An Unusual Entity: Asymptomatic Isolated Superior Sternal Cleft: Case Report

Nadir Bir Antite: Asemptomatik İzole Superiyor Sternal Kleft

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ABSTRACT Congenital anomalies of the sternum include a broad spectrum of deformities that are classified into four groups as: cervical ectopia cordis, thoracic ectopia cordis, thoracoabdominal ectopia cordis, and cleft sternum. The sternal cleft is a rare congenital anomaly caused by fusion failure of the sternum. Two types of sternal cleft have been identified, as complete (the rarest form) and incomplete (the upper cleft sternum or bifid sternum). Isolated sternal clefts present a good prognosis due to the absence of cardiac anomalies and have a better chance for primary repair of the defect. Surgical correction is indicated to protect the heart and great vessels from injury, to improve respiratory dynamics and to remove cosmetic concerns. Here, we present a clinically asymptomatic newborn with isolated incomplete V-shaped sternal cleft and a skin defect covered with a membrane. No other abnormalities or dysmorphic features were detected on the physical examination. Abdominal sonography and cranial sonography, which were obtained in order to screen for the midline anomalies, were normal. Echocardiography revealed patent ductus arteriosus and patent foramen ovale. The magnetic resonance imaging demonstrated bilateral paired sternal cartilaginous centers on either side of the wide gap in the anterior chest wall. This was compatible with the appearance of a superior congenital sternal cleft. Primary surgical repair was performed at the eighth month of age.

Key Words: Thoracic wall; abnormalities; sternum; congenital

ÖZET Sternumun konjenital anomalileri geniş bir spektrumu kapsamaktadır ve dört grupta sınıflanmaktadır: servikal ektopia kordis, torasik ektopia kordis, torako-abdominal ektopia kordis ve sternal kleft. Sternal kleft sternumun birleşmesindeki yetmezlik sonucu gelişen nadir görülen konjenital bir malformasyondur. Komplet (en nadir tipi) ve inkomplet (superiyor veya inferiyor) sternal kleft olarak iki tipi tanımlanmıştır. İzole sternal kleft kalp anomalilerinin olmaması nedeni ile iyi prognoza ve primer onarım şansına sahiptir. Cerrahi onarım, kalp ve büyük damarları yaralanmadan korumak, solunum dinamiklerini iyileştirmek, kozmetik kaygıları ortadan kaldırmak amaçları ile yapılmaktadır. Bu yazıda klinik olarak asemptomatik, izole, V şeklinde superiyor sternal kleft ve üzeri membran kaplı cilt defekti olan bir yenidoğan vakası sunulmuştur. Fizik incelemesinde sternal kleft dışında herhangi bir anomali veya dismorfik bulgu saptanmayan bebeğin orta hat anomalilerini taramaya yönelik yapılan abdominal ve kranial ultrasonografisi normaldi. Ekokardiyografide patent duktus arteriozus ve patent foramen ovale saptandı. Manyetik rezonans görüntülemede torakal sternal kleft ile uyumlu açıklık tespit edildi. Primer onarım vaka sekiz aylıkken yapıldı.

Anahtar Kelimeler: Göğüs kafesi duvarı; anormallikler; sternum; konjenital

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Congenital anomalies of the sternum include a broad spectrum of deformities that are difficult to classify. In 1990, Shamberger and Welch classified them into four groups as: cervical ectopia cordis, thoracic ectopia cordis, thoracoabdominal ectopia cordis, and cleft sternum.[1]

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The sternal cleft is a rare, generally asymptomatic congenital anomaly caused by fusion failure of the sternum. A familial basis of sternal cleft has not yet been demonstrated, and probably occurs due to a multifactorial etiology.[2] Two types of sternal cleft have been identified, as complete and incomplete. The incomplete form is usually isolated, with orthotopic normal heart and normal skin coverage.[1] If the cleft reaches the xiphoid process, it is named V-shaped, and when a bony bridge joins the two edges, ending at the third or fourth costal cartilage, it is termed broad and U-shaped. In those rare symptomatic cases, cyanosis, dyspnea and pulmonary infections are the main clinical features.[3] Isolated sternal clefts present a good prognosis due to the absence of cardiac anomalies and they have a better chance for primary repair of the defect. Surgical correction is indicated to protect the heart and great vessels from injury, to improve respiratory dynamics and to address cosmetic concerns.[1,4]

Herein, we present a clinically asymptomatic newborn with isolated incomplete V-shaped sternal cleft and a skin defect covered with a membrane.

CASE REPORT

In June 2010, a one-day-old female newborn was referred to our hospital with a congenital defect of the anterior chest wall. The defect was not diagnosed in the antenatal period. The full-term infant girl was born to nonconsanguineous parents following an uncomplicated pregnancy by cesarian section. Her birth weight was 2750 g. She was found at birth to have an obvious deformity of the chest wall with widely spaced rib ends anteriorly and a 1 x 1.5 cm skin defect covered with a membrane, through which cardiac pulsations could be easily appreciated (Figure 1). No other abnormalities or dysmorphic features were detected on the physical examination. On admission, the patient was quiet with an oxygen saturation of 95% in room air. She had a respiratory rate of 45 beats/min, heart rate of 138 beats/min, and blood pressure of 55/30 mmHg. Laboratory investigations were within normal limits. Abdominal sonography and cranial sonography, which were obtained in order to screen for the midline anomalies, were normal. Echocardiography revealed patent ductus arteriosus and patent foramen ovale. The magnetic resonance imaging demonstrated bilateral paired sternal cartilaginous centers on either side of the wide gap in the anterior chest wall (Figure 2). The xiphoid process was positioned at the midline and covered by normal-appearing cutaneous tissues. This was compatible with the appearance of a superior congenital sternal cleft. During the one-week follow-up, the infant experienced no clinical problem. The skin defect was small, and there were no associated anomalies, so we preferred to follow the patient closely for spontaneous epithelization of the skin defect. Primary surgical repair was performed at the eighth month of age due to failure of spontaneous epithelization (Figure 3).

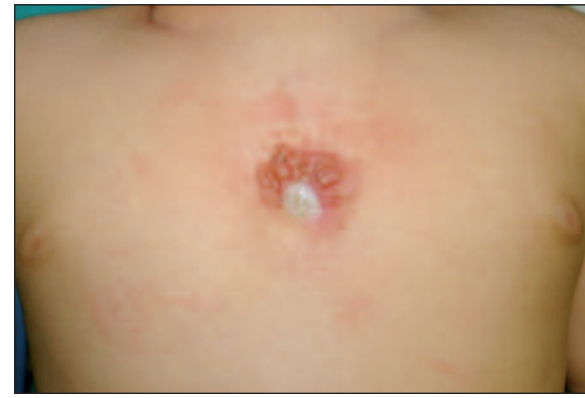

FIGURE 1: The photograph of the infant's anterior chest wall shows incomplete sternal cleft with widely spaced rib ends anteriorly and a 1x1.5 cm skin defect covered with a membrane through which cardiac pulsations could be easily appreciated.
(See for colored form http://pediatri.turkiyeklinikleri.com/)

DISCUSSION

The sternum originates from mesodermal cells in embryonic life. Mesodermal cells on either side of the anterior chest wall move toward the midline and fuse in the midline by the tenth week of gestation. The etiology of sternal cleft is still unknown. Most cases are sporadic, although a probable autosomal recessive inheritance was found in one family.[5] There were no relatives with chest wall deformities or cardiac anomalies in the family of our patient. Although most of the re-

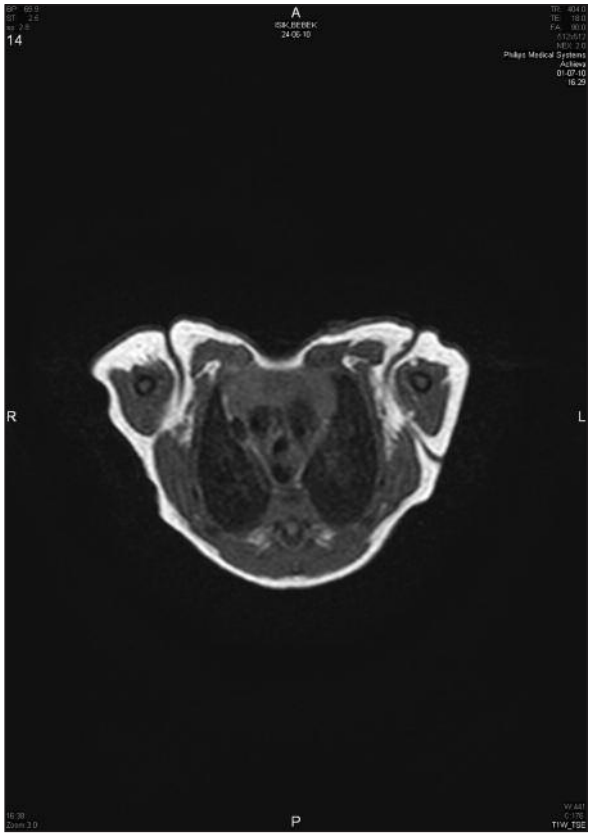

FIGURE 2: Axial T1-weighted MR image demonstrates superior sternal cleft.

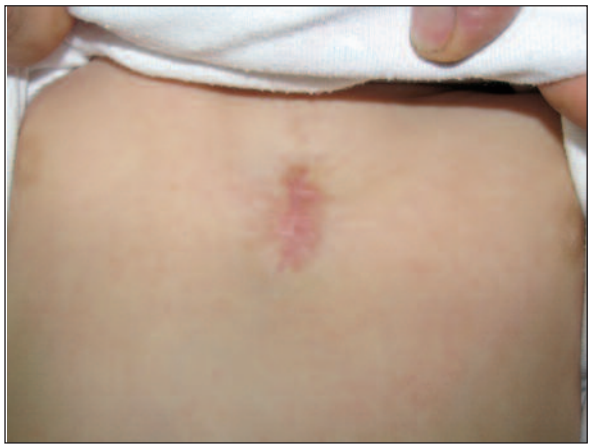

FIGURE 3: The photograph of the infant's anterior chest wall shows the current situation after primary surgical repair at the eighth month of age. Informed consent was obtained from the parents of the patient.
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ported series are small, there is a marked female predominance (8:1).[5] Similarly, in the series of de Campos et al., only one patient out of eight was male. Likewise, our patient was a female infant.[6]

Congenital sternal cleft is classified as complete or incomplete form. Very rarely, the sternal bars fail to fuse in the midline, leading to complete sternal cleft.[7] This leaves the underlying mediastinal structures including the heart and great vessels vulnerable to external trauma. Incomplete clefts are subdivided into superior and inferior forms. Inferior incomplete cleft is almost always associated with other developmental defects of the anterior chest wall such as ectopia cordis or Cantrell's pentalogy. Cantrell's pentalogy is a malformation that results in the combination of defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. These five defects form the basis of the pentad, which was also determined as thoracoabdominal ectopia cordis.[6] Superior defects are usually isolated anomalies sometimes associated with cervicofacial hemangiomas. The most frequently associated anomaly is a medial raphe. Medial raphe shows a high association with cervicofacial hemangiomas, and rarely, with severe cardiovascular malformations.[5,8] A congenital sternal cleft not associated with cardiac defects or ectopia cordis is a rare and relatively benign malformation of the chest wall. The lesion of our patient was isolated, and no other anomalies such as hemangiomas or cardiovascular malformations were accompanying.

A number of methods for repair of cleft sternum have been reported, including primary approximation, sliding or rotating chondrotomies, and the use of prosthetic grafts or flaps of bone, cartilage, autogenous tissue, or the pectoralis major muscle.[9] Of these techniques, primary closure is generally safe and easy. However, the age of the patient is very important for primary repair. After the neonatal period, and certainly after one year of life, primary repair is difficult.[2,4,10,11] The skin defect of our patient was small and underlying structures of the cleft sternum were covered with epithelium. Thus, we initially preferred to follow the patient for spontaneous epithelization of the skin defect with daily dressing. However, since this did not occur, primary surgical repair was performed at the eighth month of age. In our opinion, because primary closure is generally safe and easy, timing is crucial for surgery both for technical facility and good aesthetic results.

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