

Case Report:

“Complete sternal cleft with patent foramen ovale and meckel’s diverticulum”

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Abstract:

We report herein a rare case of complete congenital sternal cleft (absent sternum) in association with Meckel’s diverticulitis and patent foramen ovale. Sternal defects are classified as partial (superior and inferior) and complete. Complete clefts are rarer. Till now only 25 cases has been reported in literature^{1,2}. In neonates with absent sternum, the sternal bars can be easily approximated by simple suture, due to the flexibility of the cartilaginous thorax. There is also little danger of cardiac compression when the repair is performed early in life. Clinical outcome may be unfavourable when an associated anomaly, particularly, an intra cardiac anomaly coexists with the defect. If reconstruction is delayed, the increased rigidity of the chest wall and the physiologic accommodation of the thoracic organs to the circumference of the chest render simple approximation impossible, without serious compromise of the heart and lungs.

Key words: Congenital sternal cleft, patent foramen ovale, Meckel’s diverticulitis.

Introduction

The first known case of sternal cleft was described by Torres in 1740³. Since then, few publications have appeared concerning sternal cleft, which have led to introduce a classification and some therapeutical procedures⁴. Sternal defects are rare chest wall deformities that present with a broad spectrum of clinical severity from the potentially lethal ectopia cordis to the benign sternal cleft. The incidence of sternal defect is unknown; however, Acastello and colleagues stated that it represents 0.15% of all patients with a chest wall malformation in their patient population (5182 patients over a 25-year period)⁵.

In most infants, cleft sternum usually does not cause any detectable symptoms. Clavicles and nipples may be widely apart. On occasion, respiratory symptoms may result from the paradoxical motion of the sternal defect. The primary indication for repair is to protect the heart. Repair is best performed in infants because the

chest wall is most pliable. As the age advances, the increased rigidity of the chest wall and the physiologic accommodation of the thoracic organs to the circumference of the chest render simple approximation impossible⁶.

Case report

A 30 years old lady was admitted in Mahatma Gandhi Medical College and hospital, Sitapura, Jaipur in September 2012, with chief complaints of acute abdomen and was diagnosed with acute Meckel’s diverticulitis with mesenteric lymphadenopathy on Contrast enhanced CT scan of abdomen. Her past medical history was unremarkable. In the routine check-up patient is diagnosed to have sternal defect of about 2.5 cm through which cardiac pulsations could easily be seen. Patient had widely apart nipples and clavicles. A midline thoracic depression was evident during inspiration (figure 1). During expiration or coughing, a bulge appeared in the same area (figure 2). Patient didn’t have any

respiratory or cardiac problem. There was no abnormal lung and heart sounds on auscultation.

Chest radiograph AP and lateral views were done before surgery (figure 3). Helical CT chest was also performed on 128 slice CT with 3D reconstruction showing sternal cleft of 2.3 cm (figure 4). The diagnostic images showed the typically widely separated clavicles and absence of manubrium and xiphoid process. The electrocardiogram and an echocardiogram of the patient were also done. The electrocardiogram was normal but the Doppler echocardiography showed the presence of patent foramen ovale (figure 5).

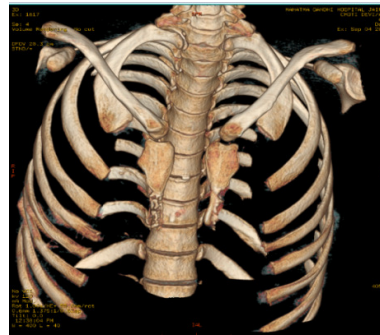
Patient was operated for Meckel's diverticulum.



(Figure. 3 patient's chest radiograph lateral view showing absent sternum and gas under diaphragm)



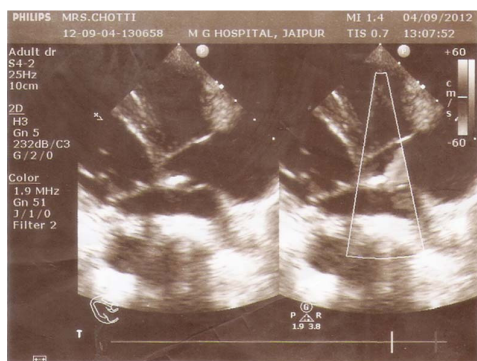
(Figure. 1 patient's chest in inspiratory phase showing depression in midline)



(Fig.4 Helical CT chest with axial scans and 3D reconstruction showing typically widely separated clavicles and absence of manubrium and xiphoid process)



(Figure. 2 patient's chest in expiratory phase showing bulging in midline)



(Figure. 5 Doppler echocardiography showing the presence of patent foramen ovale)

Discussion

Sternal defects are divided into four categories based on tissue coverage of the heart: (i) sternal cleft or bifid sternum, (ii) thoracic ectopia cordis, (iii) cervical ectopia cordis, and (iv) thoracoabdominal ectopia cordis⁷. Cleft sternum is the least severe of the 4 anomalies. In sternal clefting, the thoracic viscera are covered only by soft tissue, and they may be bulging and pulsatile, but the heart and lungs are normally positioned anatomically and do not require repair when the sternal defect is closed. Sternal defects are classified as partial (superior and inferior) and complete. A partial split is more common than complete split. A Superior defect may be U-shaped, with a defect ending at the level of the fourth costal cartilage, or V-shaped if it extends to the xiphoid process. Superior and complete sternal clefts are generally isolated abnormalities. Inferior or distal sternal clefts are very rare and are usually seen in association with other characteristic abnormalities of midline fusion as in *Cantrell Pentology*. Several associations are seen with cleft sternum. A band like scar (raphe) often extends from the sternum to the umbilicus or superiorly to the neck. Gorlin et al. reviewed the association of sternal defects with supraumbilical raphe and found 42 examples in the literature from 1842 to 1992⁸. A second group of

31 patients reported during 1880 to 1994 had a marked female predominance (29F:2M) and had facial hemangiomas appearing within the first week of life⁹. Within this second group is an apparent syndrome, recently termed *PHACE syndrome*, which includes the association of posterior fossa brain abnormalities (typically Dandy-Walker Malformation), hemangiomas, arterial malformations, coarctation of aorta, cardiac malformations and eye malformations. This syndromes also shows female predominance^{10,11}.

In ectopia cordis the heart and thoracic viscera are ectopic, either lying on the outer surface or displaced superiorly to the neck or inferiorly to the abdomen. The thoracoabdominal type of ectopia cordis is usually found in association with inferior sternal cleft, diaphragmatic, pericardial, cardiac and anterior wall defects and is termed as *Cantrell Pentology*¹².

Sternal clefts are rare congenital malformations that result from defective embryologic fusion of paired mesodermal bands in the ventral midline by the ninth weeks of gestation¹³. Cells from the lateral plate mesoderm migrate ventrally in the sixth intrauterine week to form two parallel mesenchymal bands or bars one on each side. These bands fuse craniocaudally in the midline by the ninth intrauterine week to become the body of the sternum and part of the manubrium. Three other small mesenchymal primordia, which arise between the developing clavicles, complete the cranial part of the manubrium. The sternum chondrifies and then ossifies from multiple ossification centres which appear in sequence from cranial to caudal, beginning at the sixth intrauterine month. At birth the sternum is mainly cartilage^{14, 15, 16}.

In our patient two congenital anomalies were present apart from sternal cleft the patent foramen ovale and Meckel's diverticulum. The association of patent foramen ovale with sternal cleft has been

shown earlier¹⁷, but to the best of our knowledge none of the reports have shown the association of Meckel's diverticulum with sternal cleft. It could be a matter of chance that the two deformities are found together but their association cannot be ruled out, as Meckel's diverticulum could only be looked

for if there is a picture of diverticulitis. Secondly, due to paucity of the cases of sternal cleft the true association is difficult to be established. But this study could pave the way towards establishing such association.

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