

Large epidural hematoma in a child with progeria syndrome complicated by posterior cerebral artery stroke: A case report

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Hutchinson-Gilford progeria Syndrome (HGPS) is a genetic disease in which patients manifest with a prematurely aged appearance and cardiovascular disease due to atherosclerosis. A 13-year-old male patient with HGPS was admitted with a history of trauma in the right temporoparietal region of the head, evolving with headache, worsening of previous left hemiparesis and right blepharoptosis. Head tomography showed a large epidural hematoma at the right parietal-occipital convexity. A craniotomy was performed for drainage of the hematoma. After good evolution, the control tomography showed effective drainage of the hematoma with new ischemia in the right posterior cerebral artery. In this case, the syndrome-related cerebrovascular atherosclerosis possibly contributed to the formation of hematoma and postoperative ischemia.

Keywords: Hutchison-Gilford progeria syndrome, epidural hematoma, Traumatic brain injury

INTRODUCTION/BACKGROUND

Progeria, also known as Hutchinson-Gilford progeria Syndrome (HGPS), is a rare genetic disorder with a global incidence of 1 in 8 million live births(1). Patients usually demonstrate a prematurely aged appearance. Children with HGPS have advanced atherosclerosis with risks of myocardial infarction and stroke. The mortality usually is secondary to heart disease(2).

There are many physical consequences of HGPS that can interfere with an anaesthetic act and neurosurgical procedure(1). Although these patients present as elderly persons with accompanying comorbidities, it is important to interact with them at the appropriate developmental mental level. Because of the extremely low prevalence, experience is limited even in the hands of experienced pediatric neurosurgeons and anesthesiologists(3). We describe here a rare case of a large epidural occipital hematoma in a patient with HGPS complicated by a post-operative stroke of the posterior cerebral artery.

CASE REPORT

A 13-year-old male patient was admitted to our hospital with a history of a fall when he tried to go down just 01 flight of stairs. He was known as a patient with HGPS disease making daily use of lonafarnib (200 mg/day). In 2018 he developed a large stroke in the territory of the

right middle cerebral artery and resulted in a left hemiparesis (Modified Rankin Scale 3).

The crash was over the right temporoparietal side of the head. There were no signals of loss of consciousness. He described a progressive and strong headache, vomiting, and mild weakness in the left arm, worse than the previous status. After 03 hours the patient expressed a right ptosis. The initial Glasgow coma scale, at admission, was 10 and the right pupil was dilated with no foto motor reflex. A computed tomography scan(CT scan) of the head showed a large epidural hematoma centred at the parietal prominence exerting a significant mass effect in a rotational direction toward the midbrain. (Fig.1)

The patient was prepared for emergency surgery. Before the start of anaesthesia, the fiberoptic bronchoscope was checked for an eventuality of use. The patient had a class-3 Mallampati airway score suggesting a difficult airway. Direct laryngoscopy under deep inhalational anaesthesia was followed with the placement of the endotracheal tube with the help of a stylet and a tracheal tube introducer (Bougie). There was only one attempt for tube position.

The first observation in the operating room, from the neurosurgical perspective, was the dry stretched-out skin of the scalp with little capability of subgaleal use of lidocaine + epinephrine solution to control bleeding from the skin incision. There were prominent veins at the scalp and



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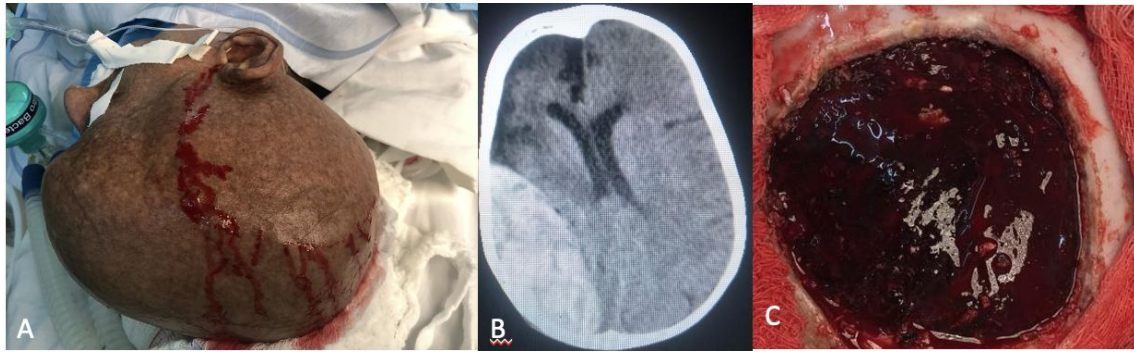


Figure 1- Proeminent veins and alopecia of the progeric patient before the skin incision(A); CT scan showing a large epidural hematoma with shift line deviation(B); Intraoperative aspect of the epidural hematoma after craniotomy

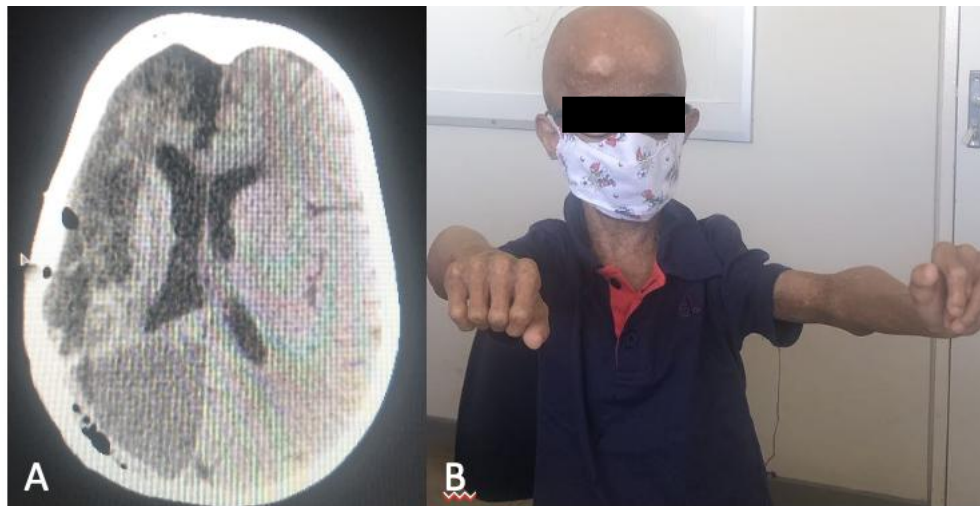


Figure 2 - CT scan one day after surgery showing a new área of ischemia at the right posterior cerebral artery territory.(A); The patient 32 days after surgery(B).

alopecia (Fig.1). The positioning of the head, turned to the left, was easy with no limitations. A “horseshoe” shaped incision was made with the centre projected in the lambdoid suture. Parieto-occipital craniotomy was designed with two burr holes and the hematoma had typical aspects of an epidural hematoma (Fig.1). Venous bleeding was detected in the vicinity of superior sagittal venous sinus which was controlled with gelfoam and surgical .

The patient was extubated after the surgery. CT-scan on the following day showed a good result with minimal residual epidural hematoma (Fig.2) It was clear a new area of ischemia at the right posterior cerebral artery territory reflected a new stroke. The patient did well and was discharged after 04 days. The neurological status during follow-up didn't change from the previous performance although the patient realized a loss in the visual field as a result of the new stroke.

DISCUSSION

HGPS clinically is characterized by phenotypic changes developing in childhood that are generally found in the elderly, including alopecia, subcutaneous fat loss, decreased bone density, and a dramatic form of premature accelerated atherosclerosis(4).

The lamins A and C are structural proteins components of the nuclear lamina, a protein scaffolding network underlying the inner nuclear membrane. The LMNA gene encodes Lamins A and C and silent and sporadic mutations are the main genetic abnormality in HGPS syndrome. The resultant protein, called progerin, causes the disease – Progeria. The genetic abnormality that underlies HGPS was elucidated in 2003(4).

Progerin is present in the cells of non-HGPS patients with coronary artery disease[4]. The use of inhibitors of farnesyltransferase, an enzyme that acts to produce the

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mutant Lamin A, has been studied as a potential treatment to ameliorate some of the devastating manifestations of HGPS. The vasculopathy in HGPS disease is characterized by finds that are present in senile atherosclerosis including loss of medial smooth muscle cells, adventitial thickening, intimal calcification, and atherosclerotic plaquing in the great vessels, small arteries, and arterioles(1, 4).

In our case, a direct consequence of the cerebral herniation and compression at the posterior cerebral artery branches was a secondary stroke resulting in a large area of infarction after the treatment of the Epidural hematoma. Traditionally in cases where the evacuation of the EH is prompt, there is no consequence in the development of stroke or territorial infarction.

Even trivial trauma can result in serious consequences for HGPS patients. Unstable gait, vision defects and skeletal deformities may facilitate the development of traumatic brain injury in typical situations found in children, like falls(5). Manderá et al reported a case in a 10-year-old boy with HGPS in which the severity of the head injury was not only explained by the trauma and was explained by the interaction between the injury and the atherosclerotic changes and characteristics of cerebral arteries in progeric patients(3). The extensive loss of mural smooth muscle cells is one of the main finds in the cerebral arteries of HGPS patients. The response of these cells over the hemodynamic shear stress seems to facilitate ischemic changes(2).

Facing cases of HGPS in an emergency is a challenge because of the low incidence of progeria and lack of familiarity with this condition. The neurosurgeon and the anesthesiologist in this case report were informed of the Progeria disease a few minutes before the surgery, an emergency challenge. The only tool used for intubation was a bougie and the drugs were managed using references of the body weight and not the age of the patient (1). Measures to avoid hypothermia were not made and should be taken in patients with alopecia and a decreased layer of subcutaneous tissue (1). The only concern as a neurosurgeon was the possible difficulty in closing the dura mater, making an analogy with older patients. The knowledge of to be facing the risk of an ischemic stroke secondary to the epidural hematoma was not clear at first.

CONCLUSION

In progeric patients, the development of brain herniation after a traumatic brain injury can facilitate the occlusion of large cerebral arteries and result in a stroke. The treatment must be tailored with particularities related to the physiopathology of HGPS disease with influence on the anesthesiology and neurosurgical actions.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Authors declare that this work exempted from ethics committee authorization

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

Funding

This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors

CONTRIBUTIONS

-Angelo Raimundo da Silva Neto: Conceptualization, Data curation, Methodology, Supervision, Writing – original draft, Writing – review & editing

-Felipe Augusto Silva Aires: Investigation, Methodology

-Elisa Torquato Cardoso Lopes: Investigation, Methodology

-Eric Cymon do Vale Beserra: Conceptualization, Investigation, Methodology

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