Case Report

Isolated Central Sterna Clefts: A Rare Congenital Malformation

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Abstract

Sternal cleft is a rare congenital anomaly which is generally observed at birth. The aetiology remains obscure. Superior clefts are more frequent than inferior ones, and isolated central clefts are extremely rare. Surgery is recommended to protect the heart and other mediastinal contents from trauma and also to improve respiratory dynamics. We present the case of a newborn with isolated central clefts and we will give a review of the literature.

Keywords: Congenital malformation, new-born, sternal cleft, surgery

Introduction

Sternal cleft is a rare congenital malformation due to partial or total failure of sternal fusion at an early stage of embryological development. We present the case of a new born with partial sternal cleft (isolated central clefts). We describe the method of repair of this anomaly and evolution.

Case

A 4400 g full term girl was born by normal delivery. There was no prenatal diagnosis. The clinical examination showed [Figure 1] a baby with impressive paradoxical movements of the thorax during inspiration/expiration. The gap occupied the lower half of the sternum. The xiphoid process was present. There was a wall defect of approximately 3 cm width, covered by a thin skin. The skin behind the defect was angiomatous. The beating heart was visible through the skin. The rest of the exam showed a heart murmur, low implanted ears and macroglossia.

The laboratory tests were within normal limits, malformatives investigations showed only a 3 mm ventricular septal defect.

The surgery was performed on day 12. The skin was incised along the defect and excided after her dissection and separation from the under sterna fascia. Closure by separated sutures could bring two sterna edges side by side. The patient's compliance at the thoracic closure was tested. The sutures were tight and the sterna bars were brought together on the midline [Figure 2]. The vital signs remained stable.

Postoperatively, the baby remained intubated and ventilated 1 day to achieve a good tolerance of the intrathoracic pressures. The recovery was uneventful. The patient was discharged 5 days later.

The follow-up at 6 years revealed no complications. Moreover, echocardiography revealed spontaneous ventricular septal defect closure.

Discussion

Sternal clefts are congenital malformations of the anterior chest wall in which the left and right hemi sternum fail to fuse or develop during foetal life. The malformation is rare, with <100 cases reported in the literature.[1,2] The sterna cleft may be partial or total, depending when the development process stopped. The defect could be superior, inferior or complete.[3]

The superior cleft is often associated with other malformations such as facial haemangioma or abdominal raphe, the inferior cleft to the pentalogy of Cantrell (ectopic cordis, intracardiac defects, sterna cleft, omphalocele and pericardial defect allowing communication with peritoneal cavity). The complete

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Figure 1: Pre-operative photograph of the patient's anterior chest wall shows incomplete sternal cleft

sternal cleft is the less frequent form. Rare association with other malformation has been met: gastroschisis, VACTERL syndrome, pectus excavatum and chest wall haemartoma.[3-5]

Many aetiological factors had been incriminated (nutritional deficiency in the mothers, consanguineous families, high frequency from Middle East) but none of them was clearly pointed out.[1,6]

The diagnosis can be easily done clinically at the birth by inspection and palpation. Prenatal diagnosis was taken into consideration but its place is not very clear yet. It seems to be more easily identified when it is associated with cardiac anomaly, because of the attentive research of other existing malformations.

Although this malformation could be diagnosed at birth, the presentation varies from the neonatal period to adolescence, being asymptomatic. There is a general agreement that sternal cleft should be repaired in the neonatal period with autogenous tissue because the flexibility of the thorax is maximal and compression of the underlying structures is minimal. Hence, the closure of the defect is better tolerated in the first 3 months of life. Even in this period, some technical difficulties could be encountered (e.g., necessity of partial removal of the thymus for space constraints in newborn).[4]

For the correction of the defect many surgical techniques were used: closure of the defect using pectoral's major advancement flap, a sternal reconstruction with autologous tissue,[1] cartilage and sterna periosteum mobilisation and approximation of sterna sternocleidomastoid muscles with closure achieved with costal homograft and prosthetic mesh. In older patients, autologous grafts or a synthetic patch such as marlex, acrylic, silicone elastomer or Teflon are used to cover the defect. In our case, the surgery consisted only in chest wall closure and it was well supported.[3,7,8]

Isolated sternal cleft repair is best performed in early infancy, within the 1st month of the life, because the thoracic cage is more flexible as a result of elasticity and expandability of cartilage in the neonatal period, which allows direct sternal repair.[9]

Combined repair of both sternal cleft and congenital heart disease in early months of life has been reported with good

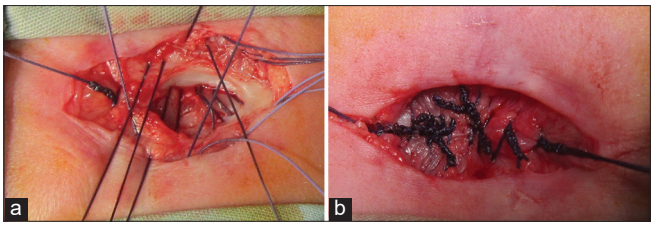


Figure 2: (a) Operative view of the sternal cleft, (b) Immediate postoperative result

result. In contrast, postponement of sternal reconstruction after congenital heart disease repair is another strategy in early infancy but may compromise respiratory function with prolonged length stay in the intensive care unit.[9]

Conclusion

Depending on the size of the defect, the sternal cleft may be treated or not. The indication for the surgery are cosmetic but are also to improve respiratory dynamics and to protect the mediastinal structures from direct injuries, in order to allow a normal life.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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