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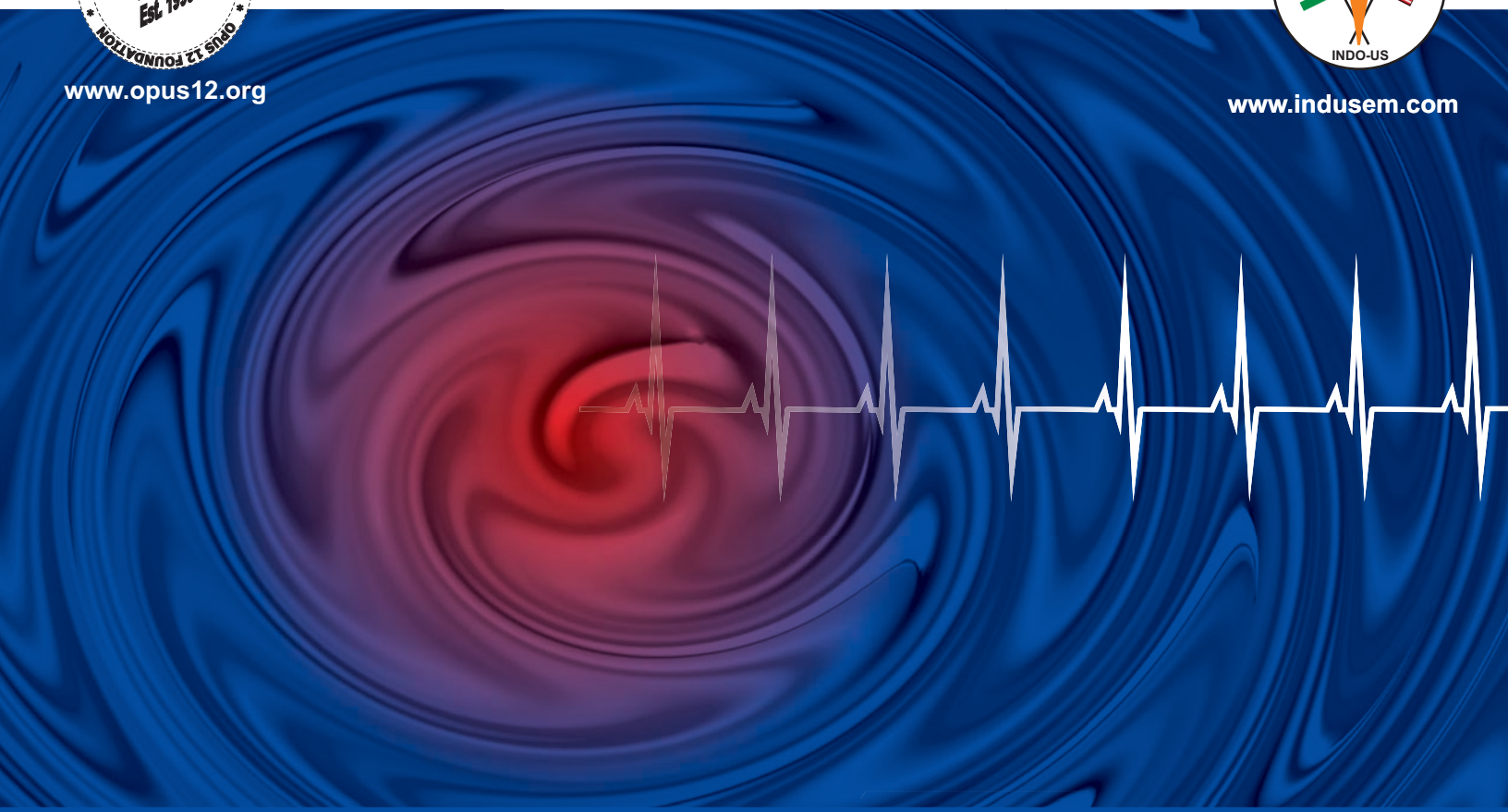

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Case Report

A fatal case of Perthes syndrome

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ABSTRACT

Perthes syndrome, or traumatic asphyxia, is a clinical syndrome associating cervicofacial cyanosis with cutaneous petechial haemorrhages and subconjonctival bleeding resulting from severe sudden compressive chest trauma. Deep inspiration and a Valsalva maneuver just prior to rapid and severe chest compression, are responsible for the development of this syndrome. Current treatment is symptomatic: urgent relief of chest compression and cardiopulmonary resuscitation if needed. Outcome may be satisfactory depending on the duration and severity of compression. Prolonged thoracic compression may sometimes lead to cerebral anoxia, irreversible neurologic damage and death. We report a fatal case of Perthes syndrome resulting from an industrial accident.

Key Words: Cardiac arrest, hypoxic coma, traumatic asphyxia

INTRODUCTION

D'Angers was the first to describe Perthes Syndrome in 1837.[1] A clinical trial involving cervico-facial cyanosis, cutaneous petechiae and subconjonctival haemorrage following thoraco-abdominal compression characterizes Perthes syndrome. Neurological symptoms are common. In 1900, Perthes proposed a physiopathological mechanism including an abrupt rise in superior vena cava pressure during compressive chest trauma.[2] Neurological damage results from combination of decreased cerebral blood flow with asphyxia. We present the case of a 31-year-old patient who developed fatal traumatic asphyxia secondary to an industrial accident.

CASE REPORT

A 31-year-old worker in an air conditioning systems factory presented in cardiac arrest after having his chest compressed between two metal jaws of a hydraulic press. The machine was inadvertently restarted while he was performing maintenance. There were no immediate witnesses to the accident. A colleague found the patient unconscious and not breathing a few minutes later. Cardiopulmonary resuscitation was initiated with a post-collapse interval of several minutes. On-site basic life support by colleagues lasted for 12 minutes before arrival of medical services. Following advanced life support, return of spontaneous circulation occurred after 8 minutes. On arrival at the hospital, he was still unconscious but stable in terms of haemodynamics and ventilation. Physical appearance was characterized by purple cyanotic craniofacial congestion with cervicofacial and subconjunctival hemorrhage [Figure 1].

Total body CT scan revealed a condensation in right lower lobe. He was transferred to the intensive care unit for further medical care. Neurological outcome was poor, because of the occurrence of status epilepticus and post-anoxic encephalopathy. This was correlated with NSE levels in blood of 64.8 ug/l at 48 hrs and evoked potentials showing the absence of cortical response. Brain death was diagnosed and the patient was transferred to operating room for organ donation six days after admission.

DISCUSSION

Perthes syndrome is a rare occurrence. Only 205 cases are reported[3,4] and an incidence of one case per 18,500 accidents.[5] Perthes syndrome is mainly found among victims of car crashes (40%). Other causes are divided between work-related accidents, sports accidents and crushing during crowding or mass movements.[3,6,7] Whatever the cause, a phenomenon of compression of the chest is always found.

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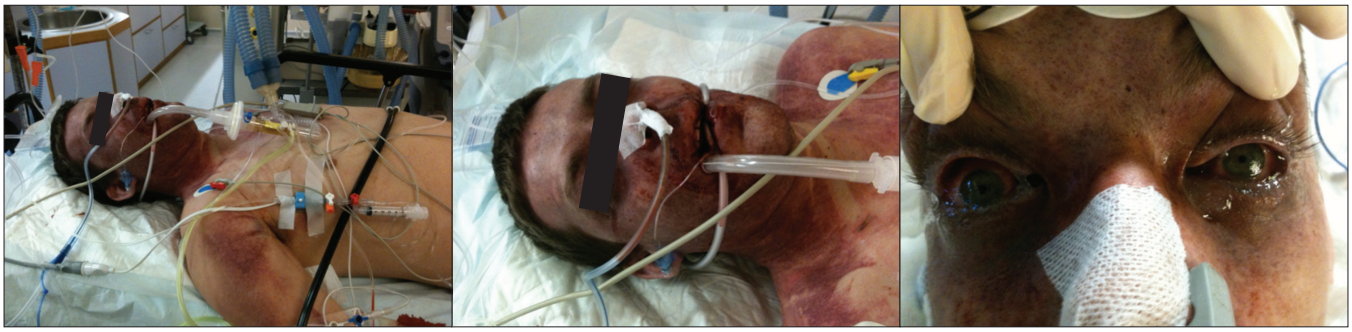

Figure 1: Physical appearance at hospital admission was characterized by purple cyanotic craniofacial congestion with cervicofacial and subconjunctival hemorrhage

The pathophysiologic mechanism deduced by Perthes seems to be applicable regardless of the etiology. The victim perceives the beginning of the compressive force, which triggers a fear response of blocking the thorax in forced inspiration with the glottis closed at the time of chest impact.[8] The compression increases the pressure in the superior vena cava (SVC), causing venous backflow with rupture of capillary venules in the drainage area of the SVC.[3,4,7] The absence of such a phenomenon in dependent territories may be explained by the collapse of the inferior vena cava during the Valsalva maneuver. This would prevent the transmission of the wave of increased venous pressure, protecting the dependent venous territory.

Neurological lesions are common (90%), and result in disorders of consciousness including deep coma.[3,4,6,9] In our case, neurological complications were severe as shown by the Glasgow Coma Score E1VtM3 at admission, progression to status epilepticus, and the high level of NSE in blood at 48 hrs and lack of response to evoked potentials. The frequency of neurological disorders contrasts with the paucity of histological lesions in the brain.[4] Neurological manifestations result from cerebral anoxia as a consequence of reduced cerebral blood flow and the asphyxia secondary to traumatic chest compression. Usually, neurologic manifestations resolve within 24 to 48 hrs with early and appropriate treatment.[9] Poor neurologic outcome in the current case may be explained by the long interval between the injury and first lifesaving first aid.

Cervicofacial cyanosis is found in 95%, cervicofacial petechiae in 84% and subconjunctival hemorrhage in 91% of cases described.[3,8] The presence of ophthalmological findings is almost constant. The severity of these ophthalmologic findings is proportional to the duration and intensity of chest compression.[4]

In addition, nonspecific thoracic lesions have been described.[3,4] Such lesions may include pulmonary contusion, rib fractures,[3,10] hemopneumothorax or diaphragmatic rupture.[6,8]

Myocardial contusions are rare.[10] Transthoracic cardiac ultrasonography of our patient was normal.

The prognosis of Perthes syndrome is variable. Chest compression greater than 10 minutes and the presence of associated injuries are associated with poor prognosis. In our example of fatal Perthes syndrome, a delay in starting cardiopulmonary resuscitation (CPR) and a long duration of CPR were responsible for cerebral anoxia and a poor outcome.

CONCLUSION

Perthes syndrome should be considered in patients presenting with association of an ecchymotic mask with cutaneous petechial haemorrhages and subconjonctival bleeding as a complication of compression of the thorax. Outcome is variable favourable depending on the severity and duration of compression.

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