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Rare disease

Congenital medium sternal cleft with partial ectopia cordis repair

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Abstract

Congenital sternal malformation is a rare anomaly often diagnosed as an asymptomatic condition at birth. The authors report a clinical case of a full-term female neonate with congenital sternal cleft and partial ectopia cordis. Successful surgical repair was accomplished at 6 days of age. When surgery is performed shortly after birth, the procedure is easier and better results are achieved.

BACKGROUND

Congenital sternal cleft is a rare chest malformation of unknown origin. Little is known about its incidence and pathogenesis, and no familiar, teratogenic or nutritional factors have been implicated. In literature reviews this malformation is reported as sporadic. Depending on the degree of separation, sternal clefts can be classified as complete or incomplete. They may result in a high mortality rate in the postnatal period if associated with cardiac anomalies. However, isolated sternal clefts have a favourable prognosis. It is known that surgical repair should preferably be performed in the neonatal period when the thorax is more compliant.

We report, herein, a case of a female neonate presenting with an incomplete isolated medium sternal cleft with partial ectopia cordis. The patient underwent a successful surgical repair. She remains well 2 months after surgery.

CASE PRESENTATION

A female neonate, 37 week's gestational age, with uneventful pregnancy was born from a eutocic uncomplicated delivery, to a 32-year-old GII, PII Caucasian woman. At delivery the birth weight was 3008 g, with Apgar scores of 9 and 10 at the first minute and fifth minute, respectively. Noted was a midline thoracic wall defect with a 2.5×3.5 cm diameter overlying thin hypopigmented bulging membrane, evident during expiration, and weeping. In inspiration a depression appeared in the same area; the anomaly was well tolerated without symptoms ([fig 1](#)). The rest of the physical examination and the investigation performed on this case, including karyotyping, chromosome banding studies, skeleton x ray and transfontanelar, cardiac and abdominal ultrasonography, were unremarkable.

At the age of 6 days, the patient underwent a primary surgery repair. The skin was incised in the midline and skin flaps with approximately 3 cm each were raised on both sides of the incision, with isolation of a small cleft zone in the middle of the sternum, without any anomaly of the superior and inferior sternum portions. A vertical sternum osteotomy was performed from the xiphoid appendix, passing the cleft zone until the manubrium. A partial ectopia cordis with a pericardium to the skin sinus tract was found and excised ([fig 2](#), arrow). The pericardium foramen was sutured with 4-0 absorbable suture. The borders of sternum were freshened to facilitate the approximation of the sternal bars and fixed in X with four Tycron sutures. A periosteum graft, excised from the sternum, was

implanted to correct the defect. A complete closure of the defect was done with subcutaneous and skin flaps closed, with drainage at the surface (fig 2). No evidence of cardiac compression was noted, and the patient remained haemodynamically stable throughout surgery and in the postoperative period. The patient is doing well after 2 months from the procedure.

DISCUSSION

Congenital sternal malformation is rare and sporadic, and probably with multifactorial aetiology without hereditary background. It is generally found at birth asymptotically.¹ In some series there is a marked female predominance although most of the series are small.^{2,3}

Congenital sternal cleft is classified as complete or incomplete,^{4,5} with complete form being the rarer.⁶ Incomplete clefts are subdivided into superior type and inferior ones. Inferior incomplete cleft may occur as an isolated entity, although it is usually associated with other development defects of the anterior chest wall such as ectopia cordis, or, in Cantrell's pentalogy, with a combination of defects involving the abdominal wall, sternum, diaphragm, pericardium and heart.³ It is undisputed concept that sternal cleft should be corrected in the neonatal period. At this stage, the sternal bars can be easily approximated by simple suture, due to the maximal flexibility of the cartilaginous thorax with a minimal risk of underlying cardiovascular structures.^{3,4}

In conclusion our case is especially unusual because it is a small medium sternal cleft associated with partial ectopia cordis, and does not fit the usual findings and/or classical sternal cleft classification.

LEARNING POINTS

- Congenital sternal malformation can be associated with a high mortality rate in the postnatal period if associated with cardiac anomalies.
- It is important to recognise this condition because of the increased risk of mediastinic structures trauma.
- For a better outcome, the sternal cleft should be corrected in the neonatal period.

Footnotes

Competing interests: none.

Patient consent: Patient/guardian consent was obtained for publication.

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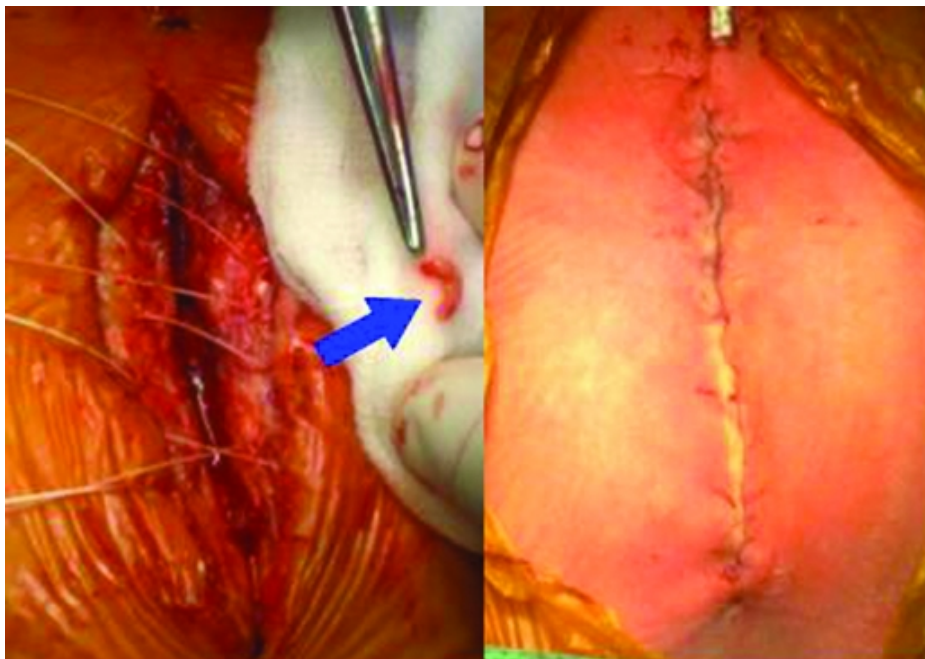
Figures and Tables

Figure 1



Bulging and depressive membranous defect during expiration and inspiration with a hypopigmented cutaneous rim.

Figure 2



Sternum osteotomy, excision of pericardium to the skin sinus tract and postoperative photograph of the patient.