Case Report

Primary closure of sternal cleft in a 4-year-old child

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Abstract

A cleft sternum is a rare congenital anomaly often diagnosed as asymptomatic at birth. Clinical outcome may be unfavorable when an associated anomaly, particularly an intracardiac anomaly, coexists with the defect. Primary repair should be employed in the neonatal period because the flexibility of the chest wall is maximal and compression of underlying structures is minimal. However, patients with sternal cleft may even present late in the childhood or adolescence period. We herein report a case of a 4-year-old girl with sternal cleft who showed a favorable clinical outcome following successful primary surgical repair with the use of autogenous tissues.

Keywords: sternal cleft, repair, autogenous tissue

Introduction

Sternal cleft is the separation of the sternum with orthotopic normal heart and normal skin coverage(1). Sternal clefts are extremely rare and very few cases have been reported(2). They may be classified as complete or incomplete with the complete cleft being the rarer form. They may be attended with a high mortality in the early postnatal period resulting from the coexisting cardiac anomalies and the difficulty of repositioning the heart without circulatory impairment(2). Isolated sternal clefts, however, have a favorable prognosis because they present without intracardiac anomalies, which allows a potential for primary repair of the defect. Repair of a sternal cleft should preferably be employed early in the neonatal period when the thorax is relatively more compliant, and the primary closure is generally safe and easy(2,3). Some cases, however, may be omitted at birth, and they may present late in the childhood or adolescence period. We report here a case of 4-year-old girl presenting with a sternal cleft who was admitted late in her childhood. The patient
underwent a successful surgical repair with
the use of autogenous tissues.

Case Report
A female, 4 years of age, from Shendi, came
complaining of abnormal swelling in the
anterior chest wall since birth. The patient is
an outcome of SVD at home, cried and
breastfed immediately; she is well nourished
and weighed 16 kg.
The mother noticed that her baby had defect in
her upper chest that protruded with breathing
and crying. There were no other complaints or
symptoms related to other systems.
The patient looked well, not pale, jaundiced or
dehydrated.
Her pulse rate was 100 bpm and respiratory
rate was 34 cpm.
Chest examination revealed a wide defect in
the upper sternum with thin skin layer
covering the defect, the skin moved
paradoxically with the breathing movement,
great vessels pulsation could be felt in the
defect cardiac examination was normal. The
abdomen was normal (Fig 1).

Fig 1: The sternal defect before surgery

Laboratory investigations were within the
normal levels, chest CT showed incomplete
sterna cleft, otherwise chest is normal.
Echocardiography showed normal heart.

Management
The skin overlying the sternal defect was
incised as an elliptical shape incision. The
pericardium, heart and the thymus were
normal. The sternal bars were present on each
side showing a U-shaped incomplete sternal
cleft extending to mid sternum with the
sternocleidomastoids widely separated.
Initially, the redundant endothorathic fascia
was excised and sutured continuously. The
periosteum of each sternal bar was incised on
its lateral border, and the flaps were sutured
together in the midline with 3/0 absorbable
sutures. Followed by many layers of fascial
coverings which were approximated and
sutured with 3/0 absorbable sutures. Similarly,
the sternal bars were approximated with four
interrupted 0 nonabsorbable sutures in the
midline. Pectoralis major muscles flaps were
also raised and approximated in the midline,
and finally, sternohyoid, sternothyroid and
sternocleidomastoid muscles were
approximated medially to avoid a possible
lung herniation. The skin and the
subcutaneous tissues were primarily closed
(Fig 2).

Fig 2: The defect after healing of the wound:

No evidence of cardiac compression was
noted, and the patient remained
ehemodynamically stable both throughout the
procedure and in the postoperative period. The
patient was doing well in the subsequent
postoperative follow-up visits.
Discussion

Sternal cleft is a rare congenital anomaly, which refers to the upper sternal cleft or bifid sternum. It has a multifactorial etiology without any familial basis, and it is generally observed at birth without symptoms \(^4\). It results from the fusion failure of the sternum. Sternal cleft may be V-shaped, when the cleft reaches the xiphoid process, or broad and U-shaped, with a bony bridge joining the two of the developing clavicles. The sternal bars may sometimes fail to join in the midline, which results in a complete sternal cleft \(^5\). Although the fusion of the sternal bands normally starts from the cephalic end, an upper failure of fusion should result in a cleft of the whole sternum; nevertheless, the upper sternal cleft with fusion of the distal part is the most common form of sternal clefts \(^6\). 

A possible explanation lies in a primary absence of the cephalic single element, the pre sternum, or in a secondary splitting \(^7\). Kaplan and associates \(^8\) hypothesized that ectopia cordis results from mechanical compression secondary to rupture of the chorion or yolk sac. Rupture of these structures at three weeks’ gestation would interfere with cardiac descent, internal cardiac development, and midline fusion of thoracic structures. Later rupture between the sixth and the ninth weeks might result in cleft sternum alone due to thoracic compression after cardiac descent and development. An early disturbance in the development of midline mesodermal structures that interferes with fusion of the lateral sternal bands and overlying cutaneous tissue has also been postulated to explain the association of sternal malformation and vascular dysplasia \(^8\).

Prenatal diagnosis has not been defined in the literature; however, prenatal ultrasonogram, reevaluated after birth, showed the presence of an upper sternal cleft in only one case \(^9\). The diagnosis of sternal cleft is easily done at birth by inspection and palpation. Diagnostic investigations are thus directed to exclude the edges, ending at the third or fourth costal cartilages as in our case \(^5\). The embryology of the sternal cleft remains obscure. In embryonic life, the sternum originates from the lateral plate mesoderm. Cells from two bands of mesoderm on either side of the anterior chest wall migrate toward the midline, and become fused by the tenth week to form the sternum. The manubrium is formed by primordia between the ventral ends of the developing clavicles. The cardiopulmonary system progressively accommodates to the size of the thorax following the first 3 months of age, and the chest wall becomes firm. Thus, numerous authors agree that the optimal choice of treatment is the primary direct closure in the neonatal period with autogenous tissues, when flexibility of the chest wall is maximal and compression of underlying structures is minimal \(^2,3,6\). Nevertheless, hypoplastic nature of the sternal remnants and the width of the cleft may sometimes preclude primary repair \(^10\). Hence, more complicated procedures such as implantation of autologous grafts such as costal cartilages, parietal skull, tibial periosteum, and the use of prosthetic materials such as stainless steel mesh, Marlex, acrylic, silicone elastomer or Teflon have been suggested as alternative approaches \(^2,3,6\). Although our patient presented late in her infancy, we were able to accomplish a successful primary repair by performing incision of periosteum of each sternal bar on lateral border and using the autogenous tissues. 

Although surgical repair should be performed in the neonatal period in patients with sternal cleft, a safe and favorable operation may also be performed with the use of autogenous tissues even in late infancy.
References