.[6] In addition, the fluid-attenuated inversion recovery sequence is useful in detecting these lesions in their acute stage and revealing their watershed distribution.[7] On diffusion-weighted imaging (DWI),

the areas of cytotoxic edema appear as a "starfield," i.e., multiple scattered white spots against dark background.[8] In addition, hyperintensities on DWI indicate low diffusivity, appearing as hypointensities on apparent diffusion coefficient map.[9]

Based on the size on T2WI, the lesions can be graded from 1 to 3 as follows: Grade 1 = small spotty hyperintensities, grade 2 = several small spotty or macular hyperintensities, grade 3 = large macular hyperintensities. Takahashi *et al*. showed that the maximum grade of lesion on MRI during the course of cerebral FES correlated with patient's GCS at the time of presentation. However, the initial MRI done soon after onset of cerebral FES may not reveal the true severity of neurological insult as the cerebral lesions take time to develop; hence, serial scans are useful. Furthermore, their study showed that resolution of lesions on MRI coincided with neurological recovery; complete disappearance of lesion indicating good outcome and chronic infarcts, hemorrhages and brain atrophy pointing to poor outcome.[10]

These findings were also seen in our case. Based on clinical features and initial MRI [Figure 1], the patient was diagnosed with cerebral FES, and supportive care therapy started. The serial MRI at 3 weeks [Figure 2] showed grade 3 lesions corresponding with patient's initial low GCS and severe cerebral insult. In most cases of cerebral FES, recovery is seen within 4–6 weeks; however, there are a few reports of prolonged coma and delayed recovery up to 2 months.[11] In our case, the patient responded well to supportive intensive care therapy and his neurological recovery though slow was consistent as he could return to his full functional status after 6 months. The follow-up MRI scan [Figure 3] showed resolution of most of the earlier lesions correlating with his neurological recovery and good outcome.

A high degree of suspicion for cerebral FES is warranted in patients of orthopedic trauma without head injury who develop altered sensorium. Early recognition of cerebral FES is important for appropriate medical management and to avoid adverse outcomes and improve prognosis. Immobilization of the fracture site, maintenance of intravascular volume, hemodynamic, and ventilatory support are the main elements of treatment. Care should be taken to prevent secondary brain injury due to hypoxia, hypotension, or seizures. The case indicates potentially good outcomes even in severe cases of cerebral FES though extended intensive care, and supportive, rehabilitative care may be needed for this.

Acknowledgment

We would like to thank Dr. Anu Singhal and Dr. Mamta Singh from Department of Radiology, Employees' State Insurance Postgraduate Institute of Medical Sciences and Research, for their support in writing this article.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Müller C, Rahn BA, Pfister U, Meinig RP. The incidence, pathogenesis, diagnosis, and treatment of fat embolism. Orthop Rev 1994;23:107-17.
2. Stein PD, Yaekoub AY, Matta F, Kleerekoper M. Fat embolism syndrome. Am J Med Sci 2008;336:472-7.
3. Meyer N, Pennington WT, Dewitt D, Schmeling GJ. Isolated cerebral fat emboli syndrome in multiply injured patients: A review of three cases and the literature. J Trauma 2007;63:1395-402.
4. Jacobson DM, Terrence CF, Reinmuth OM. The neurologic manifestations of fat embolism. Neurology 1986;36:847-51.
5. Butteriss DJ, Mahad D, Soh C, Wall T, Weir D, Birchall D. Reversal cytotoxic cerebral edema in cerebral fat embolism. Am J Neuroradiol 2003;24:97-101.
6. Marshall GB, Heale VR, Herx L, Abdeen A, Mrkonjic L, Powell J, *et al.* Magnetic resonance diffusion weighted imaging in cerebral fat embolism. Can J Neurol Sci 2004;31:417-21.
7. Brant-Zawadzki M, Atkinson D, Detrick M, Bradley WG, Scidmore G. Fluid-attenuated inversion recovery (FLAIR) for assessment of cerebral infarction. Initial clinical experience in 50 patients. Stroke 1996;27:1187-91.
8. Parizel PM, Demey HE, Veeckmans G, Verstreken F, Cras P, Jorens PG, *et al.* Early diagnosis of cerebral fat embolism syndrome by diffusion-weighted MRI (starfield pattern). Stroke 2001;32:2942-4.
9. Kim HJ, Lee CH, Lee SH, Moon TY. Magnetic resonance imaging and histologic findings of experimental cerebral fat embolism. Invest Radiol 2003;38:625-34.
10. Takahashi M, Suzuki R, Osakabe Y, Asai JI, Miyo T, Nagashima G, *et al.* Magnetic resonance imaging findings in cerebral fat embolism: Correlation with clinical manifestations. J Trauma 1999;46:324-7.
11. Gregorakos L, Sakayianni K, Hroni D, Harizopoulou V, Markou N, Georgiadou F, *et al.* Prolonged coma due to cerebral fat embolism: Report of two cases. J Accid Emerg Med 2000;17:144-6.