ΜΑΖΙΚΟΣ ΛΙΠΩΔΗΣ ΕΓΚΕΦΑΛΙΚΟΣ ΕΜΒΟΛΙΣΜΟΣ ΜΕΤΑ ΑΠΟ ΚΑΤΑΓΜΑ ΙΣΧΙΟΥ ΧΩΡΙΣ ΕΝΔΕΙΞΕΙΣ ΔΕΞΙΑΣ-ΑΡΙΣΤΕΡΗΣ ΔΙΑΦΥΓΗΣ

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ΠΕΡΙΛΗΨΗ

Εισαγωγή: Το σύνδρομο λιπώδους εγκεφαλικής εμβολής αποτελεί μια σπάνια επιπλοκή των καταγμάτων των μακρών οστών με χαρακτηριστικά νευρο-απεικονιστικά ευρήματα. **Παρουσίαση περιστατικού:** Γυναίκα 83 ετών, υποβλήθηκε σε χειρουργική αντικατάσταση του ισχίου λόγω κατάγματος μετά από πτώση. Λίγη ώρα μετά την αποσωλήνωση της, η ασθενής εμφάνισε οξεία αναπνευστική δυσχέρεια και εν συνεχεία έπεσε σε κώμα. Διαπιστώθηκε πετεχειώδες εξάνθημα δέρματος. Η μαγνητική τομογραφία εγκεφάλου ανέδειξε πολυάριθμες, διεσπαρμένες, αυξημένου σήματος, μικρής διαμέτρου, εμβολικού τύπου αλλοιώσεις στις ακολουθίες διάχυσης καθώς και χαμηλού σήματος αλλοιώσεις στις ακολουθίες Grandient Echo εντοπιζόμενες στο σπληνίο του μεσολοβίου και στην παρεγκεφαλίδα. Το διοισοφάγειο υπερηχογράφημα καρδιάς δεν ανέδειξε βατό ωοειδές τρήμα ούτε άλλη διακολπική επικοινωνία. Βάση της οξείας έναρξης της συμπτωματολογίας, της ταυτόχρονης προσβολής του δέρματος, του αναπνευστικού συστήματος και του εγκεφάλου, έγινε η διάγνωση του συνδρόμου λιπώδους εμβολής. **Συμπέρασμα:** Η μαζική λιπώδης εμβολή του εγκεφάλου αποτελεί μια σπάνια και δυνητικά καταστροφική μετατραυματική επιπλοκή η οποία μπορεί να συμβεί ακόμη και μετά από έλασσον τραύμα και χωρίς την παρουσία δεξιάς-αριστερής διαφυγής.

MASSIVE CEREBRAL FAT EMBOLISM AFTER A HIP FRACTURE WITHOUT EVIDENCE FOR RIGHT-TO-LEFT SHUNT

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ABSTRACT

Introduction: Cerebral fat embolism (CFE) syndrome is a rare complication of long-bone fractures with characteristic neuroimaging findings. **Case Report**: An 83-year-old woman underwent total hip replacement surgery after suffering a hip fracture due to a fall. Soon after extubation, the patient acutely developed shortness of breath and eventually fell into a coma. A petechial rash was noted on her skin. Brain-MRI showed numerous, scattered, hyperintense, small-sized, embolic lesions on Diffusion-Weighed-Imaging sequences, and hypointense lesions located in the splenium of the corpus callosum and the cerebellum on Gradient-Echo sequences. Transoesophageal echocardiography did not reveal patent foramen ovale or other interatrial communication. Based on the abrupt onset, simultaneous involvement of the skin,

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brain and respiratory system, fat embolism syndrome was diagnosed. **Conclusion**: Massive CFE is a rare and potentially devastating complication following acute trauma, which may develop even after a minor trauma and without the presence of a right-to-left shunt.

Key Words: cerebral fat embolism, bone fracture, patent foramen ovale, cerebral microbleeds

INTRODUCTION

Cerebral fat embolism (CFE) syndrome is a rare complication of long-bone fractures with characteristic neuroimaging findings.[1] Its incidence is 0.25% to 1.25%, but could be as high as 10% in cases with multiple fractures resulting in unstable pelvic injuries.[2] It is part of the fat embolism syndrome (FES) which was first described by Zenker in 1862.[3] Subsequent studies have shown that FES is more common in patients with multiple at-risk fractures compared to single at-risk fractures.[4] The symptoms develop within 24-48 hours after injury and are related mainly to neurological, respiratory and dermatological sequelae.[2] Fat embolism of the brain occurs in 60% of FES cases, whereas the clinical manifestations vary from mild presentations to coma and death.[5] No specific treatment exists for CES and management is largely supportive.[5] A meta-analysis of 7 clinical studies found that prophylactic administration of corticosteroids might reduce the risk of FES development in patients with long bone fractures. However, the quality of the analysed studies was poor.[6] When CFE is diagnosed, high dose corticosteroid treatment is commonly prescribed to reduce the extent of the cerebral oedema.

CASE PRESENTATION

An 83-year-old woman, after suffering a hip fracture, underwent total hip replacement surgery without complications. Within a few hours after completion of the surgical procedure, the patient developed acutely shortness of breath, became unresponsive, and eventually fell into a coma with a flaccid tetraplegia and a Glasgow Coma Scale of 6 (eye opening and production of sounds after stimulus). An urgent brain Computed Tomography scan was unremarkable. Chest X-ray examination showed bilateral diffuse basal pulmonary infiltrates (**Figure 1A**). Blood tests were within normal values, except from low hemoglobulin levels (10.2 gr/dl), and mildly increased C-reactive-protein (2.6 mg/dl). The Electrocardiogram disclosed sinus tachycardia (>100 bpm). Brain Magnetic resonance Imaging (MRI) showed numerous, scattered, hyperintense, small sized, embolic lesions on Diffusion Weighed Imaging (DWI) sequences, as well as hypointense lesions located in the splenium of the corpus callosum and the cerebellum on Gradient Echo sequences (**Figure 2A-F**). Brain MR-Angiography and Venography excluded the presence of stenoses or occlusion of the large intracranial arteries, whereas cerebral venous sinuses were patent. Interestingly, a petechial rash was also noted on the patients' inner thighs and axillae (**Figure 1B**). To exclude infective endocarditis or other potential embolic cardiac sources, a transoesophageal echocardiography was ordered, which did not disclose evidence for intracardiac thrombi or cardiac valve vegetations. Notably, there was no patent foramen ovale or other interatrial communications. Blood cultures were also negative. Lower limb venous duplex ultrasound was negative for deep venous thrombosis. The Electroencephalogram showed diffuse slowing with an intermittent and unstable alpha rhythm detected in the posterior cerebral regions, as well as bilateral frontal and generalised periodic discharges of biphasic delta waves.

Based on the history of long bone fracture, the abrupt clinical onset, and the simultaneous involvement of the skin, brain and respiratory system, fat embolism syndrome was suspected and subsequently was diagnosed based on the proposed diagnostic criteria by Gurd and Wilson published in 1974, since

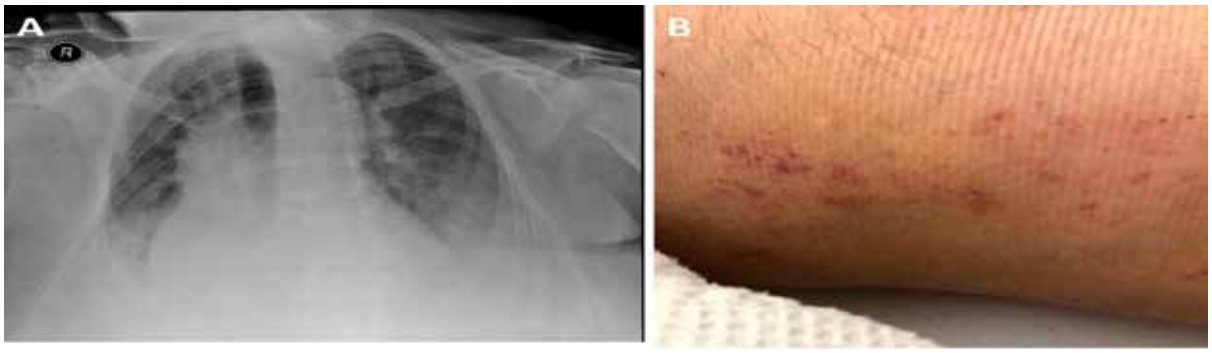

Figure 1. Chest X-ray showing bilateral diffuse basal pulmonary infiltrates (A). Petechial rash on the patients' inner thighs (B).

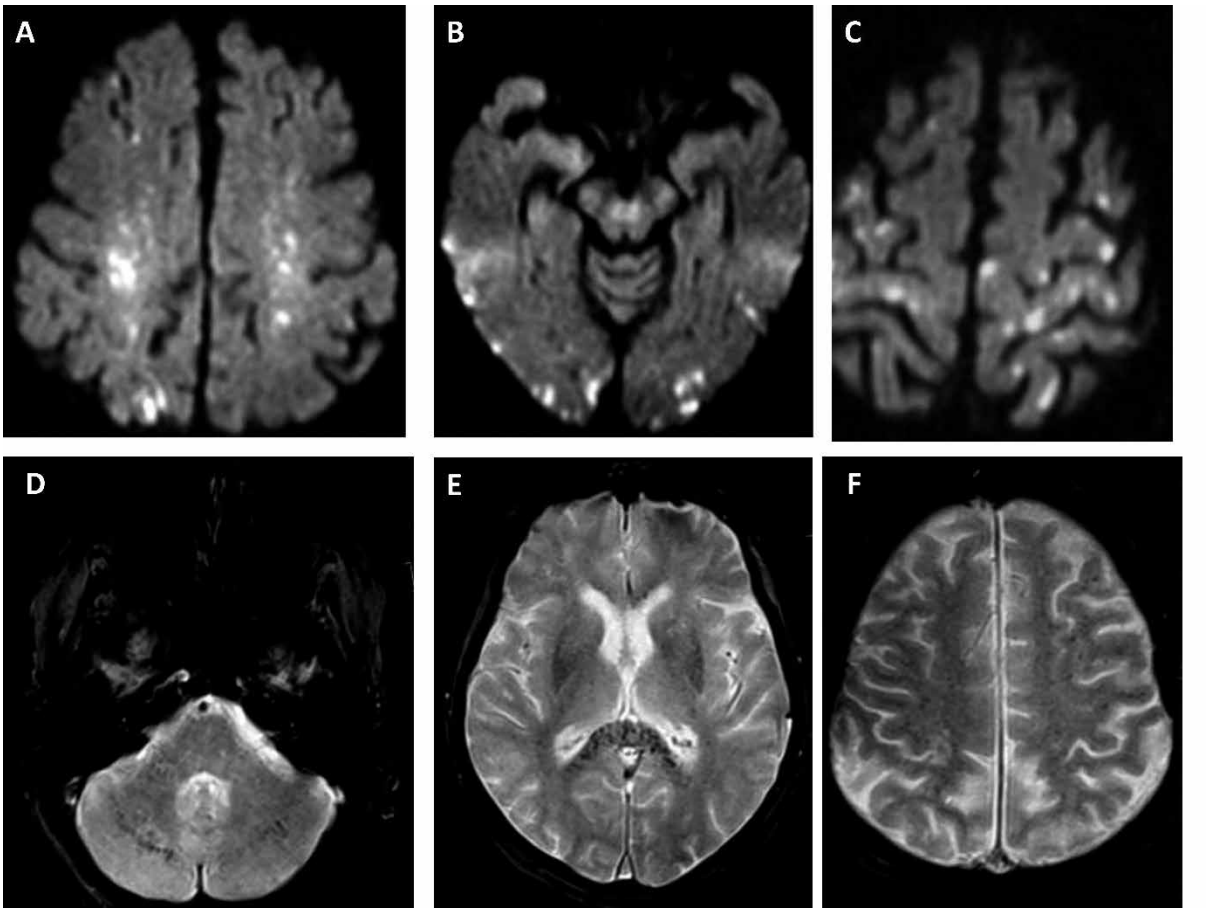

Figure 2. Brain Diffusion-Weighted-Imaging-Sequences showing scattered, hyperintense, embolic lesions (A-C) with a characteristic "starfield" pattern. Brain Gradient-Echo-Sequences showing numerous hypointense foci involving the cerebellum, the splenium of the corpus callosum, and the cortex with a characteristic "walnut-kernel" microbleed pattern, corresponding to petechial cerebral microhaemorrhages (D-F).

the patient fulfilled all three major diagnostic criteria (respiratory distress, cerebral symptoms, petechial rash).[7] The diagnosis was further supported by the characteristic "starfield" pattern of the DWI positive lesions and the simultaneous "walnut-kernel" microbleed pattern on GRE sequences.[1] The patient received empirical treatment with intravenous methylprednisolone infusions, 500mg per day for 5 days. Unfortunately, the patient died 2 weeks later.

DISCUSSION

The pathophysiology of fat embolism syndrome remains controversial. According to one theory, the release of free fatty acids results in an inflammatory response and microvascular disruption.[8] However, the most popular theory states that lipid fragments from the bone marrow, under increased intramedullary pressure, enter the damaged venous sinusoids and are transferred to the lungs where they get embedded into the pulmonary microvasculature. In turn, fat droplets may gain access into the arterial vascular bed and obstruct microvessels of other organs, including the brain. The fat emboli pass into the arterial system through right-to-left shunt, typically through a patent foramen ovale (PFO), or directly through the pulmonary capillary bed.[5] In the absence of PFO, it is believed that a high velocity trauma is required to force the fat emboli to reach the lungs and then pass through the pulmonary microvasculature into the arterial vasculature, and subsequently migrate to the brain.[9,10] Indeed, a recent review found a significant reduction of the incidence of fat embolism syndrome after the age of 30, underscoring the importance of high energy traumas that are far more common in younger ages.[10]

Our case report challenges both concepts regarding CFE pathogenesis. First, CFE may occur without PFO or other type of right-to-left communications. This has been shown in a recent review of all available case reports, where PFO was present in only 12% of patients with CFE, less frequently than the reported incidence in the general population.[10] Second, massive CFE may even occur in the absence of a history of high velocity/high energy impact. Indeed, our patient suffered a simple, partial fall, with pres-

sure from the body-weight exerted briefly on the hip. This low energy impact resulted in a single long bone fracture that was managed acutely and successfully. However, it eventually caused this rare complication of a massive CFE with fatal outcome.

In conclusion, CFE is a rare and potentially devastating complication following acute bone trauma. Clinicians should be aware of this infrequent complication, in order to promptly recognise its symptoms and signs and manage the patients accordingly. Importantly, as also shown by the present case report, massive CFE may even develop after a minor trauma and without the presence of a right-to-left shunt.

DISCLAIMERS

There is no conflict of interest, and no funding related to the present study.

ACKNOWLEDGEMENTS

None.

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