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Case Report / Olgu Sunumu

Repair of a complete sternal cleft in a five-month-old female infant

Beş aylık kız bebekte komplet sternal kleft onarımı

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ABSTRACT

Failed midline ventral fusion of sternal bars is the cause of complete sternal cleft, which is a rare congenital anomaly that may cause cardiopulmonary compromise. Very few cases of complete sternal cleft have been reported so far in the literature. Surgical correction is recommended to protect mediastinal structures and to restore respiratory dynamics. Herein, we present a case of complete sternal cleft in a five-month-old female infant which was repaired using patch, titanium plate, and bilateral pectoralis muscular flap.

Keywords: Chest wall deformity, sternal cleft, sternal reconstruction.

ÖZ

Sternum barlarının orta hatta birleşmesinin başarısız olması nadir bir doğumsal anomali olup, kalp ve solunum problemlerine neden olabilen komplet sternal kleftin sebebidir. Literatürde şimdiye kadar çok az sayıda komplet sternal kleft olgusu bildirilmiştir. Mediasten yapılarını korumak ve solunum dinamiklerini eski haline getirmek için cerrahi düzeltme önerilir. Bu yazıda, beş aylık bir kız bebekte yama, titanyum plak ve bilateral pektoralis musküler flep ile tamir edilen komplet sternal kleft olgusu sunuldu.

Anahtar sözcükler: Göğüs duvarı deformitesi, sternal kleft, sternum onarımı.

Congenital sternal cleft (SC) is a rare idiopathic congenital chest wall malformation occurring in 2:100,000 live births, with a female predominance and comprising 0.15% of all chest deformities.[1] The SC results from a partial or total failure of sternal fusion of the mesenchymal cells in the ventral midline in the early embryological development, and is classified as "complete" or "partial".[2] Complete SC is extremely rare, and only 35 cases have been reported in the English literature.[2-7] Sternal cleft may be isolated or associated with other defects, such as pectus excavatum, aortic arch malformation, and a component of pentalogy of Cantrell or posterior fossa anomalies, hemangioma, arterial-cardiac-eye anomalies (PHACES) syndrome.[2] It may cause paradoxical respiratory movements and the risk of harmful events is increased on mediastinal viscera. Surgical repair is recommended, if possible in the earlier period of life, to improve pulmonary and cardiac mechanics, protect mediastinal structures, and cosmesis.[2-4]

In this article, we report a rare case of complete SC in a five-month-old girl repaired using patch, titanium plate, and bilateral pectoralis muscular flap.

CASE REPORT

A five-month-old female patient was referred due to a midline sternum and skin defect on the anterior

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chest wall. The defect was diagnosed postnatally in an external center in Iraq. Her physical examination demonstrated a membranous skin covering the defect and visible cardiac pulsations through the defect (Figure 1a). There were paradoxical movements of the anterior thorax during breathing. The patient was born at the 37th gestational age with a low birth weight. She was the only child, and the mother had no family history (including consanguine marriage) or congenital anomalies. As the mother reported, the baby had no further complaints such as difficulty in breathing, cyanosis, and feeding problem. Echocardiography demonstrated only a mild patent ductus arteriosus. Contrast-enhanced computed tomography of the chest and three-dimensional reconstruction revealed an absent sternum and persistent left superior vena cava (Figure 1b). The examination of the rest of the body and the laboratory tests were all normal. Based on these findings, the patient was diagnosed with an isolated complete SC and a surgical repair was planned.

The skin on the defect was raised separating it from the pericardium, but a part of the pericardium was resected and sutured primarily due to dense adhesion to the skin. Ends of the ribs were exposed bilaterally (Figure 2a). The horizontal and vertical diameters of the defect was measured as 6.0 and 5.0 cm, respectively. Primary closure of the defect was not performed due to the risk of the compression of the mediastinal viscera. A patch (Gore-Tex® Acuseal Cardiovascular Patch, W.L.Gore&Associates, AZ, USA) was placed above the pericardium and, then, attached to the ribs with prolene sutures (Figures 2b). Since the use of the patch only provides a semirigid reconstruction, the patient may be under risk for anterior chest wall instability including chest wall retractions leading to higher oxygen requirements, we placed a titanium plate on the patch as a more rigid option, as previously described.[2] The plate was sutured only to the patch and not attached to any structure of the patient to provide the normal growth of the neighboring structures of the patient (Figure 2c). Bilateral pectoralis muscular flaps were advanced to the midline over the patch and plate, and sutured to each other (Figure 2d). Two drains (Hemovac®, Zimmer Biomet, USA) was placed one beneath the muscular flap and the second between the flap and the skin, and the skin was closed with primary suture (Figure 2e).

The postoperative course was normal and the wound healed without any problem. The patient was extubated on the same day and discharged on Day 15 with appropriate antibiotic treatment. A control echocardiography on Day 8 demonstrated no cardiac compression. Follow-up at the first year after the operation demonstrated that she was growing up without any health problem, and the control echocardiography was normal.

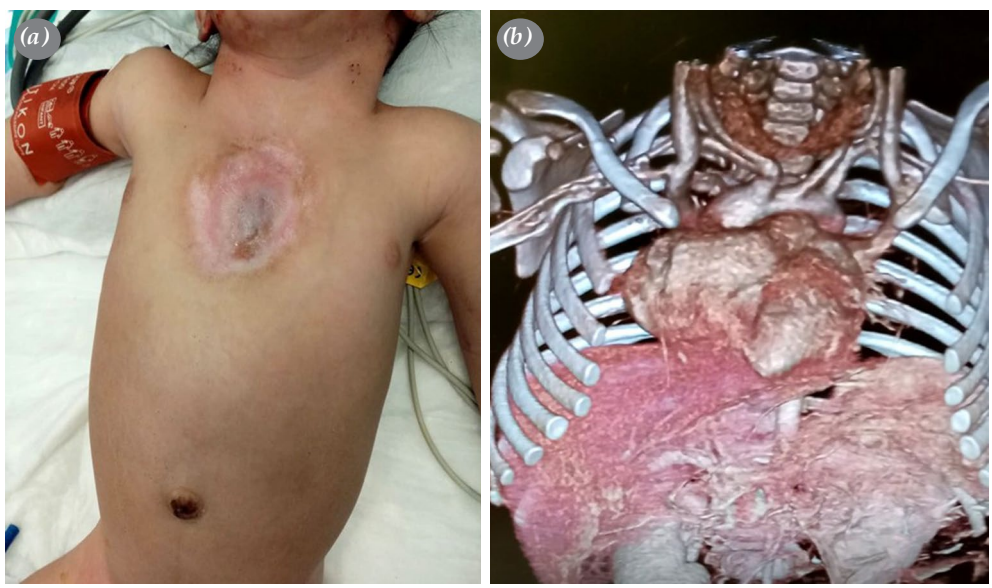

Figure 1. (a) Preoperative image of the patient demonstrating the defect: a membranous skin over the pericardium. **(b)** Three-dimensional reconstruction of the computed tomography of the chest: no sternum is visible.

Figure 2. (a) The defect after resection of the skin and suturing the pericardium. **(b)** Reconstruction using patch. **(c)** Titanium plate sutured on the patch. **(d)** Bilateral pectoralis muscular flaps reinforcement. **(e)** Postoperative image of the patient.

DISCUSSION

Medial migration of cells from plates of lateral plate mesoderm on either side of the developing chest wall forms the sternum, which is completed by 10 weeks. A defect in this fusion causes SC, which can be complete or partial.[2,3] Complete SC is extremely rare. Review of the English literature regarding the patients with SC revealed only 35 cases.[2-7] Therefore, our patient is the 36th reported case of complete SC.

Since 50 to 72% of SCs are associated with other defects, such as PHACES syndrome, pentalogy of Cantrell, cardiac defects, supraumbilical raphe, aortic malformation, pectus excavatum, maxillofacial defects, Dandy-Walker syndrome, a thorough screening for these abnormalities is required before a surgical intervention.[2,3,7] Our patient had no other congenital anomalies, except for a persistent left superior vena cava.

Surgical repair is indicated to protect the mediastinal viscera, to improve respiratory dynamics and cosmesis. Even for asymptomatic patients, sternal repair is needed to maintain normal mediastinal and pulmonary growth and to offer a good cosmesis.[2-4] Our patient had cosmetic problems without any additional health problem.

Timing of repair is dictated by the physiological status of the patient. If the patients have altered pulmonary mechanics and intracardiac defects, surgical intervention should be performed as soon as possible. On the other hand, when there are no additional anomalies requiring urgent repair, surgery may be delated for several months so that the baby gains weight.[2-5] Our patient had no health problem, except for low birth weight, and was referred to our clinic when she reached normal body weight.

Primary suturing is more likely preferred in patients younger than four months, since chest wall is more elastic.[2,7] It was mentioned that after three months of age, the chest wall becomes relatively rigid, and more complicated techniques may be needed, such as use of protheses, partial or total thymectomy, sliding chondrotomies and clavicle dislocation.[4,6] Many different materials for sternal reconstruction have been used including permanent patches, and plates made of titanium, polyethylene, poly-L-lactic acid.[2,3] The use of the prostheses is a valid alternative to the primary closure in older or difficult cases.[3] Primary repair may cause mediastinal compression; if such signs occur during surgery, alternative approaches should be performed.[4] Flap closure should be considered in all cases to protect the repair and decrease tension

on the overlying skin closure. Pectoralis, rectus, and latissimus muscle flaps are used for this purpose.[2] Since our patient was five-month-old and the defect size was large, we repaired the defect using patch and titanium plate, and pectoralis muscle flap.

A common complication is prolonged postoperative ventilatory requirement, that mostly develops following primary repair. Another common complication is the development of seroma, when prostheses and grafts are used.[2] The development of chest wall deformities such as pectus excavatum is a long-term complication and damage to the mammary gland in female patients may occur, when pectoralis muscle flap is used.[3] The postoperative course of our patient was uneventful. No signs of cardiac compression were detected in echocardiography obtained one year after surgery. No signs of chest wall deformity were reported by her mother, either.

In conclusion, sternal cleft is a rare chest wall deformity that necessitates surgical repair. Accurate physical examination is mandatory to rule out possible associated syndromes and anomalies. Primary closure is advised in the early months of life, but in older patients and patients with larger defects, the use of prostheses and more complicated techniques should be required.

Patient Consent for Publication: A written informed consent was obtained from the parents and/or legal guardians of the patients.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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