A RARE CONGENITAL MALFORMATION: ISOLATED STERNAL CLEFT

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Abstract

A twenty-five days old girl, admitted to pediatric pulmonology clinic, with collapse on the anterior chest wall while breathing. Physical examination revealed total retraction of the chest wall especially on the upper part, paradoxical chest wall movements with respiration, and a raphe extending from the umbilicus to the lower end of the sternum. Hypoplasia of the inferior and non-development of the superior segment of the sternum was shown on 3-dimensional(3D) thorax CT. At the age of fifty-six days, she had been operated. Due to the large defect, primary closure couldn't be performed and the sternum was reconstructed with the cartilage obtained from the tissue around the ribs. The sternal cleft is a rare malformation; complete assessment, based primarily on thoracic imaging is required for diagnosis. Also cardiac, abdominal and cranial imaging for concominant malformations should be performed; our patient had none. Immediate surgical management is essential to avoid possible complications.

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To the editor,

Isolated sternal cleft is rare a congenital malformation. It can be isoleted or associated with various cardiac, abdominal, cranial and eye malformations. Diagnosis with radiology. Management is only surgery as soon as possible. Here in case we report a neonate with isoleted incomplet sternal cleft.

CASE:

A twenty-five days old girl, admitted to pediatric pulmonology clinic, with collapse on the anterior chest wall while breathing. Physical examination revealed total retraction of the chest wall especially on the upper part, paradoxical chest wall movements with respiration and a raphe extending from the umbilicus to the lower end of the sternum (Video-1). Other system findings were normal. Evaluation of sternum related bone pathologies, thorax computed tomography (CT) and 3-dimensional (3D) reconstruction imaging was planned. On the thorax CT hypoplasia of the inferior and non-development of the superior segment of the sternum was found (Figure-1). Lung parenchyma was normal. Abnormal findings were not detected in abdominal, cranial ultrasonography, echocardiography and eye examination which were evaluated in terms of pathologies that could accompany with strenal cleft. At the age of fifty-six days, the patient had been operated. Due to the large defect, primary closure couldn't be performed as a surgical procedure and the sternum was reconstructed with the cartilage obtained from the tissue around the ribs.

DISCUSSION:

Sternal cleft is a rare congenital malformation, incidence of 2/100,000 [4], caused by failure in the fusion of sternal rods, on the cranio-caudal direction, in the intrauterine 8-10th weeks [1]. It was first described by De Torres in 1739 [2]. Malformation may be total/parcel of the lower and/or upper part of the sternum, seen in 67% in parsial form [3,4]. It is often sporadic, but similar cases have been reported within the same family [5]. Sternal cleft can be isolated or associated with syndromes such as PHACE, Cantrell Pentalogy, accompanied by various cardiac, abdominal, cranial and eye related malformations [7, 8]. Clinically, paradoxical chest wall movements marked by respiration are pathagnomonic [4]. In adulthood, many cases are asymptomatic, patients may also be diagnosed with symptoms of accompanying malformations or various lung related symptoms [4]. Radiological imaging plays a key role in diagnosis, classification and the search for associated malformations. Computed tomography is the gold standard in diagnosis [4]. Surgical correction should be performed as soon as possible (1-4 weeks in the neonatal period) to prevent possible complications in all cases with or without any symptom [6, 9, 10]. Surgical correction is required in all cases to provide bone protection to mediastinal tissues and organs, to create normal intrathoracical pressure and to eliminate chest deformity regardless of age at diagnosis [9]. In early intervention, primary repair can be applied, in this procedure the sternal bands are simply approached together [3]. In late intervention, complex reconstruction surgeries are required due to the ossification of the cartilage tissue. Surgical procedures such as oblique division of the cartilage, chondrotomy or filling the defect with auto graft or some different materials can be performed [3, 4].

The sternal cleft is a rare malformation; complete assessment, based primarily on thoracic imaging is required for diagnosis. Also cardiac, abdominal and cranial imaging and eye examination should be performed for associated malformations. Regardless of the age at diagnosis, patients should be managed by immediate surgery which is essential to avoid possible complications.

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