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Sternal cleft – A rare congenital malformation

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ABSTRACT

Sternal cleft is a rare congenital malformation. We present the case of a new-born with complete sternal cleft and skin defect, without any other associated anomaly. Operated on in the neo-natal period, the post-operative recovery was good. She developed a transitory right lateral cervical lymphangioma, which resolved spontaneously. The follow up at 4 years revealed no complications.

Sternal cleft is a rare congenital malformation. It is due to a partial or total failure of sternal fusion at an early stage of embryological development. When it is associated with cardiac anomaly, the clinical outcome may be unfavorable. Complete sternal cleft, although asymptomatic, is easily diagnosed at birth, being obvious due to the abnormal movements of the thorax. Early surgery benefits from the elasticity of the thoracic cage. It is optimally performed in the neo-natal period in order to protect the heart and mediastinal structures from direct injuries. We present the case of a new-born with complete sternal cleft and skin defect.

1. Case report

A 2440 g full-term girl, with a complex thoracic malformation, was transferred to our department. She was born by normal delivery. There was no prenatal diagnosis. The clinical examination revealed a baby in good general condition, with impressive paradoxical movements of the thorax during inspiration/expiration. The sternum was absent. In between the medial extremities of the right and left ribs there was a big wall defect of approximately 5 cm width, covered by skin. The beating heart was visible through the skin. The two hemi thoraxes were approaching during inspiration and pushed apart during expiration. The skin of the gap area presented a 3 × 4 cm defect, covered by a yellowish membrane (Fig. 1). The exam of the abdomen and of the rest of the body was without particularity. The laboratory tests were within normal limits. The cardiac ultrasound revealed no anomaly. The X-rays and ultrasound did not show any other thoracic or abdominal malformation, only a complete sternal cleft (Figs. 2 and 3). The surgery was performed on day 12. The skin was incised along the defect. The yellowish membrane was excised. The dissection was continued to the sternal bars. After isolating the bars on each side from the underlying structures, muscle flaps were created from the pectoral muscles. The sternal halves were freshened up. Non-absorbable sutures were placed as stay sutures around the sternal halves. The patient's compliance at the thoracic closure and at increased intra-thoracic pressure was tested for 10 min. A few cardiac rhythm anomalies were noted. These were well tolerated and slowly disappearing. The sutures were tight and the sternal bars were brought together on the midline. The muscular flaps were approximated on the midline, in order to cover the defect. The vital signs remained stable. Aspirativ drainage was left in situ for 3 days. Post-operatively, the baby remained intubated and ventilated for 3 days in order to achieve a good tolerance of the intra-thoracic pressures. The recovery was uneventful. The patient was discharged 10 days later (Fig. 4). Follow-up was done at 3, 6, 9 months and yearly thereafter for a further 3 years. Ventilatory,

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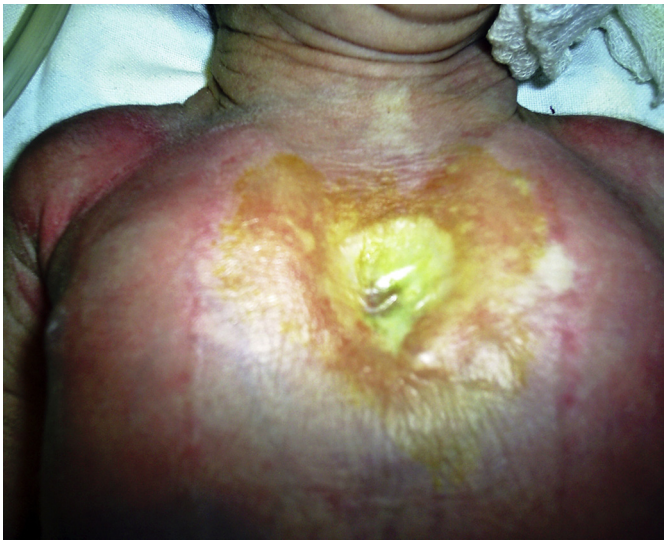

Fig. 1. Pre-operative image of the defect: the "V" shape and the yellowish membrane.

cardiac and weight parameters remained within normal limits for age. In the first few months the child slowly developed a small right lateral cervical lymphangioma, which diminished and disappeared spontaneously.

2. Discussion

Sternal cleft is a rare anomaly and corresponds to type 11 from the Willital classification of chest deformities [1]. Only sporadic cases have been described in the literature. The sternum is formed from 2 mesenchymal bars, independent from the ribs, which fuse between the 7th and 10th week of gestation. It starts cranially, at the manubrium and finishes distally at the xiphoid process. The sternal cleft may be partial or total, depending when the development process stopped. The defect could be superior, inferior or complete. The ossification of the sternum starts in the 5th month of embryonic life [2]. The superior cleft is often associated with other malformations like facial hemangioma or abdominal raphe [3,4]; the inferior cleft to ectopia cordis alone or as the pentalogy of Cantrell (ectopia cordis, intracardiac defects, sternal cleft, omphalocele, pericardial defect allowing communication with peritoneal cavity) [5]. The complete sternal cleft is the less frequent form. Rare association with other malformation has been met: gastroschisis [6], VACTERL syndrome [7], Dandy–Walker syndrome [8], pectus excavatum [9], and chest wall hamartoma [10]. In our case the sternal cleft was the only anomaly present.

Haque [11] noted a nutritional deficiency in the mothers of a few of his patients and stipulated that riboflavin could be incriminated for the appearance of skeletal defect. But most important, he diagnosed the malformation in two consanguineous families and in one the malformation reappeared suggesting that this is autosomal recessive transmitted. Gorlin et al. [8] found a cleft sternum in a pair of sisters, one of them having associated facial hemangioma and the other a tear-dropped umbilicus. Many aetiological factors had been incriminated but none of them was clearly pointed out. A high frequency of reported cases from the Middle East was noted and this should be taken into consideration for genetic counseling concerning transmission. Our patient was an isolated case, without any health problems in the family.

Some authors advocate a female predominance of the malformation. This is not proven due to the sporadic cases. Our patient is female but is also a unique case.

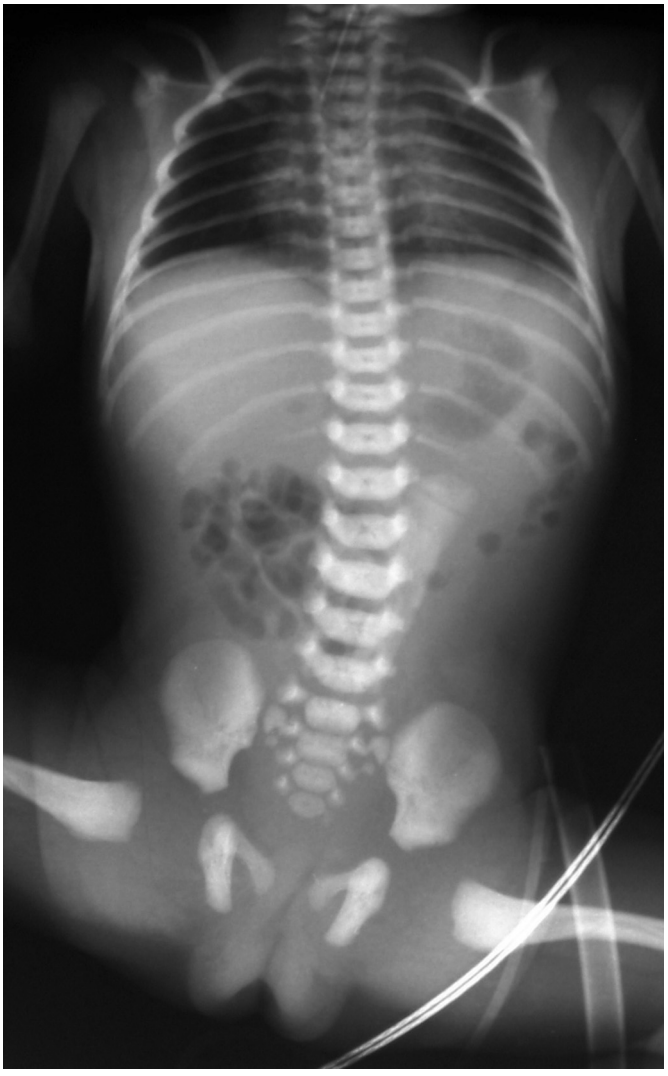

Fig. 2. X-ray: the big distance between the medial extremities of the clavicles.

Prenatal diagnosis was taken into consideration but its place is not very clear yet. Heron et al. [2] mention that their patient was diagnosed with sternal cleft by ultrasound at 21 weeks' gestation as well as Knox [12]. It seems to be more easily identified when it is associated with cardiac anomaly, because of the attentive research of other existing malformations.

The diagnosis can be easily done clinically at birth by inspection and palpation. The particular mechanics of the thoracic wall and the unusual movements of the heart under the skin are suggestive. The space between the costal ridges 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 cartilage [13]. The imaging identifies any associated anomaly besides the obvious ones like abdominal raphe, facial hemangioma and gives a detailed description of the position of clavicles, sternal bars and ribs.

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