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Case Report / Olgu Sunumu

Surgical repair of a sternal cleft malformation

Sternal kleft malformasyonunun cerrahi onarımı

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ABSTRACT

Sternal cleft is a unique congenital chest wall malformation that results from failure of sternal fusion early in the embryological development. Surgical correction is advised both to protect mediastinal structures and to restore respiratory dynamics. Early surgical correction, preferably in the neonatal period, is recommended in order to benefit from the elasticity of the thoracic cage. In this article, we present a two-month-old female patient with a superior V-shaped sternal cleft, which was successfully corrected with posterior periosteal flap, sliding chondroplasty and cartilage graft interposition techniques.

Keywords: Chest wall deformity, chondroplasty, sternal cleft.

ÖZ

Sternal kleft, embriyolojik gelişimin başlarında sternal füzyonun başarısızlığından kaynaklanan, nadir görülen, doğumsal bir göğüs duvarı malformasyonudur. Cerrahi düzeltme hem mediastinal yapıları korumak hem de solunum dinamiklerini yeniden sağlamak için tavsiye edilir. Göğüs kafesinin esnekliğinden faydalanmak için tercihen yenidoğan döneminde erken cerrahi düzeltme önerilir. Bu yazıda, posterior periosteal flep, sliding kondroplasti ve kıkırdak greft interpozisyonu teknikleriyle başarılı bir şekilde düzeltilmiş, V şeklinde sternal klefti olan iki aylık bir kız hasta sunuldu.

Anahtar sözcükler: Göğüs duvarı deformitesi, kondroplasti, sternal kleft.

Sternal cleft (SC) is a rare idiopathic congenital chest wall malformation that results from a partial or total failure of sternal fusion mesenchymal cells in the ventral midline early in the embryological development.[1] It occurs in 2:100,000 live births, with a female predominance.[2] They are often associated with other malformations such as hemangiomas, pentalogy of Cantrell, PHACES syndrome, gastroschisis, omphalocele, pectus excavatum and ectopia cordis.[3,4]

Complete SC is clearly diagnosed at birth due to abnormal movements of the thorax. Surgical correction is recommended to protect mediastinal structures and heart from direct injuries and to restore respiratory dynamics. Early surgery, optimally in the neonatal period, is preferred in order to benfit from the elasticity of the thoracic cage and achieve primary closure.[5-7] Apart from primary closure, different surgical techniques have been described such as bone graft interposition, prosthetic closure, muscle flap interposition, sliding chondroplasty, and clavicle dislocation.[2,8,9] In this article, we present a two-month-old female patient with a superior V-shaped SC which was successfully corrected with posterior periosteal flap, sliding chondroplasty and cartilage graft interposition techniques.

CASE REPORT

A two-month-old female patient was referred to our department with the clinical diagnosis of a superior SC. She was diagnosed postnatally by clinical examination in another institution. Her diagnosis was confirmed by thorax computed tomography (CT) including three-dimensional reconstruction views (Figure 1). The cardiac ultrasound showed no anomaly except a small patent foramen ovale. Her physical examination revealed paradoxical movements of the anterior thorax during inspiration and expiration. There was a big wall defect of approximately 3×4 cm between the medial

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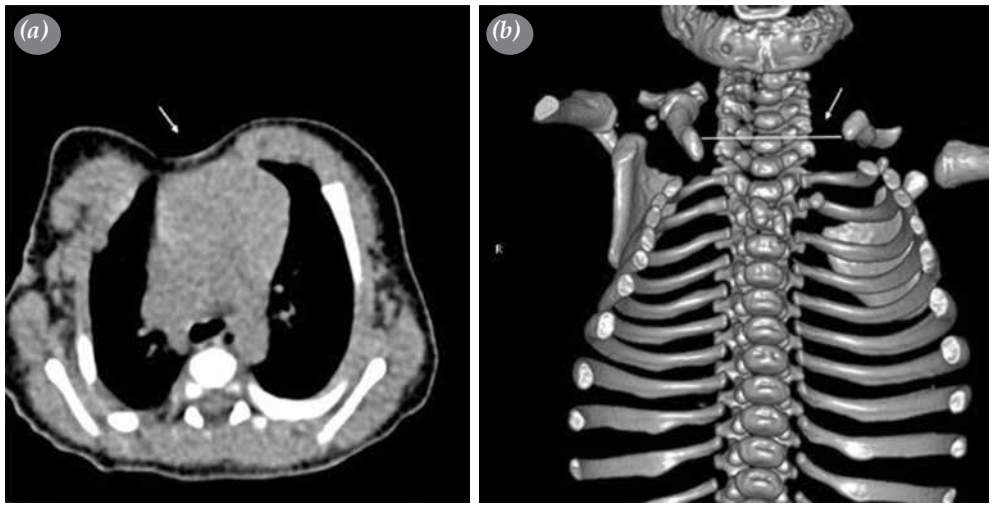

Figure 1. Thorax computed tomography **(a)** and three-dimensional reconstruction **(b)** views of defect.

extremities of the right and left ribs, covered by skin. The beating heart was visible through the skin. The examination of the rest of the body and the laboratory tests were normal. A written informed consent was obtained from the legal guardians of the patient.

A vertical midline skin incision along the defect was performed. She had a V-shaped superior sternal cleft. The horizontal and vertical diameters of the defect was measured as 3.6 and 3.0 cm, respectively. Bilateral sternal bars were released from the underlying structures, thymus gland was completely removed and the mediastinal tissues were freed from each sternoclavicular joint. The fused inferior portion of the sternum was separated by a midline osteotomy by removing costal cartilages bilaterally.

The perichondrium was elevated completely on each sternal half to expose the underlying cartilages and the periosteal flap was folded medially to contribute to the closure of the defect. The perichondrium of three costal cartilages on each side was also elevated. Sliding chondroplasty was performed by performing Z-shaped incisions through the costal cartilages to produce

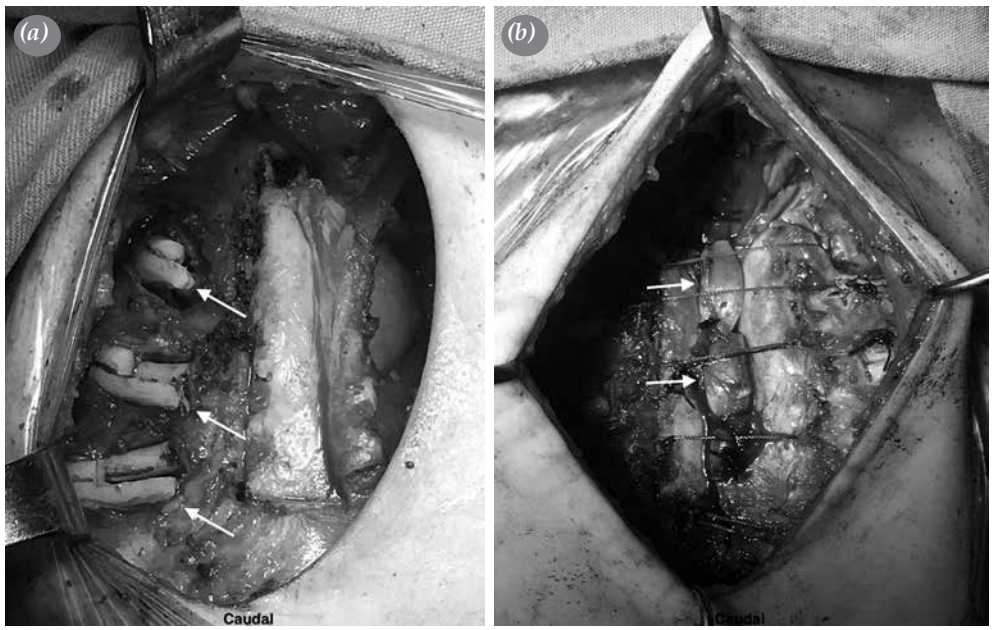

Figure 2. (a) Sliding chondroplasty of costal cartilages (arrows) and **(b)** autogenous cartilage grafts in midline of approximated sternum (arrows).

greater length and allow further approximation of the sternal halves (Figure 2a). The sternum was then approximated with several Ethibond® polyester sutures (Ethicon Inc., Somerville, NJ, USA). Previously removed costal cartilages were used as autogenous grafts in the midline of the approximated sternum (Figure 2b). To ensure that cardiac and respiratory functions were not compromised due to the increased intra-thoracic pressure, the patient was observed for five min. The periosteal flaps of cartilages on each side were primarily closed with Vicryl® polyglactin sutures (Ethicon Inc., Somerville, NJ, USA). Bilateral muscle flaps were created from the pectoral muscles and approximated on the midline. The wound was then closed in layers with absorbable sutures.

DISCUSSION

Sternal cleft is a rare midline deformity with an incidence of <1% of all chest wall deformities.[10] During the embryonic development, the sternum originates from two mesenchymal bars. Mesodermal cells on either side of the anterior chest wall fuse in the midline between the 6th to 10th week of gestation.[1,11] Sternal cleft can be classified into complete and partial forms, which the latter can be subdivided to superior or inferior types depending on when the developmental process is interrupted. Nevertheless, its exact etiology is still not known.

The diagnosis of SC is usually established by postnatal physical examination. The paradoxical respiratory movements and the beating of the heart under the skin are suggestive. Diagnosis can be confirmed by imaging techniques such as ultrasonography, CT or magnetic resonance imaging. Echocardiography should be performed preoperatively to rule out associated cardiac malformations.[2] Without surgical correction, the defect can potentially increase both the risk of trauma-related injury to the underlying structures and respiratory dysfunction due to unpaired gas exchange.[12,13]

Several surgical techniques have been described in the literature for repair of SC, including primary approximation, sliding osteochondroplasty, use of autologous grafts (rib, bone or muscle), prosthetic repair (Teflon, polypropylene, silicone or titanium) and biologic tissue grafts (extracellular dermal matrix).[2,6,14,15] The age of the patient is the most important factor for the selection of the surgical procedure. Current opinion for the management of SC is to perform primary surgical closure within the newborn period.[2,8,10] Some authors expand the ideal closure time to first three months of life.[3,16] During this period, the flexibility of thorax is maximal and the risk of compression to the underlying structures is minimal. Thus, primary closure could be performed easier, with a lower risk of cardiovascular compromise. Beyond this period or to close larger defects, additional procedures such as sliding chondroplasties, osteotomies, clavicular dislocation and the use of autologous grafts or prosthetic materials are recommended due to the increased pressure of the reduced space and lower compliance of the chest wall which could impair cardiovascular function.[2,10,16,17] Although our patient was a two-month-old child, her rib cage was too stiff and the V-shaped defect was a large. Midline closure was not possible without performing sliding chondroplasty and use of autologous cartilages. It is also recommended to perform a partial or complete thymectomy to create more space for primary closure and to reduce the risk of mediastinal compression.[18] In addition, the approach of elevating perichondrial flaps is advantageous to approximate cartilages and to facilitate primary union.[19]

The complication rate of SC repair is low. Mortality is seldom reported and related to coexisting additional malformations rather than to surgical intervention. However, patients should be followed-up for development of other chest wall deformities such as pectus excavatum for the long-term.[2,13]

In conclusion, SC is a rare congenital chest wall malformation mainly diagnosed in newborns. Surgical closure of the defect should preferably be performed in the neonatal period. Detailed physical examination and cardiovascular imaging prior to surgery are important to rule out associated malformations. Choice of proper surgical technique mainly depends on the age of the patient and the results of surgical repair and complication rates are satisfactory.

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