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Case Report

Surgical management of congenital sternal cleft in a pediatric patient with PHACES syndrome

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ABSTRACT

PHACES syndrome, first described in 1978 and named in 1996, is a rare disorder involving multiple malformations, including abnormalities of the posterior fossa, cervicofacial segmental hemangiomas, cerebrovascular arterial anomalies, cardiac or large vessel defects, ocular abnormalities and sternal defects such as the sternal cleft. It predominantly affects females, with a ratio of 9:1 and is seen in 2–3% of infantile hemangioma cases. Some patients may also present with thyroid disorders. The sternal cleft, one of the anomalies associated with PHACES syndrome, is a rare congenital malformation caused by a failure in the fusion of the sternal midline during embryonic development. It can be classified as partial or complete, with the superior partial cleft being the most common form. This defect compromises the protection of mediastinal organs and can result in varying degrees of respiratory distress. Diagnosis is typically straightforward through inspection and palpation at birth.

Keywords: Congenital malformations, PHACES Syndrome, Surgical repair, Sternal cleft

INTRODUCTION

PHACES syndrome was first described in 1978 and named with that name in 1996, which is an acronym for posterior fossa (or brain) malformations, cervical-facial segmental hemangioma, cerebrovascular arterial malformations, cardiac or large vessel defects, eye defects and sternal cleft, some patients have thyroid disorders. Described by Rotter, et al, PHACE syndrome is observed in 2-3% of cases of childhood hemangiomas, it most often affects women in a ratio of 9:1.[1]

The sternal slit is located within the defects of PHACES syndrome, characterized by defect in the fusion in the sternal midline that constitutes a rare anomaly. The first reported surgical correction was performed in 1888 by M. Lannelongue.[2] This article is based on the case of a 10-months-old male infant who had a complete sternal slit and is performed in a third-level hospital by the plastic and reconstructive surgery and thoracic surgery service.

CASE REPORT

Male patient of 10 months, product of third deed, of term (39 SDG), APGAR of 9, weighed 3180 grams and 53 cm in size, obtained by maternal cesarean section with adequate prenatal control and supplementation with folic acid, without suspected injury before birth carrier of PHACES syndrome which was diagnosed by the genetics service of the University Hospital at 7 months, carrier of hemangioma in the lower lip, left nasal wing and right kygomatic region. At birth, an obvious defect in the

anterior chest is observed, characterized by a sternal slit that covers the upper 2/3 thirds, adequate amplexion and amplexation movements, without respiratory distress data, audible and normal vesicular murmur auscultation in both pulmonary fields, normo-dynamic precord, rhythmic cardiac noises without audible murmur, which does not condition ventilatory alteration (Figure 1).

Studies were conducted to rule out reported vascular malformations such as: Echocardiogram: Aortic tortuosity at the level of the aortic arch, normal skull CT, Angioresonance with probable aortic pseudocoartation and normal cerebral angioresonance. Scheduled for surgical intervention by the plastic and reconstructive surgery and thoracic surgery service performed on August 10, 2023.

It proceeds to perform incision in the middle sternal line, it is dissected by skin planes, subcutaneous cellular tissue finding lack of complete sternal fusion (Figure 2).

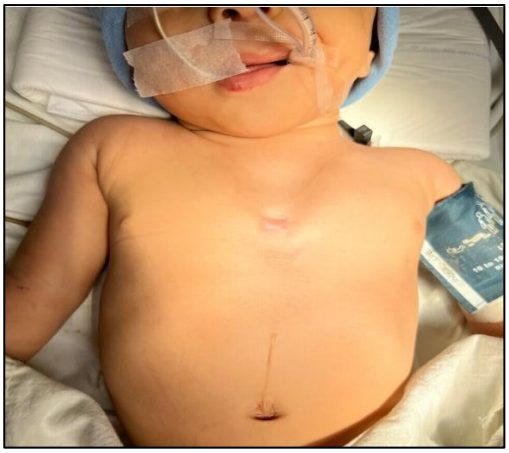

Figure 1: Male patient of 10 months, with obvious defect in the anterior chest.

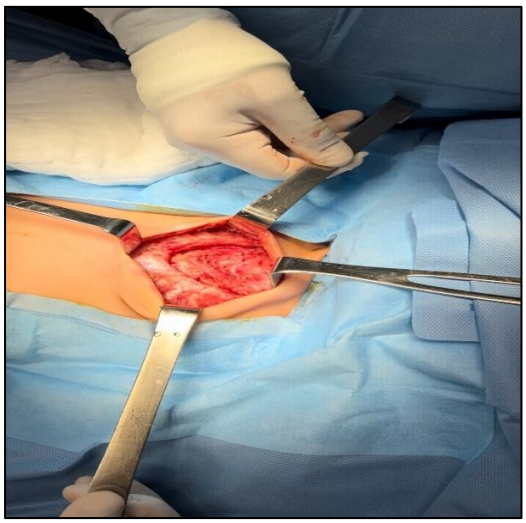

Figure 2: Obvious defect in the anterior chest.

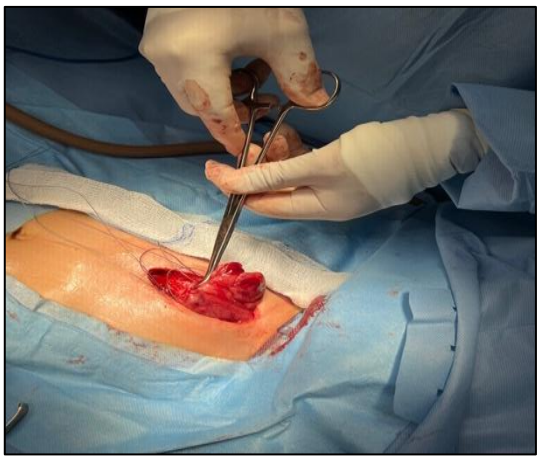

Figure 3: Timectomy.

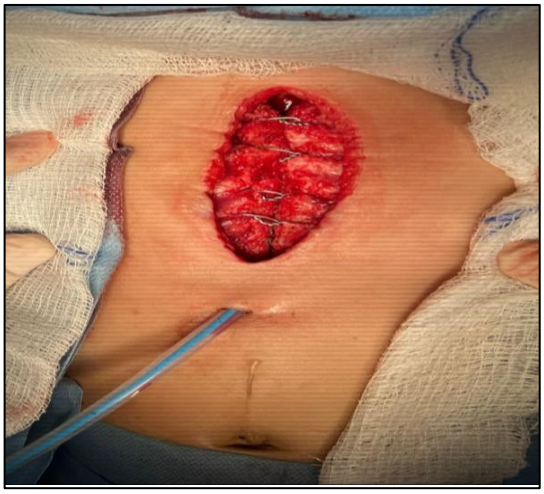

Figure 4: Sternography with wire points 2-0.

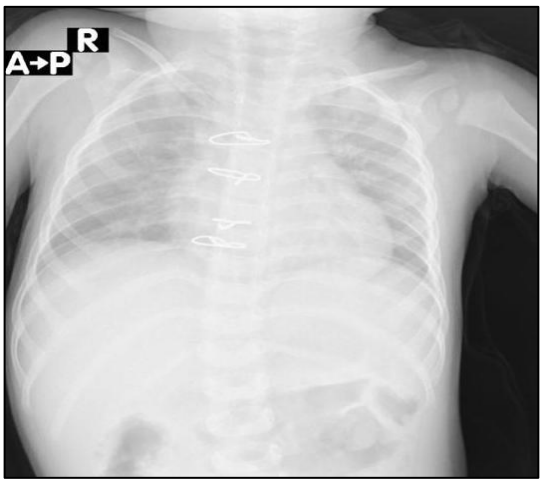

Figure 5: Control X-ray after 6 months of surgery.

It proceeds to dissect the anterior lamina of the sternum and it is dissected laterally pectoral major bilaterally to the sides exposing costal cartilages separating mediastinal pleura laterally to sternoclavicular joint, bilateral sternal osteotomy is performed, for matching of edges checking

adequate alignment is revived bilateral sternal lateral edge showing voluminous thymus so that resection of the same is performed (Figure 3).

We proceed to perform sternography with 2-0 wire points (Figure 4). It proceeds to perform closure by planes, muscular with vicryl 2-0, subcutaneous with vicryl 2-0 and subdermal skin with monocryl 3-0 and surgical act is terminated without accidents or incidents.

Patient goes to the intubated pediatric intensive care unit, with a mediastial probe which reports expenditure on the first day of 62 cc, on the second day an expenditure of 37 cc is reported, leaving it to seal on the third day and being removed on the fourth postoperative day without complications.

The patient was dis-carried from our unit on the fifth postoperative day, without complications, currently continued in follow-up by outpatient consultation without presenting ventilatory repercussions by surgical procedure (Figure 5).

DISCUSSION

The sternal slit is a rare and very low frequency congenital malformation of the chest wall, which may or may not be associated with genetic syndromes such as PHACES, which can be classified according to Corina Zamfir et al, into partial slits and total slits.[3] The partial forms can be upper, middle or lower, the most frequent form is the upper partial slit, the ossification of the sternum begins in the fifth month of embryonic life. The malformation causes a failure in the protection of the midstanic organs, which can occasionally cause respiratory distress of varying degrees.[4]

The diagnosis can be made easily, at birth through inspection and palpation, the peculiar mechanics of the chest wall and unusual movements are suggestive. Sometimes, it is necessary to perform imaging studies, such as X-rays or computed tomography, to evaluate the extent of the slit and to rule out other associated anomalies.[5] These studies also make it possible to differentiate congenital sternal slit from other similar conditions, such as pectus excavatum or costal malformations.

Most patients with congenital sternal slit do not require treatment, as the condition does not usually affect the child's overall health.[6] In cases where the slit is deep or is associated with other anatomical defects, surgical options may be considered to correct the aesthetic or functional deformity Surgical treatment is generally limited to severe cases or those with significant comorbidities.[7] The indications for surgical management in patients who present this malformation are to remove the bone protection of the mediastinal structures, prevent abnormal movement (thoracic dissociation) at the time of breathing, eliminate the visible deformity and allow the normal development of the rib cage.[8]

For which several techniques are described that can be classified into two groups: Those that act by modifying the costal and sternal anatomy and Those that limit themselves to hiding the defect through fillings, the most used are the autografts of cartilage, iliac crest, ribs, parietal skull and tibial perios with good result.[9] The need for partial or total removal of the thymus due to space limitations has been demonstrated in pediatric patients described The prognosis for children with congenital sternal clenss is generally excellent. Most cases are asymptomatic and do not affect the patient's development or quality of life. While patients with more severe forms of the condition may require surgery, recovery is usually satisfactory and complications are rare.[10]

CONCLUSION

Congenital sternal slit is a rare and generally benign anomaly that affects a small number of pediatric patients. Although most cases do not present symptoms or complications, it is essential to make an adequate diagnosis to differentiate it from other conditions and to identify possible comorbidities. The treatment is usually conservative and the prognosis is favorable for most patients, however the repair at an early age as in the patient described, is simple, with good functional result and good aesthetic result in addition to achieving adequate ventilation, on the contrary when the repair of the defect is deferred to older ages, more invasive procedures are required.

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