

Perthes syndrome: a case report

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Abstract

The association of cervicofacial petechiae and subconjunctival hemorrhages with neurological manifestations defines Perthes syndrome or traumatic asphyxia syndrome. This syndrome appears after sudden and brief posttraumatic thoracic or thoracoabdominal compression on a hyperinflated chest like a Valsalva maneuver. In general, the prognosis is favorable. The accompanying lesions and the length of the chest compression are predictors of the outcome.

INTRODUCTION

Perthes syndrome, also known as post-traumatic superior cava syndrome, is an uncommon condition. It results from a violent and brief thoracic or thoracoabdominal compression on a hyperinflated thorax. This syndrome associates cervicofacial petechiae and subconjunctival hemorrhage with neurological manifestations. The evolution is favorable if the compression is relieved rapidly and cardiopulmonary resuscitation is initiated early, otherwise anoxic encephalopathy with serious sequelae may develop. We report a pediatric case of Perthes syndrome with a favorable evolution.

CASE REPORT

A 3-year-old child, with no particular medical or surgical history, was admitted to the emergency room following a domestic accident: the father, while reversing the car, hit his son who was trapped under the vehicle with thoracic compression. The family quickly discharged the child and brought him to the emergency department two hours after the accident.

On admission, the boy was drowsy; the Glasgow Coma Scale was at 14/15. He was polypneic at 38 cycles/minute with peri buccal cyanosis. The pulmonary auscultation was symmetric, and the oxygen saturation was at 93% on room air and 98% with 3 l/min of oxygen. His blood pressure was 90/50 mmHg, and his heart rate was 130 bpm. He had thoracic and epigastric abrasions. The abdominal examination was normal. We noted diffuse petechial lesions on the face and the chest and bilateral subconjunctival hemorrhage. (Figures 1 and 2)

The blood count, as well as biochemical analyses, were normal. Three hours after the accident, a body scan was performed and only identified pulmonary contusion. The eye fundus was normal. Neurological symptoms disappeared within the first day of admission. The evolution was favorable within 48 hours; the boy was discharged. Cutaneous petechial lesions and the subconjunctival hemorrhage disappeared progressively.



Figure 1: diffuse petechial lesions on the face

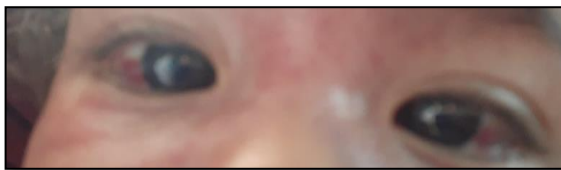


Figure 2: bilateral subconjunctival hemorrhage

DISCUSSION

Olliver first described Perthes syndrome, also known as post-traumatic superior cava syndrome, in 1837. It associates petechiae, subconjunctival hemorrhage, and neurological manifestations [1]. The incidence of Perthes syndrome is low. Some authors report an incidence of one case per 18,500 accidents [2, 3]. It is mainly reported in road accidents when the victim is trapped between the seat and the dashboard (40%). The other causes of Perthes syndrome include work accidents, sports accidents, crushing during jostling, and child victims of sexual abuse. Whatever the traumatic cause is, the common mechanism is thoracic compression or crushing [3].

In 1900, Perthes elucidated the mechanism of the post-traumatic superior cava syndrome [4]. The victim blocks the thorax in forced inspiration with a closed glottis at the onset of thoracic impact because of a panic reaction [5]. Traumatic

compression on a thorax which is already hyper-inflated increases the pressure in the superior vena cava. It results in venous stasis and capillary and venous ruptures in the overlying territories. The fall of the cerebral blood flow and asphyxia secondary to the traumatic thoracic compression induces cerebral anoxia [3]. The absence of venous stasis in the underlying territories can be explained by the collapse of the inferior vena cava within the Valsalva maneuver, protecting the underlying venous territory [6, 7].

The clinical feature of these phenomena is the manifestation of cyanosis, petechiae, and subconjunctival hemorrhage, with respiratory, neurological, and visual manifestations. The duration and intensity of compression influence the occurrence of lesions [8]. These lesions can be associated with other direct lesions related to the trauma itself.

Cutaneous-mucosal lesions, namely cervicofacial cyanosis, cervicofacial petechiae, and bilateral subconjunctival hemorrhage are reported in more than 92% of cases [3]. Our patient presented all these signs.

Ophthalmologic manifestations are always present. The patient can describe a decrease in visual acuity, diplopia, or scotoma. The examination may reveal anisocoria, traumatic mydriasis, and a decrease in the photo motor reflex. The fundus examination is normal in half of the cases. It may show a retinal hemorrhage, exudates, hyperemia, or retinal edema. These disorders are commonly transient, and they regress completely, but slowly [3]. However, some cases of late blindness due to central retinal

vein thrombosis and optic atrophy have been reported [9]. In our case, the fundus examination was normal.

Neurological damage is the most serious and frequently occurs (90%) in Perthes syndrome. The patient may present consciousness disorders, from agitation to severe coma. These neurological symptoms typically disappear within one to two days [3, 10]. In our case, they disappeared within the first day of admission.

In addition to these specific lesions, other thoracoabdominal lesions, which are non-specific, can be present. They reflect the violence of the trauma and may include pulmonary contusion, rib fractures, hemopneumothorax, joint damage, hemoptysis, or diaphragmatic rupture [11, 12]. Cardiac damage is exceptional. Even when there are intrathoracic lesions present, costal bone lesions can be prevented in children thanks to their high thoracic elasticity [7]. Our patient presented only pulmonary contusion.

Hemodynamic instability can be attached to myocardial contusion, or hemorrhage secondary to a lesion of the large vessels [13]. Abdominal injuries may include hemoperitoneum, digestive hemorrhage, laceration, or perforation of abdominal organs [14]. Genito-urinary lesions, tympanic perforation, hematuria, and medullary injuries were also described [15].

The management of Perthes syndrome should be rapid and start at the site of the accident. Prehospital measures aim to release the thoracic compression and start cardiopulmonary resuscitation early to reduce the risk of anoxic brain injury [3, 11].

Perthes syndrome has a generally positive prognosis, with a 90% chance of survival [3]. The occurrence of severe accompanying lesions and prolonged chest compression for more than ten minutes are predictors of a bad prognosis [14].

CONCLUSION

Perthes syndrome, or post-traumatic superior cava syndrome, is rare. The physician should suspect it when there is an association between an ecchymotic mask and neurological manifestations, occurring after a sudden and brief compression trauma of the thorax. The treatment of Perthes syndrome consists of the rapid lifting of the compression and early cardiopulmonary resuscitation to reduce the risk of cerebral anoxia.

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