

Unusual Cause of Hypotension in a Polytrauma Victim: A Case of Fahr's Syndrome

Sir,
Shock in polytrauma patient is believed to be due to hemorrhage until proven otherwise.^[1] A 28-year-old male patient presented to the trauma center with a history of blunt trauma to the chest and abdomen following fall from height. He had a medical history of seizure disorder for the past 5 years for which he was on antiepileptic drugs.

On arrival at the emergency department, the patient was conscious, responding but tachypneic (respiratory rate 30 breaths/min) with bilateral equal air entry and in the state of shock (blood pressure was 80/40 mmHg, and heart rate was 130/min). Pupils were bilaterally equal and reactive. He was resuscitated in the emergency department (ED) with 2 L of crystalloids and two units of packed red blood cells.

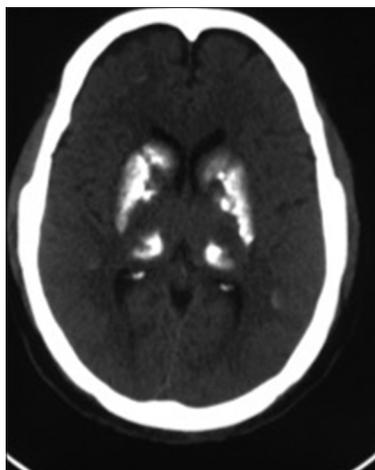


Figure 1: Bilateral basal ganglia and thalamic calcification

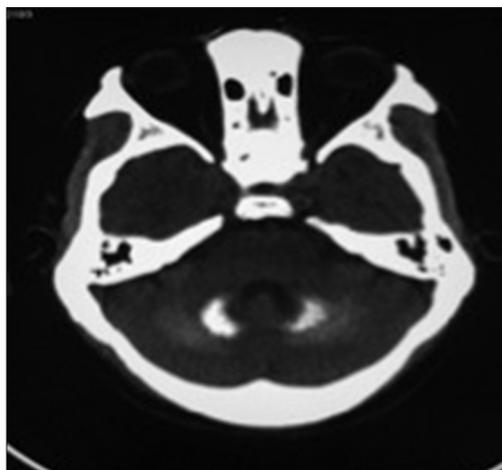


Figure 2: Periventricular calcification in the cerebellum

Chest X-ray, electrocardiography, pneumoscan, and extended focused assessment with sonography for trauma scans did not reveal any obvious abnormality. Noncontrast computed tomography (NCCT) head and cervical spine ruled out intracranial/extracranial bleed or spinal cord injury. Even after adequate fluid resuscitation and blood transfusion, he continued to be in shock requiring multiple vasopressor (noradrenaline and adrenaline) supports. Bedside echocardiography ruled out any cardiac dysfunction/tamponade/embolism. The patient was taken up for emergency exploratory laparotomy to rule out any retroperitoneal bleed or bowel injury. However, laparotomy was negative. Emergency blood biochemistry revealed that the patient had low serum calcium levels (ionized calcium 0.19 mmol/L, total calcium 4.2 g/dl, and total protein 6.8 g/dl). Once intravenous calcium was supplemented, the patient's shock started improving. After 3 days of calcium therapy, the patient was weaned off completely from vasopressors. The cause of low serum calcium levels was investigated. NCCT films were reviewed which showed bilateral basal ganglia and cerebellar calcification [Figures 1 and 2] which were missed initially in the ED. Serum parathyroid hormone (PTH) levels were found to be low (4 pg/ml). The patient's kin gave a history of him being emotionally labile with a history of sudden crying, aggressive behavior, and frequent mood changes. Based on the characteristics of NCCT findings, coexistent neuropsychiatric symptoms, and with hypocalcemia and hypoparathyroidism, a diagnosis of Fahr's syndrome was made.

Fahr's syndrome is a rare neurodegenerative disorder associated with intracranial calcification involving the basal ganglia and dentate nuclei. It is commonly associated with a myriad of parathyroid function abnormalities such as idiopathic hypoparathyroidism, secondary hypoparathyroidism, pseudohypoparathyroidism, pseudopseudohypoparathyroidism, and hyperparathyroidism.^[2] The incidence of idiopathic hypoparathyroidism as found in our case can be as high as 23%. Hypoparathyroidism often leads to metabolic abnormalities of hypocalcemia and hyperphosphatemia, and can lead to calcification at ectopic sites.^[3] Neurological abnormalities,

such as cognitive impairment, seizures, pyramidal signs, and psychiatric features, are often associated with disorder.^[4] In our case, the initial NCCT findings of intracranial calcification were missed in the ED. However, when we investigated the cause of hypocalcemia, coexistent low PTH levels, characteristic personality changes, and history of seizure disorder provided enough clues to lead us to a diagnosis of Fahr's syndrome. The case highlights the importance of looking for calcium levels and evidence of parathyroid dysfunction when faced with intracranial calcification in NCCT head. A missed diagnosis of hypocalcemia led to exploratory laparotomy. The cause of refractory hypotension in our patient was metabolic, which reverted once body calcium stores were replenished.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Access this article online	
Quick Response Code: 	Website: www.ijccm.org
	DOI: 10.4103/ijccm.IJCCM_2_18

How to cite this article: Yadav A, Aggarwal R, Kundu R, Soni KD. Unusual cause of hypotension in a polytrauma victim: A case of fahr's syndrome. *Indian J Crit Care Med* 2018;22:621-3.

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