Congenital Superior Sternal Cleft Isolated in a Newborn: Report of a Rare Case and a Literature Review

Yu Sha*, Yi Deng*, Minglin Ou*

* Yu Sha, Yi Deng, and Minglin Ou contribute equally

Corresponding Author: Minglin Ou, e-mail: minglinou@163.com, minglinou@glmc.edu.cn

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Patient: Female, newborn

Final Diagnosis: Congenital sternal cleft

Symptoms: Sternal cleft

Medication: —

Clinical Procedure: —

Specialty: Pharmacology and Pharmacy

Objective: Rare disease

Background: Sternal cleft is a greatly rare congenital thoracic deformity, arising from a failure of the sternal bars fusion process that should be completed in the fetal period, the incidence of which is less than 0.15%.

Case Report: Herein, we present a case report of a newborn girl having a superior congenital sternal cleft. After the baby was born, scar-like tissue was found in the middle of the chest and extended to the root of the umbilical cord. Based on the imaging data, this newborn was diagnosed with sternal cleft belonging to the superior sternal cleft and not associated with other congenital deformities.

Conclusions: As a rare congenital thoracic deformity, postpartum diagnosis of the sternal cleft mainly is currently based on medical imaging, including thoracic computed tomography (CT), three-dimensional (3D) reconstruction CT, and magnetic resonance imaging (MRI). Sternal cleft not only affects the aesthetic appearance but also leads to the destruction of the bone structure of the thorax, resulting in opposing thoracic movements. Therefore, early diagnosis and early treatment play significant roles in the treatment of this congenital sternal deformity. Regardless of whether there are clinical symptoms of sternal cleft, primary repair surgery must be done as soon as possible and during the neonatal period is best, in which simple surgical techniques achieve remarkable effects.

Keywords: Diagnosis • Infant, Newborn • Sternal cleft

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1 Central Laboratory, Guangxi Health Commission Key Laboratory of Glucose and Lipid Metabolism Disorders, The Second Affiliated Hospital of Guilin Medical University, Guilin, Guangxi, PR China
2 Department of Neonatology, The Second Affiliated Hospital of Guilin Medical University, Guilin, Guangxi, PR China

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Background

The sternal cleft, a greatly rare congenital malformation of the chest wall, was first reported in 1739 [1], with an incidence of less than 0.15% [2]. There is currently no evidence of a familial susceptibility to sternal cleft, and its pathogenesis is unclear, but it is more common in girls [3]. Therefore, there is a hypothesis indicating that sternal cleft may be related to Impairment in HoxB gene expression, based on murine models [3]. The sternal cleft arises from a failure of the sternal bars fusion process that should be completed by the tenth week of gestation under normal physiological conditions. This malformation is usually divided into 3 types: superior sternal cleft, total sternal cleft, and inferior cleft, according to the location and degree of the fissure. And the sternal cleft can be isolated or associated with other congenital deformities and the former is rare. Currently, postpartum diagnosis of sternal cleft mainly relies on medical imaging. Sternum cleft not only affects the aesthetic appearance but also leads to the destruction of the bone structure of the thorax, resulting in contradictory thoracic movements (depression during inspiration and bulge during expiration) and, in severe cases, may result in impaired gas exchange and difficulty breathing, impaired heart and lung function, chest infection, and even death [4-9]. Therefore, it is necessary for patients with sternal cleft to undergo surgical correction as soon as possible to restore the protective function of the thorax to protect the heart, lungs and other mediastinal organs. Here, we present a case report of a newborn girl having an isolated congenital sternal cleft, belonging to the superior sternal cleft. Taking this case as a starting point, we systematically discuss the formation mechanism of this deformity, possible causes, methods of diagnosis, the best time of corrective surgery, and surgical methods.

Case Report

We report the case of a newborn female diagnosed with congenital sternal cleft, which was part of the superior sternal cleft and not associated with other congenital deformities. After the baby was born, scar-like tissue was found in the middle of the chest and extended to the root of the umbilical cord, with a vesicle at the end of it near the head (Figure 1). The vesicle contained a colorless, transparent liquid, not infiltrated into the basal layer, presumably caused by extrusion during fetal development.

Based on the imaging data, we diagnosed a sternal cleft. Thoracic CT with no contrast agent injected was done, showing that the sternum was separated and the soft tissue in the middle of the chest was sunken inward (Figure 2). According to the results of the CT examination, we obtained specific data on the patient’s sternal fissure by measuring the fissures between the 4 unlosed sternal segments that together constitute the sternal body, at the same level of the sternal separation. It included the distance between the inner edge and

Figure 1. A newborn girl with an isolated congenital superior sternal cleft in the middle of the chest and extended to the root of the umbilical cord.

Figure 2. Thoracic CT with no contrast agent injected indicating the unlosed sternum (arrow).
the center of the sternal segment of the split sternal body. The specific data from top to bottom were: the distance between the inner edges of the first sternal segment was 12.8 mm and the distance between centers of the first sternal segment was 15.8 mm; the distance between the inner edges of the second sternal segment was 4.5 mm and the distance between centers of the second sternal segment was 6.8 mm. The distance between the inner edges of the third sternum was 2.6 mm and the distance between centers of the third sternum was 4.4 mm; the distance between the inner edges of the fourth sternum was 4.1 mm and the distance between centers of the fourth sternum was 6.4 mm (Figure 3). The extent of this newborn’s sternal cleft was shown by 3D CT reconstructions, and no rib or spinal anomalies were found (Figure 4). Thus, this case we reported as an isolated superior cleft sternum.

After the newborn was admitted to the hospital, she had a normal body temperature and strong sucking ability, and no vomiting or cough. In addition, breathing was regular with normal chest wall movement (a bulge during expiration and depression during inspiration) and the structure of the lungs was normal, confirmed by CT with no contrast agent injected.
Medical ultrasound examination of her urinary system and digestive system revealed no obvious abnormalities in the liver, spleen, bladder, and kidneys. Except for the unclosed foramen ovale (2.2 mm) in the heart during echocardiography, there were no other obvious abnormalities found. Flow cytometry showed absolute and relative counts of lymphocyte subsets and NK cells all belonging to immune cells, and no obvious pathological abnormalities. Regrettably, after the diagnosis of sternal cleft, the newborn’s guardian declined sternum repair surgery for reasons unknown to us.

**Discussion**

The sternum consists of 3 portions: the manubrium, the mesosternum (sternal body) made up of 4 segments, and the
metasternum (xiphoid process). In the embryonic stage, the left and right sternal plates are formed at the primordial sternal base. At the 9th to 10th weeks of gestation, the 2 sides of the chest plates fuse from top to bottom in the midline to form osteochondral. After birth, the cartilage has multiple ossification centers, ossified into several sternal segments, and finally fused into the sternum composed of 3 parts: the stem of the sternum, the body of the sternum, and the xiphoid process. If the midline sternal fusion blocks the process, a sternum cleft will form, which is a sternal deformity.

Clinically, according to the location and degree of the fissure, sternal clefts are usually divided into 3 types: superior sternal cleft, total sternal cleft, and inferior cleft. The sternal cleft can be isolated or associated with other congenital deformities. Specifically, the abnormalities, usually related to the superior sternum cleft, may be midline raphe from the tip of the cleft to the umbilicus, cerviocacal hemangiomas, and PHACES (posterior fossa malformations, facial hemangiomas, arterial anomalies with coarctation of aorta, cardiac defects, eye abnormalities, sternal cleft, and supraumbilical raphe) syndrome [10]. Total sternal cleft and inferior sterna cleft are usually related to serious developmental defects, especially cardiac ectopic and Cantrell pentalogy syndrome (substernal fissure or defect, anterior diaphragm defect, missing pericardial parietal layer, pericardial cavity communicating with an abdominal cavity; absent midline of supraumbilical abdominal wall with separate or continuous presence of umbilical bulge; multiple heart malformations) [11].

The diagnosis of the cleft sternum is mainly through medical imaging, which can determine the extent of the defect. Specifically, thoracic CT and DR are used to determine the extent of the lesion. Three-dimensional reconstruction CT is used to determine the location of the defect and whether the related ribs and spine are abnormal. MRI can detect whether there is cartilage development and the degree of development in the space of the sternal cleft. A contrast-enhanced CT, brain MRI, and echocardiography assist in determining whether the sternal cleft is accompanied by cardiovascular malformations and the degree of malformations. Prenatal diagnosis is essential for pregnant women. If a prenatal diagnosis is performed and a sternal cleft is found, then it is safest to have a cesarean section to prevent vaginal delivery from causing trauma to the external chest and abdominal structures.

However, at present, most of the current diagnosis of sternal clefts is postpartum diagnosis, and there are still relatively few methods available for screening during pregnancy or prenatal diagnosis with better results than postnatal diagnosis. Sternal clefts can be identified prenatally on mid-trimester ultrasonography [12]. However, ultrasonography imaging is limited by image resolution, and maternal obesity, unfavorable fetal position, and multiple pregnancy also affect the accuracy of ultrasonography [13]. Therefore, mild sternal cleft is not easily detected on ultrasonography. MRI is an effective means of prenatal screening for sternal fissures, but MRI is not yet used as a routine prenatal examination item due to the high cost of examination, so it is not widely used in prenatal diagnosis [14]. Despite the increased use of fetal MRI over the past decade, prenatal ultrasound is the most widely used tool in clinical practice. MRI is used for further evaluation only if the ultrasonography is abnormal [14,15]. In addition, X-ray, CT, and other imaging methods that produce radiation and lead to abnormal fetal development should not be used during the obstetric examination of pregnant women, although these methods are very effective in finding a sternal cleft. In addition, because this is a rare disease, physician inexperience is also an important reason for neglect of sternal cleft in prenatal diagnosis [13]. There are also no serological biomarkers for prenatal diagnosis of sternal clefts. Thus, a sternal cleft may not be diagnosed until after birth or even in adulthood. Incorporating MRI into prenatal diagnosis and increasing the awareness of prenatal doctors about the rare disease of sternal cleft is of great significance for prenatal detection of sternal cleft. Based on this, an appropriate delivery method can be selected, which can reduce the risk of a fetus who has sternal cleft in delivery. The prenatal diagnosis method for sternal cleft still needs further development and improvement.

Sternum cleft not only affects the aesthetic appearance, but also leads to destruction of thorax bone structure, resulting in opposing thoracic movements, and the mediastinum structure not effectively protected. Therefore, corrective surgery is necessary for patients with a cleft sternum to remove deformities, restore the bone-protected mediastinal structure, and set normal intrathoracic pressure [16]. Surgical correction of sternal cleft is the key treatment method, and it depends on the age of the patient. Regardless of whether there are clinical symptoms of sternal cleft, primary repair surgery must be done as soon as possible, ideally during the neonatal period because the sternum is more flexible then and the surgical techniques required are simple. Furthermore, the sternum fissure can be directly sutured in newborns within 1 month after birth. However, if corrective surgery is not performed during the neonatal period, as age increases, sternal cartilage ossification will cause sternal reconstruction to become complicated, and the requirements for surgical techniques are greatly increased, including sliding or rotating cartilage incision, sliding chondrotomy, partial or total thymectomy, clavicle dislocation, bone or cartilage graft insertion, and muscle flap insertion [17,18]. In older patients, the chest cavity is stiff and difficult to repair. Sliding chondrotomy and clavicle dislocation are effective and beneficial in elderly patients with chest wall stiffness because they increase chest wall compliance [19,20]. Partial or complete thymectomy is also sometimes required.
to reduce the risk of mediastinal compression during closure. If the sternal fissure is too wide or the risk of cardiovascular compression is too high, it can be closed with a prosthesis or bone graft [17,21]. Corrective surgery to reconstruct the thoracic structure must avoid compression of the heart and blood vessels. Consequently, timely diagnosis and corrective surgery can repair the thoracic structure and restore its protective function, achieving aesthetic effects.

Conclusions

Sternal cleft is a rare congenital disease that affects esthetics and can seriously endanger the patient’s life. Timely corrective surgery is very important for patients with congenital sternal cleft, preferably during the neonatal period. At present, the postpartum diagnosis of congenital sternal cleft mainly relies on imaging examinations including thoracic CT, 3D reconstruction CT, and MRI, but CT is usually not used for prenatal diagnosis because the radiation generated can affect normal fetal development. Ultrasound is currently the main prenatal diagnosis method for sternal clefs, but due to the limited resolution of ultrasound images, mild sternal clefs are not easily found. Although MRI is an effective prenatal method for sternal clefs, it cannot be used as a routine method for prenatal diagnosis due to its high cost and can only be used after the abnormality is detected by ultrasound. Thus, there is an urgent need for appropriate prenatal diagnostic methods that can detect mild microsternal clefs.

Ethical Approval

The ethics committee of the Second Affiliated Hospital of Guilin Medical College has approved the study.

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