

Case Report

An Infant with Congenital Sternal Cleft and Ventricular Septal Defect

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Abstract

Congenital midline sternal cleft is a rare anomaly, and can be recognized at birth due to the typical appearance as a sternal depression covered with atrophic skin with a median abdominal raphe extending down to the umbilicus. Such infants should also be examined for lesions in the brain, heart, and eyes as sternal clefts are usually described as part of PHACE Syndrome and Pentalogy of Cantrell, or as part of sternal malformation/vascular dysplasia association. Isolated sternal cleft without any associated anomalies are very rare. Here, we describe a female infant who was noted as having a sternal cleft and an examination for associated malformations revealed a ventricular septal defect.

Abbreviations

VSD- Ventricular Septal Defect, CT- Computerized Tomography, MRI- Magnetic Resonance Imaging.

Keywords: Sternal Cleft; Abdominal Raphe; Heart Malformation.

Introduction

Congenital midline sternal cleft is a rare anomaly that is usually described as part of a defined syndrome like PHACE Syndrome (posterior fossa brain abnormalities, hemangiomas, arterial anomalies in the cranial vasculature, coarctation of the aorta/cardiac defects, eye abnormalities and sternal defects) [1] and Pentalogy of Cantrell, [2] or as part of sternal malformation/vascular dysplasia association [3]. The incidence of congenital sternal cleft has been reported to be fewer than 1 in 100000 live births [4]. A depression in the sternum, covered with atrophic skin with a median abdominal raphe extending from the sternal depression to the umbilicus is the presenting feature. Such children may also have internal vascular lesions in the brain, heart, and eyes [3]. Here, we describe a female infant who was noted as having a sternal cleft and an examination for associated malformations revealed a ventricular septal defect.

Case Report

A 2680 g healthy female baby was born via cesarean section to a 30-year old woman with a previous abortion. Antenatal workup was normal. The baby had a 5.5 cm-long midline abdominal raphe running down to the umbilicus from the sternum, where the chest

wall appeared depressed (Fig 1). The baby had no other abnormal superficial lesions and clinical examination was normal. Chest radiograph, cranial and abdominal ultrasounds and ophthalmic examination were normal. Echocardiography revealed a 6 mm inlet ventricular septal defect (VSD). The baby had an otherwise uneventful course and was discharged after proper counseling. At the latest follow-up visit at fourteen weeks, the baby was healthy and had normal growth and development.



Fig 1: The patient's chest wall showing atrophic skin over the sternal depression and midline abdominal raphe extending down to the umbilicus

Discussion

The sternum is derived from the mesoderm. In the sixth week of

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gestation, paired concentrations of longitudinal thoracic lateral bands start migrating medially and fuse craniocaudally by the tenth week. Deviation from the normal process of ventral midline thoracic fusion can present as an irregularly-shaped xiphoid, ectopia cordis, or a partial upper or complete sternal cleft [5].

The typical clinical features include a sternal cleft that is covered with atrophic skin, a midline abdominal raphe extending from the sternal defect to the umbilicus with an orthotopic heart [3, 5]. The defect can be shaped like a narrow 'V' or a wider 'U'. A partial sternal defect involves the upper sternum and manubrium. In thoracic and thoraco-abdominal ectopia cordis, the defects are mostly in the lower sternum [5].

Sternal cleft may appear as part of the Pentalogy of Cantrell, which includes omphalocele, anterior diaphragmatic hernia, sternal defect, ectopia cordis and cardiac defects [2].

It may also appear as part of the sternal malformation/vascular dysplasia association with associated cutaneous craniofacial hemangiomas and occasionally, internal vascular malformations on the respiratory tract and viscera [3].

Associated abnormalities that have been reported include absence of anterior pericardium, cleft lip and palate, bifid uvula, micrognathia, and glossoptosis [3]. Associated cardiac defects include transposition of great vessels [6] and truncus arteriosus [7].

The defect may present as part of PHACE Syndrome [1]. Sternal cleft, [4] as well as PHACE Syndrome, [3] have a marked female predilection. It has been suggested that the sternal malformation/vascular dysplasia association and PHACE Syndrome are a single spectrum of anomalies and that these be classified as PHACE Syndrome [3].

In our patient, cutaneous hemangiomas had not appeared till fourteen weeks age, and, although PHACE Syndrome can be diagnosed even without characteristic cutaneous hemangiomas [8], only a cardiovascular anomaly (VSD) was present in addition to the sternal defect, and did not fulfill the criteria. However, considering the variability in presentation of the hemangiomas [1], the possibility of a diagnosis of PHACE Syndrome at a later date cannot be ruled out.

Isolated sternal cleft, without any associated anomalies, has also been reported, though it is quite rare [4]. Thoracic computerized tomography (CT) imaging can be used to visualize the sternal cleft and confirm the diagnosis [3].

The cutaneous hemangiomas are seen on the face, neck and upper trunk, and are superficial and plaque-like to begin with, but later grow and resemble mixed or deep hemangiomas. These do not appear at birth and manifest in early infancy, usually

around 2 months [1, 3]. Hemangiomas of respiratory tract can cause respiratory compromise and those of gastrointestinal tract, bleeding and infection [3].

Thus, a newborn with sternal defect requires evaluation to find associated malformations of the brain, eyes, heart and great arteries, including cranial ultrasound and magnetic resonance imaging (MRI) to rule out posterior fossa defects and intracranial vascular anomalies, complete cardiac examination including echocardiogram, and ophthalmologic examination to detect ocular abnormalities. Afterwards, close follow-up is mandated for development of internal and external hemangiomas, which may lead to potentially life-threatening events.

For the sternal cleft itself, surgery is indicated, and early repair is recommended if it is associated with other chest defects to best gain advantage from the flexible nature of the neonatal rib cage [9]. The pliant cartilaginous thorax in neonates allows easy approximation of the sternal bars by simple suture and reduces chance of cardiac compression. The other corrective procedures include chondrotomy, and the use of prosthetic or natural grafts [10].

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