Complete sternal cleft with tetralogy of Fallot

Debasis Das[1], Nilanjan Dutta[1], Pratik Pandey[1], Adhish Basu[2], Manish Kumar Sharma[3], Rangan Koley[3], Amitabha Chattopadhyay[4]

[1]Department of Cardiac Surgery, Narayana Superspeciality Hospital, Howrah, West Bengal, India, [2]Department of Plastic Surgery, Narayana Superspeciality Hospital, Howrah, West Bengal, India, [3]Department of Cardiac Anaesthesia, Narayana Superspeciality Hospital, Howrah, West Bengal, India, [4]Department of Pediatric Cardiology, Narayana Superspeciality Hospital, Howrah, West Bengal, India

ABSTRACT

Complete sternal cleft is a very rare congenital midline defect of the sternum. It is not uncommonly associated with intracardiac defects. We report a case of a 2-year-old child with complete sternal cleft and tetralogy of Fallot who presented with cyanotic spells. The child underwent total correction, followed by chest wall reconstruction on the next day.

Keywords: Cleft Sternum, surgical technique, tetralogy of Fallot

INTRODUCTION

Cleft sternum (CS) is a rare thoracic malformation. Isolated sternal defects have fairly good prognosis. However, complete CS can present with any form of intracardiac defects including life-threatening disorders such as ectopia cordis or Cantrell's pentalogy. The presence of any such association confers negative outcome on surgical corrections. An association of complete CS with tetralogy of Fallot (TOF) is extremely rare, and not many cases have been reported.

CASE REPORT

A 2-year-old girl presented to our emergency room in cyanotic spells. She was a known case of TOF and midline thoracic defect for 3 months of age. The child had to be intubated in the emergency room considering severe desaturation not responding to usual measures. There was a visible midline cardiac pulsation on the chest wall with an overlying central raphe [Figure 1] without any palpable sternum.

Echocardiography showed large malaligned subaortic ventricular septal defect (VSD), severe infundibular and valvular pulmonary stenosis, confluent- and adequate-sized branch pulmonary arteries, right aortic arch, and bilateral superior vena cavae. A computed tomography scan of the thorax was done to rule out any other associated malformations. The scan confirmed a complete CS [Figure 2]. The child underwent cardiac catheterization study to confirm intracardiac anatomy and to rule out aortopulmonary collaterals.

A combined staged surgical procedure in the form of intracardiac repair followed by chest wall reconstruction was planned. Meanwhile, the child was kept ventilated.

The patient underwent Dacron patch closure of VSD, infundibular resection, pulmonary valvectomy, and transannular autologous pericardial patch augmentation of the right ventricular outflow tract. Branch pulmonary arteries were adequate and confluent. A conscious decision was taken by the team members to postpone the sternal reconstruction by 24 h in view of a major intracardiac procedure having been performed. Sternal reconstruction was done subsequent morning. We performed bilateral sliding chondrotomies of the first four ribs and local

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Address for correspondence: Dr. Debasis Das, Department of Cardiac Surgery, Narayana Superspeciality Hospital, 120/1 Andul Road, Howrah - 711 103, West Bengal, India. E-mail: dasdeba@yahoo.com

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advancement of the pectoralis major muscle to close the defect primarily with No. 2 stainless steel wires [Figure 3a and b]. The child was electively ventilated and extubated on the 4th postoperative day. Her hospital stay was prolonged (20 days) due to the need for intense chest physiotherapy and oxygen dependency. The patient was discharged with stable sternum without any deformity. She is doing well on the last follow-up as well, 1 year after the procedure.

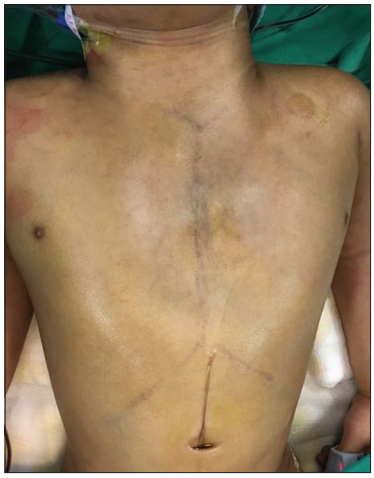

Figure 1: Preoperative image showing a supraumbilical median raphe

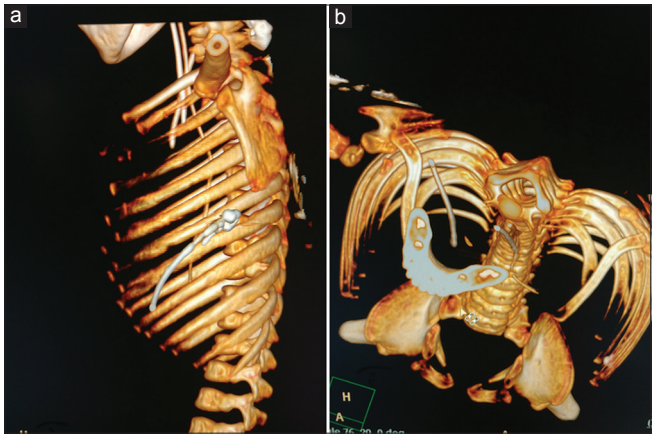

Figure 2: (a) Three-dimensional reconstructed lateral image computed tomography scan of the thorax. (b) Three-dimensional reconstructed anterior–posterior image computed tomography scan of the thorax

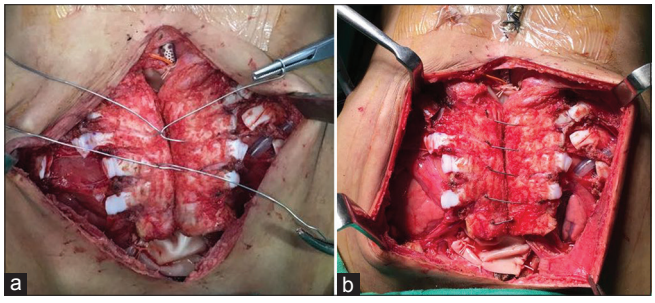

Figure 3: (a) Postchondrotomy sternal plate mobilization and wiring. (b) Completed bony cover over the heart

DISCUSSION

CS accounts for only 0.15% of all chest wall malformations.[1] It can be complete and incomplete type. Complete CS is very rare and involves splitting of the sternum from the manubrium to the xiphoid. This variety is also strongly associated with complex intracardiac defects such as TOF, Cantrell's pentalogy, ectopia cordis, and many others. Noncardiac anomalies may be cervicofacial hemangiomas, diastasis recti, and cleft mandible.[1] Incomplete CS presents with chondral bridges either in the upper part (inferior-type incomplete CS) or in the lower part (superior-type incomplete CS) of the sternum. Incomplete CS is very often associated with vascular dysplasias. A supraumbilical raphe is present in 30% of cases of CS.

The etiology of CS is speculative. There are two theories involved:

- Failure of fusion of lateral sternal bands with midline mesodermal structures between the 6th and 9th gestational weeks[2]
- Early amnion rupture may result in multiple defects by pressure necrosis, incomplete embryogenesis, and tearing and tethering by amnion bands.[3]

Surgery is advocated to protect the underlying heart from external trauma to allay parental anxiety and to prevent recurrent respiratory tract infections.[4] It is well accepted that isolated CS is best repaired primarily in the neonatal period because extreme compliance and pliability of bony thorax permit direct sternal closure. However, the presence of complex cardiac defects postpones the sternal repair until the age and weight of elective operation of the cardiac malformation are reached.[5]

Numerous methods of surgical correction are available. The factors which decide an appropriate correction technique are the age of the patient and degree of the cleft. The first bony repair was attempted by Burton in 1947 by placing the cartilage graft over the defect.[4] Maier and Bortone achieved primary closure without the use of any graft in a 6-week-old child.[6] Older age at presentation necessitates adoption of other repair techniques. Sabiston described multiple bilateral sliding chondrotomies to approximate the defect.[4] Meissner reported "door wing plasty" which included the division of coastal cartilages laterally and turning them medially to bridge the defect.[4] Acastello *et al.*[1] surgically dislocated the sternoclavicular junction for sternal approximation without tension. When direct closure is not possible without rise in intrathoracic pressure, autografts such as cartilages, ribs, and tibia can be used.[1,4,6] Prosthetic materials such as Teflon, mesh,

silicone, acrylic, and titanium plates can also be used.[1,6] However, these materials are associated with infections, extrusions, and failure to remodel themselves to patient's growth.[7] Direct approximation of the sternal halves should, therefore, be attempted whenever possible. Bové *et al.*[8] described a 2-month-old infant with upper CS (incomplete) and TOF.

Our patient was a 2-year-old girl. We corrected intracardiac defects followed by sternal repair on the next day. The chest wall reconstruction was intentionally deferred in an attempt to make the procedure safer. Being a late presenter, we felt concerned about acute tamponade effect after primary closure of the sternal defect. Sternal closure was achieved by bilateral sliding chondrotomies (Sabiston's technique) of four ribs followed by local advancement of the pectoralis major muscle flap. We have been able to perform primary repair in a late presenter without any use of autologous or prosthetic materials. Mobilization of the pectoralis major and double-breasting provided strength to the repair and reduced the lateral tension.

In conclusion, this case is extremely rare as regards to late presentation and association of complete sternal cleft with TOF. Although primary repair of complete CS was successful in this case of late presentation, it may not be possible always.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Acastello E, Majluf R, Garrido P, Barbosa LM, Peredo A. Sternal cleft: A surgical opportunity. J Pediatr Surg 2003;38:178-83.
2. Hersh JH, Waterfill D, Rutledge J, Harrod MJ, O'Sheal SF, Verdi G, *et al.* Sternal malformation/vascular dysplasia association. Am J Med Genet 1985;21:177-86, 201-2.
3. Van Allen MI, Myhre S. Ectopia cordis thoracalis with craniofacial defects resulting from early amnion rupture. Teratology 1985;32:19-24.
4. Shamberger RC, Welch KJ. Sternal defects. Pediatr Surg Int 1990;5:156-64.
5. Salley RK, Stewart S. Superior sternal cleft: Repair in the Newborn. Ann Thorac Surg 1985;39:582-3.
6. Dòmini M, Cupaioli M, Rossi F, Fakhro A, Aquino A, Chiesa PL. Bifid sternum: Neonatal surgical treatment. Ann Thorac Surg 2000;69:267-9.
7. Yavuzer S, Kara M. Primary repair of a sternal cleft in an infant with autogenous tissues. Interact Cardiovasc Thorac Surg 2003;2:541-3.
8. Bové T, Goldstein JP, Viart P, Deuvaert FE. Combined repair of upper sternal cleft and tetralogy of fallot in an infant. Ann Thorac Surg 1997;64:561-2.