Cerebral fat embolization with paroxysmal sympathetic hyperactivity syndrome and septic shock at high altitude: A case report and literature review

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Case report

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Abstract

Background

Cerebral fat embolism (CFE) syndrome at high altitude was rare complicated with paroxysmal sympathetic hyperactivity (PSH) syndrome and septic shock. It is a challenge to differential diagnosis and treatment at high altitude.

Case presentation

This case presents a CFE with PSH and septic shock of a 23-year-old man occurred at high altitude of 3800 meters above sea level, transferred by airplane successfully and cured in the department of neurosurgery, Xi’an Tangdu Hospital.

Conclusions

It is key that CFE with PSH can be rapid diagnosed and treatment bundles of septic shock should be initiated as soon as possible. Early neurological rehabilitation played an important role for good outcome.

Background

Cerebral fat embolism (CFE) usually occurs after trauma or during surgical procedures and it occurs in 0.5–3.5% of cases [1-2]. A limited case CFE with paroxysmal sympathetic hyperactivity (PSH) has been reported [3]. We presented a complicated young case of CFE complicated with PSH and septic shock at high altitude.

Case Presentation

A 23-year-old man had a right closed femoral midshaft femur, tibia and fibula fracture (Fig. 1) from a motor vehicle accident on a height of 3800 meter above sea level. He was rescued one hour later and conscious (GCS 15). 4 hours later, he was transferred to local hospital and place a proximal tibial traction pin. 11 hours after the accident, the patient was agitated and lost consciousness with convulsions. GCS was reduced to 6. The head CT scan showed normal (Fig. 2A). 3 days later, he had a fever over 39°C and the convulsions were more frequently than before. The convulsion was controlled by propofol and sodium valproate, but consciousness was not improved (GCS 6) after suspending sedatives. Then, we did the neuroimaging examination for consciousness disorder. The head CT scan indicated brain swelling (Fig. 2B). MRI showed innumerable foci of hyperintense lesions in a "starfield" pattern on T2, FLAIR and DWI sequence images, located at the periventricular, subcortical, basal ganglia, cerebellum and deep white matter predominantly (Fig. 3). The diagnosis maybe difficult to differentiate diffused axion injury (DAI) and CFE if only based on neuroimaging evidence. Finally, CFE was diagnosed based on clinical features and neuroimaging. The reason was illuminated in the discussion section.
His core temperature was over 39°C after 3 days post trauma. The chest CT scan indicated pneumonia (Fig. 4A) and antibiotics was administrated. More seriously, hypotension was presented (85/50 mmHg) and respirate rate was 30/min, indicating that patient got to be septic shock caused by pneumonia. The Sequential Organ Failure Assessment (SOFA) score was 12. As to be more critical than before, he was transferred to Tang Du Hospital by air.

When arrived, he was comatose and convulsive with ictus from 8 to 10 times daily. The manifestation was the intermittent hypertension (blood pressure up to 175/90 mmHg), tachycardia (pulse up to 155/min), febrile (up to 39°C), tachypnea (respire rate up to 45/min) and diaphoresis. Flexor posturing was existed at the same time. The duration of episodes was within 5 minutes. Moreover, continuous electroencephalogram (cEEG) suggested a moderate inhibition of cortex function without epileptiform discharges. PSH was diagnosed based on clinical features, imaging and cEEG. The treatment included propranolol and midazolam. To better control the episodes, fentanyl, dexmedetomidine, and bromocriptine were administered. The episode of PSH was gradually improved, no more than 2 times per day.

His pneumonia was severe and septic shock was present. Chest CT showed multiple lung effusion with pleural effusion 8 days post trauma (Fig. 4B), so thoracic close drainage and mechanical ventilation was performed. The maximum temperature was up to 40 °C, and blood pressure was reduced to 80/55 mmHg. Abnormal laboratory findings included PCT at 8 ng/ml, peripheral leukocyte count of 36 × 10⁹/L (Fig. 5A, 5B). The patient got received anti-sepsis bundles in NICU. Firstly, fluid resuscitation was done (30 ml/kg) within 3 hours and mean blood pressure was increased to 65 mmHg by norepinephrine (10 µg.kg⁻¹.min⁻¹). Secondly, sputum culture was done before imipenem was used initially (1 g intravenous (IV), Q8h). 5 days later, antibiotics was adjusted to vancomycin (1 g IV, Q12h) because result of sputum culture was MRSA. Thirdly, airway management was performed strictly, including turning over and slaping his back for sputum draining, subglottic aspiration and maintain balloon pressure of endotracheal tube up to 25 cmH₂O to prevent aspiration of oral secretion. The pneumonia and septic shock were improved remarkably 14 days post anti-sepsis bundles (Fig. 4C).

After the pneumonia was cured and withdraw mechanical ventilation successfully, the fracture of right femoral shaft, tibia and fibula were surgically fixed by orthopedic surgeon. Then hyperbaric oxygen therapy was supplement to the therapeutic regimen and the patient got conscious gradually.

The patient was awake 90 days post trauma. After 2 months of rehabilitation treatment, motor and language skills are fully restored. Then he was discharged home to continue ambulatory rehabilitation. At the 1 year of follow-up, his modified Rankin Scale (mRS) was 2.

**Discussion**

A typical clinical feature of fat embolism syndrome (FES) was a triad of pulmonary, central nervous system (cerebral fat embolism), and cutaneous manifestations several hours post major trauma [1,4]. Fat
emboli can pass through the pulmonary vasculature, resulting in systemic embolization, most commonly in the brain (CFE) [5].

We presented the complicated CFE case with PSH syndrome and septic shock post trauma. He was cured in NICU by intensive care and rehabilitation treatment and got a good outcome (mRS 2) finally.

CFE is highly variable and nonspecific including headache, lethargy, irritability, delirium, stupor, convulsions, or coma [6]. In this case, we spend some time to differentiate CFE with diffuse axion injury (DAI) or seizures post trauma. Aaron M et al. studied the differences of neuroimaging between CFE and DAI based on MRI analysis according to number, size/shape and the distribution of microhemorrhages. It is found that CFE had significantly more hemorrhages than DAI, particularly in the frontal, parietal and occipital lobes, the corpus callosum and cerebellum. CFE microhemorrhages were punctate/round, whereas DAI hemorrhages were medium sized and linear. DAI is more likely to demonstrate hemorrhages larger than punctate, which is found in CFE. Diffuse confluent diffusion restriction favors CFE, whereas a few scattered foci favor DAI [7]. Our diagnosis of CFE was mainly based on his clinical feature and the MRI feature.

PSH syndrome is similar to seizure post head trauma, sepsis, pulmonary embolism, malignant hyperthermia, overdose of sympathomimetic or anticholinergic agents or autonomic dysreflexia as seen with acute spinal cord injury. Patient's medical history, signs, laboratory results (creatine kinase, serial serum lactate, and repeated blood cultures) and cEEG monitoring can facilitate the diagnosis of PSH. Treatment of CFE with PSH is based on supportive and symptomatic therapy. Opioids, β-blockers, α1-agonists and bromocriptine are some of the options available to manage PSH [3].

Conclusions

This is a complicated case of CFE with PSH and septic shock post trauma at high altitude. It is key that rapid diagnoses and appropriate drug treatment were available as soon as possible. Early neurological rehabilitation played an important role for good outcome.

Abbreviations

CFE: cerebral fat embolism; PSH: paroxysmal sympathetic hyperactivity; FES: Fat embolism syndrome; GCS: glasgow coma score; SOFA: sequential organ failure assessment; cEEG: continuous electroencephalogram; CT: computed tomography; DAI: diffuse axion injury; MRI: magnetic resonance imaging; mRS: modified Rankin Scale; NICU: neurosurgical intensive care unit.

Declarations

Ethics approval and consent to participate
The Ethics approval was gotten from Ethics Committee of Tangdu Hospital. Patient Consent Form has been signed by the patient. The original of the signed form is held by the institution and can be made available to the editors upon request.

**Availability of data and materials**

Not applicable.

**Consent for publication**

Consent Form has been signed by the patient. The original of the signed form is held by the institution and can be made available to the editors upon request.

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**Competing interests**

The authors declare that they have no competing interests.

**Authors’ contributions**

ML, GZ and HG contributed to write the report. ML contributed in clinical treatment of the case. SNG contributed in drafting the manuscript. GDG and YQ contributed in revising the manuscript. All authors read and approved the final manuscript.

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**Figures**

**Figure 1**

X-ray examination images of the right femoral shaft, as well as tibia and fibula obtained 2 hours after the accident.
Figure 2

A. Head CT obtained 11 hours after the accident, the patient was agitated and lost consciousness with convulsions. B. The head CT scan obtained 3 days after the accident, which indicated brain swelling.
Figure 3

MRI scans obtained 3 days after the accident. (FLAIR, DWI, SWI sequence in order)

A

B

C

Figure 4

A. The chest CT scan obtained 3 days post trauma. B. The chest CT scan obtained 8 days post trauma. C. The chest CT scan obtained 22 days post trauma (14 days post anti-sepsis bundles).
Figure 5

A, B show the changes in procalcitonin and white blood cells from 11 days to 28 days post trauma.