Case report - Thoracic general

Primary repair of a sternal cleft in an infant with autogenous tissues

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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; Surgery; Congenital anomaly

1. 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 being the complete cleft the rarer form. They may characterize 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 [1]. 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 herein a case of a 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, and she remains well 19 years after the operation.

2. Case report

A 4-year-old girl presented with a bony defect in the central upper part of her chest wall since her birth. On physical examination, a wide gap at the upper site of the sternum was observed (Fig. 1A). Pulsations of the great vessels could easily be seen through the defect, which was covered by a thin layer of skin. Diastasis of the upper third of the rectus abdominis muscle was also observed. Laboratory examinations were within the normal limits. A chest roentgenogram and computed tomography scans (Fig. 2) showed widening between the medial ends of the clavicles with the absence of the manubrium and ossification centers in the upper third of the sternum resulting from the agenesis of the sternum. Ultrasonography revealed the thymus underneath a thin cutaneous coverage. Echocardiography did not reveal any coexisting cardiac abnormality. The patient had undergone an operation with the diagnosis of sternal cleft. The skin overlying the sternal defect was incised along the midline corresponding with the virtual suprasternal notch to the epigastric region. The pericardium, heart and the thymus were normal, however abdominal rectus muscle showed a diastasis. The sternal
bars were present on each side showing a U-shaped incomplete sternal cleft extending to the lower half of the sternum. The connection site of the sternal bars was the level of the fourth costal cartilages. Initially, a vertical wedge osteotomy was performed at the most upper part of the bridge between the sternal bands. Thereafter, chondrotomies were performed on the costal cartilages of the first four ribs on each side at the costochondral junctions to release the sternal bars. 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. Similarly, the sternal bars were approximated with four interrupted 0 nonabsorbable sutures in the midline. Pectoralis major muscles were also approximated in the midline, and finally, sternohyoid, sternothyroid and sternocleidomastoid muscles were approximated medially to avoid a possible lung herniation. The diastasis in the rectus muscle was repaired with sutures. The skin and the subcutaneous tissues were primarily closed (Fig. 1B). No evidence of cardiac compression was noted, and the patient remained hemodynamically stable either throughout the procedure or in the postoperative period. A retrosternal hematoma, collected in the postoperative course, was drained properly. The patient is doing well 19 years after the operation.

3. 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 edges, ending at the third or fourth costal cartilages as in our case [3].

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 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 infrequent associated anomalies. These consist of a bandlike scar from the umbilicus to the sternal defect or from the sternal defect to the chin and cervicofacial hemangiomas. Diastasis recti may also be associated with a sternal cleft as in our case [2].

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 peristosteum, 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 chondrotomies on the involved costal cartilage 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