CASE REPORT

Closing the cleft over a throbbing heart: neonatal sternal cleft

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SUMMARY

Sternal cleft is a rare anomaly comprising 0.5% of chest wall malformations. We present a case of a neonate with a 'V'-shaped upper partial sternal cleft at birth. A hyperpigmented cutaneous nevi was present over the cleft. Primary approximation and closure of the defect was performed at 1 week of life. We discuss the presentation and management, and review the literature.

BACKGROUND

Sternal cleft is a rare anomaly occurring in 1 in 50 000–100 000 births. We present a case of a neonate with partial sternal cleft for its rarity, and a newer association, a cutaneous hamartoma is also reported. We propose a simple algorithm for management.

During the sixth week of embryonic life, the sternum arises from a pair of longitudinal concentrations of mesenchymal tissue, called 'sternal bars', located on either side of the anterior chest wall. These sternal bars migrate and fuse in a craniocaudal direction to form the sternal plate by the 10th week.¹ Meanwhile, mesenchymal condensations become cartilaginous in the seventh week of intra-uterine life. After maturation into cartilage and fusion, the sternum undergoes several transverse divisions into a series of six cartilaginous segments called 'sternebrae'. There is one segment for the manubrium, four for the body and one for the xiphoid process.¹ Any defect in the fusion process produces sternal cleft.

Cleft sternum can be classified as complete cleft or partial cleft. Partial clefts can be superior or inferior. A superior partial cleft is either a U-shaped defect with the cleft ending at the level of the fourth costal cartilage, or a 'V'-shaped defect if the sternum is cleft to the xiphoid process. Inferior clefts are rare and are usually associated with other abnormalities of midline fusion.

CASE PRESENTATION

A term male baby was delivered by caesarean section at our hospital. The indication was premature rupture of the membrane with fetal distress. The mother was a 24-year-old primi, and her antenatal scan at 6 months at a primary health centre was reported to be normal. She took iron and folic acid tablets during pregnancy and her antenatal period was uneventful.

On examination, the baby cried at birth and his APGAR was normal. The birth weight was 2.6 kg. A 'V'-shaped concave defect in the upper anterior chest wall extending up to the xiphisternum and

covered by skin was noted. During crying, paradoxical skin movement with herniation of cardiac structures was noted over the defect. A hyperpigmented naevus was noted on the skin over the cleft in the midline (figure 1). The baby had tachypnoea with clear lung fields at birth which gradually settled over 36 h of life.

INVESTIGATIONS

Basic investigations like haemoglobin, blood count and C reactive protein were normal. Chest X-ray anteroposterior view and lateral view showed no ossification centre over the sternal area. CT of the chest with three-dimensional (3D) reconstruction revealed a central 'V'-shaped defect with sternal segments on either side and widening of both clavicular ends (figure 2). The central fusion of the lower ends of sternal bars was not present. The pericardium was intact and lung fields were normal. The radiologist opined complete cleft. But upper partial cleft and xiphisternal cartilage was confirmed intraoperatively. ECG and echo performed to rule out cardiac anomaly showed a patent foramen ovale on day 1 and closure of it after 1 week.

DIFFERENTIAL DIAGNOSIS

Sternal cleft as a part of syndromes such as pentalogy of cantrel and thoracoabdominoschisis was considered. Abdominal ultrasonogram, echo and CT of the brain ruled out diaphragmatic defects, intracardiac defects and posterior fossa abnormalities, respectively.

Figure 1 'V'-shaped cleft, bulging heart (arrow), hyperpigmented nevi over skin (arrow head).

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Figure 2 CT of the chest. (A) Three-dimensional reconstruction showing increased interclavicular distance and bilateral sternal segments (short arrows); (B) Axial section showing defective sternum with intact pericardium (long arrow) and normal lung fields.

TREATMENT

Intravenous fluids and antibiotics were given for the first 3 days. Spoon feeds with expressed breast milk were initiated after settlement of respiratory distress and the neonate was taken for surgery on the seventh day of life.

Procedure: Under general anaesthesia, an upper midline incision was made over the defect. Primary approximation of sternal bars was performed with 3–0 ethibond sutures (figure 3) and the defect closed (figure 4). Postoperatively, the baby was placed in the neonatal intensive care unit; feedings were started on day 2 and the baby was discharged on the 10th postoperative day.

OUTCOME AND FOLLOW-UP

At 3-month follow-up, the baby was thriving with no major complications.

DISCUSSION

So far, slightly more than 100 cases of sternal cleft have been described in the literature. Sternal cleft is often asymptomatic (74%) and associated with other defects (72%). A higher incidence of partial superior type sternal cleft and female predominance was described.²

The implicated aetiologies include riboflavin deficiency and *HOXB4* gene disruption.³ Superior clefts are associated with facial haemangiomas, midline supraumbilical raphe and PHACES syndrome. Inferior clefts are associated with more serious problems like cardiac defects and pentalogy of cantrell

(skin, sternum, diaphragm, pericardium and intracardiac defects).

Our case had a midline hyperpigmented, slightly elevated patch present over the lower part of the cleft. We clinically suspected it to be an accessory areola; but the excised skin examined by the pathologist contained a proliferation of connective tissue and a few glandular elements, suggestive of a hamartoma. Similarly, in a case series,² three cases were associated with precordial skin tags, which on examination were found to be hamartomas. Prajapati and Chaturvedi⁴ reported a hyperpigmented cutaneous nevi over the sternal cleft in a neonate. Mogilner *et al*⁵ reported that the presence of cutaneous nevi was associated with

Figure 3 Both sternal bars (arrows) approximated to the midline with ethibond sutures.

Figure 4 Skin closure.

a skin to pericardial sinus tract. In the present case the pericardium was intact and no sinus tract was found.

Kaplan⁶ proposed that the rupture of chorion or yolk sac during cardiac descent causes a disturbance in the mesodermal structures, leading to sternal cleft. Involvement of cutaneous tissue in this mesodermal disturbance was proposed to explain the association of haemangiomas in sternal cleft. We believe similar mechanisms explain the association of cutaneous hamartomas.

Prenatal diagnosis

Fetal sternal development is complete in 10 weeks. Thus, the sternum can be studied during 12th week examination. However, this is difficult because of the frequent flexion of the fetal head on the thorax and the thinness of the sternum compared with the thickness of the sonographic section. The second trimester and early third trimester (between 18 and 26 weeks) is the best period for sternal study, because of the absence or small number and size of ossification centres.

Figure 5 Management algorithm (USG, ultrasound sonography).

Postnatal workup

At birth, the sternum is normally seen on lateral view X-ray as four or five distinct segments or ossification centres. CT is the modality of choice to evaluate anatomic detail of the sternum.⁷ Reformatted coronal images better demonstrate the lucent defects corresponding to clefts. 3D reconstruction shows the shape, extent of defect and sternal bars or segments on both sides.

MRI (T1-weighted spin-echo pulse sequence) can confirm CT findings and provide additional information about the bone marrow and soft tissues adjacent to the sternum.

Management

Primary closure is the preferred modality in infants under 3 months because of pliability of the chest wall, and compression of structures is minimal.⁸ After 3 months, sliding or rotating chondrotomies, autologous grafts such as costal cartilages, parietal skull, tibial periosteum, ribs and the use of prosthetic materials such as marlex, acrylic, silicone elastomer, teflon and stainless steel mesh were described. We propose a simple algorithm for the management of sternal cleft (figure 5).

In a review of institutional series, three intraoperative complications (3/70, 4%) due to pericardial or pleural tears during

sternal dissection were reported. Postoperative complications were 12 (17%), mostly represented by retrosternal seromas and pneumothorax. Death was reported in cases of three patients affected with cardiac defects.² In our case a stitch abscess was noted on follow-up, which resolved with conservative management. No recurrences of sternal cleft after repair have been described in the literature. Follow-up is necessary as these patients can develop chest wall deformities such as pectus excavatum.

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Contributors ARJ was involved in case management, designed the manuscript and is the guarantor; RP revised the manuscript and followed up the case; MK and MG revised the manuscript and gave the approval.

Competing interests None.

Patient consent Obtained.

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Learning points

- Upper partial sternal cleft is associated with cutaneous hyperpigmented nevi in the chest.
- CT of the chest with three-dimensional reconstruction gives anatomic details, while echo, CT of the brain and ultrasound of the abdomen help to rule out associated anomalies.
- Primary approximation is a surgical opportunity in the neonatal period and carries good prognosis.

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