Unusual presentation of more common disease/injury

Fat embolism syndrome in a child with dystonia musculorum deformans

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A 16-year-old boy with dystonia musculorum deformans underwent an operation for removal of femoral implants and excision of the prominence at the greater trochanter of the left hip. He was found to have fat embolism syndrome at postoperative day 1 as evidenced by confusion, respiratory symptoms, chest radiograph changes, raised erythrocyte sedimentation rate, thrombocytopenia and fat in the urine and sputum.

BACKGROUND

The occurrence of fat embolism syndrome (FES) in paediatric patients is exceedingly rare. Postulations for this variation could be due to the difference in marrow composition as the proportion of haematopoietic tissue to fat in a child's bone marrow is more than that found in adults.¹ Thus, there are only case reports of this condition in paediatric patients which mainly involve spinal fusion or screw augmentations.^{1–3} The following is the first report of FES in a paediatric patient after a minor procedure with removal of femoral implant.

CASE PRESENTATION

A 16-year-old boy has background history of dystonia musculorum deformans (torsion dystonia). The patient had previous proximal femoral valgus osteotomy for the treatment of painful hip subluxation caused by spasticity. Due to impingement of the femoral implants and prominent greater trochanter (figure 1), he underwent removal of implants and excision of the prominence at the greater trochanter of the left hip under general anaesthesia. The implanted locking compression plate was removed along with its accompanying locking screws. Excision of the prominent area of the lateral aspect of the greater trochanter, about 2×3 cm, was performed to relieve the impingement. The decorticated area and screw holes were packed with bone wax to control the bleeding. For anaesthesia, the child was premedicated with oral midazolam. General anaesthesia was induced with remifentanyl, propofol and cisatracurium; maintained with air, oxygen, isoflurane and remifentanyl infusions supplemented with morphine and ketorolac. The patient was stable throughout the operation and after the operation he was monitored closely in the postanaesthesia care unit and was awake and in stable condition.

On postoperative day (POD) 1, he was alert and responsive during the morning ward round. His vital signs were stable without any supportive treatment. However, 24 h after the surgery, he developed acute confusion in the ward. Clinically he was drowsy and was disorientated to

time, place and person. Immediate medical assessment found that he was in severe dystonia and pain. No seizure was witnessed and no focal neurological signs were present. There was no wound infection indicated by the absence of erythema and discharge. There was no deep vein thrombosis as the calves were soft and not tender to palpation. The patient was afebrile, blood pressure and pulse rate were stable but he had tachypnoea of 20–30 breaths per min. He required 3 litres per min of supplementary oxygen via nasal cannula to maintain an oxygen saturation of 97% (he had oxygen saturation of 90% with 2 litres of oxygen per min).

INVESTIGATIONS

Immediate blood results showed normal blood gases, white blood cell count, haemoglobin, electrolytes and blood glucose. Abnormal blood tests include a raised erythrocyte sedimentation rate (ESR) to 44 mm/h and a 15% reduction in platelet count.

ECG showed sinus rhythm with a heart rate of 80 beats per min. Chest radiograph showed left lung consolidative changes suggestive of pulmonary oedema, atelectasis and pneumonia (figure 2). The sputum culture and blood culture were negative for any bacterial growth. EEG and cerebral spectral array (CSA) showed no seizure activity and CT brain showed a tiny hypodense lesion at the right basal ganglia and internal capsule indicating either an old cerebral insult or resultant from previous deep brain stimulation. Lumbar puncture was also done and found no evidence of CNS infection or fat. However, urine and sputum were positive for fat.

DIFFERENTIAL DIAGNOSIS

Acute confusion on POD 1 in this patient included drug effect (sedation), seizure, sepsis and FES. Drug effect is unlikely as the usual anaesthesia drugs were used and the dosage was within safety limits. There was also no change in his usual medication. It is uncommon for pneumonia to develop into sepsis and confusion so early in the postoperative period (POD1) especially with such an acute

Figure 1 Preoperative radiograph showing left femoral valgus osteotomy for the treatment of painful left hip chronic subluxation and femoral locking plate insitu.

presentation of confusion. Furthermore, there was no fever, the blood and sputum culture results were negative, ESR was not significantly increased and white blood cell count was also normal. Deep wound infection was a possible differential diagnosis at the time of presentation but the clinical appearance of the wound was well. A normal EEG and CSA have also effectively ruled out seizure as a cause for the confusion. The CT brain showed a tiny hypodense lesion at the right basal ganglia and internal capsule but it was unlikely to cause any acute cerebral dysfunction. The presence of raised ESR, thrombocytopenia, acute confusion, respiratory symptoms and fat globules in sputum and urine already fulfilled two major and four minor features of FES. Thus, FES was the most likely diagnosis for his acute presentation.

OUTCOME AND FOLLOW-UP

He was immediately transferred to the paediatric intensive care unit (PICU) for close monitoring. He was given supportive care including oxygen via nasal cannula, analgesia, the patient's usual dosage of midazolam for the dystonia, prophylactic antibiotics and chest physiotherapy. Although mechanical ventilation was not required, the oxygen requirement was increased from nasal cannula to venturi mask with a FiO_2 : 0.35–0.5 on POD3. Prophylactic antibiotics were given as we could not effectively rule out acute confusion secondary to postoperative wound infection at the initial presentation. We explored the wound on POD4 due to this uncertainty, but found no signs of infection except a seroma around the site where the implant was removed. We did retrieve a large amount of bone wax that was packed at the osteotomy site. The wound culture from the operation was negative for any organism. Thus, the antibiotics were stopped.

After a few days of supportive treatment he was fully conscious, well orientated, and the dystonia settled. He was discharged from PICU on POD7 with full neurological recovery. The oxygen requirement slowly reduced to

Figure 2 Chest radiograph on postoperative day 1 showing left-sided chest consolidative changes.

nasal cannula (2 litres per min) on POD5 and was weaned off on POD9. Chest radiograph on POD9 showed resolved consolidative changes (figure 3). He was subsequently discharged and without any more hip impingement symptoms.

DISCUSSION

Intramedullary cavity reaming for fixation of long bone fractures leading to increased risk of FES is a well-established concept with a quoted incidence of 0.9–35% and death rate of 1–20% in the literature.^{4–11} Early diagnosis and timely resuscitation to minimise stress response and hypovolaemia is of paramount importance in FES management. The role of steroid and heparin is still controversial at the moment and prophylactic treatment with glucose, alcohol or aspirin are inconclusive as well.¹²

The symptom onset of FES is usually within 12 to 48 h after the triggering event. However, these symptoms could be few and non-specific and thus, laboratory and radiological tests are usually required to strengthen the diagnosis. Positive test findings include the presence of fat globules in blood, urine and bronchoalveolar lavage and also increased pulmonary arterial pressure but none of these investigations are 100% specific.^{12 13}

There is no accepted definition for FES. Rather, it is a clinical diagnosis based on a combination of symptoms. The diagnosis of FES is essentially a clinical diagnosis made by exclusion. The universally accepted diagnostic criteria for FES is the Gurd's criteria.¹⁴ Gurd's criteria is a combination of respiratory, neurological, cutaneous manifestation

Figure 3 Chest radiograph on postoperative day 9 after weaning oxygen supplement therapy showing resolution of consolidative changes.

and laboratory findings.¹⁴ The major features include respiratory symptoms with positive radiological changes, cerebral involvement and the pathognomonic petechial rash. Minor features include tachycardia, fever, retinal changes, urinary changes, drop in haemoglobin levels, thrombocytopenia, high ESR and fat globules in the sputum. A positive diagnosis requires at least one major and four minor features and evidence of fat in the bloodstream.¹⁴

Gurd's criteria had been criticised in the past for underdiagnosing FES.^{9 12} There were various case reports concerning presentations of FES deviating from the Gurd's criteria. For instance, the report of sole clinical manifestation of fat laden macrophages in the bronchoalveolar lavage of a patient with hypoxaemia, which was a non-specific symptom frequently present after any long bone fracture.¹⁵ In addition, there were also reports of FES following spinal arthrodesis and augmentation of spine pedicle screws with bone cement resulting in fulminating pulmonary fat embolism.¹⁻³ There were also reports of isolated cerebral fat emboli syndrome with subtle clinical signs and were diagnosed by MRI and CT changes.¹⁶ Both major and minor features could also be present in other concomitant conditions. In the postoperative setting for example, features such as respiratory changes, tachycardia, fever could indicate atelectasis, and high ESR could indicate inflammatory change after operative trauma.

Our patient did fulfil the diagnostic criteria for FES according to Gurd despite the absence of petechial rash. We performed extensive investigations such as CT, EEG, CSA and lumbar puncture to rule out other causes of the acute confusion. Furthermore, the medication used for anesthesia and sedation did not deviate from the usual practice. Sepsis was unlikely as the presentation of the

confusion was very acute and all cultures were negative for any microorganism. In addition, it was unlikely that a severe sepsis causing confusion would be treated with only 4 days of antibiotics. With the presence of respiratory symptoms and fat substance in the sputum and urine, suggesting circulating fat globules in the bloodstream, FES was established. Two major features namely respiratory symptoms with chest x-ray changes and cerebral involvement were present in our patient. Positive minor features included urinary changes, thrombocytopenia, high ESR and fat globules in sputum. We considered ESR of 44 mm/h and 15% reduction in platelet count in our case to have fulfilled the diagnostic criteria. In the original description of Gurd's criteria, the actual numerical value of high ESR and thrombocytopenia was not provided.¹⁴

The cause for FES in our patient is more difficult to quantify. Our patient underwent an orthopaedic procedure that did not offend the medullary cavity which is usually the source of embolism whereby the intramedullary fat enters the systemic circulation.¹⁶ Moreover, the underlying condition of our patient, dystonia musculorum deformans or torsion dystonia is characterised by sustained muscle contractions and twisting movements with abnormal body postures and not with increased risk of FES or fat in the bloodstream.^{17 18} However we have formulated two postulations for its pathomechanism.

First, the patient positioning may have caused circulatory shifts which may have contributed to the FES. One previous case report discussed the concept of positioning in development of FES.² Spinal fusion surgery may require the patient to lie prone for a certain period of time leading to prolonged compression of the femoral or iliac veins at the pelvis and upper thighs. After turning the patient supine, the sudden relief of venous stasis may contribute to circulatory changes and a sudden massive amount of microemboli may be unleashed.² Although our patient was in the supine position throughout the surgery, he was also wheelchair-bound and there would be an unusual amount of venous stasis in the lower limb preoperatively. The venous stasis could have been aggravated by intraoperative volatile agents such as isoflurane which would contribute to further vasodilatation. The formation of the fat particles in our patient could have originated from the partial osteotomy, trauma from the removal of bone around the plate or from the removal of the plate and screws. The presence of generalised dystonia postoperatively and postoperative dynamic circulatory changes would relieve the accumulated venous stasis leading to massive microembolism, initiating the FES event.

The second possible contributing factor for the development of FES in our patient was the use of bone wax for plugging of the previous screw holes and the osteotomy site. A large amount was used for packing the osteotomy site to stop bleeding from bone marrow cavities. Especially with the large raw bone area after removal of the plate and screws, bone wax plugging could inadvertently precipitate the FES event by increasing the intraosseous pressure. Two animal studies have compared bone cement and bone wax on the development of FES by injection into vertebral bodies.^{19 20} Both studies confirmed the presence of FES in the bone wax group by autopsy with leakage of or bone wax into paravertebral veins and similar quantities of fat emboli in the lungs.^{19 20} The studies found no significant

difference between bone cement or bone wax in causing fat embolism by noting similar reduction in mean arterial blood pressure (40%) and intraosseous pressure (367.8 mm Hg).^{19 20} Although pressurisation of the intramedullary canal will undoubtedly increase the risk of FES, fat material extrusion can still be present with low pressure application.^{3 21} Hence, using bone wax in our patient could still precipitate the FES event.

The complete recovery of our patient without residual neurological deficit suggests that immediate detection of mental deterioration and the aggressive use of supportive treatment to reduce the extent of brain injury can result in good outcome of patient.²²

Learning points

- ▶ FES must be considered as a possible complication even in patients receiving low risk orthopaedic procedures.
- ▶ FES may have non-specific presentations but should be identified early with prompt investigations to rule out other causes of cerebral involvement and respiratory insults.
- ▶ Supportive treatment should also be administered without delay.
- ▶ Clinicians must be wary of susceptible patients (risk of venous stasis).
- ▶ Judicious use of bone cementation or bone wax which, pressurised or not, may precipitate FES.

Competing interests None.

Patient consent Obtained.

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