Complete Sternal Cleft in a Filipino Newborn

Barbra Charina V. Cavan[1], Jonna A. Masongsong[1], Peter Y. Mancao[2], Arnold A. Tan[2]

[1]*Department of Pediatrics, Cebu Doctors' University Hospital, Osmena Blvd., Cebu City, Philippines;*
[2]*Department of Surgery, Cebu Doctors' University Hospital, Osmena Blvd., Cebu City, Philippines*

ABSTRACT

We report the case of a female newborn with complete sternal cleft and patent ductus arteriosus. No other dysmorphic features were noted. Repair was accomplished at 18 days of age and the respiratory and cardiac conditions remained stable.

Key Words: sternal cleft, cleft sternum, chest wall deformity, congenital malformation or anomaly, surgery

Introduction

Sternal clefts are rare occurrences. Embryologically, the sternum is formed by the fusion of the paired parallel sternal bars between the 8th and 10th week of pregnancy.[1,2] Incomplete fusion or failure of the fusion of the sternal bars leads to the development of a congenital sternal cleft.[3] If fusion is partial, this is referred to as an incomplete sternal cleft, designated as superior or inferior depending on the location of the cleft. The superior cleft is the most common.[1] If the whole sternum failed to fuse, this condition is called a complete sternal cleft.[4]

Hazari et al conducted a review of the of sternal clefts from 1800 to 1998 and noted 73 cases, 40% of which had associated facial hemangiomas.[1] Sarper et al in 2002 cited a 1990 study of Shamberger and Welch which documented only 23 cases of complete sternal clefts specifically.[5] Shalak et al, also in 2002, reported one patient with complete sternal cleft with a congenital heart disease and reviewed 35 previously reported cases, including those mentioned by Hazari.[4]

Common malformations reported to be associated with sternal clefts are ectopia cordis, vascular malformations/ dysplasia such as hemangiomas, supraumbilical raphe, and even abdominal abnormalities.[1,3] Cantrell reported a syndrome also known as Cantrell's pentalogy where there is a combination of an omphalocele, diaphragmatic hernia, sternal cleft, pericardial defect, and cardiac abnormalities.[6]

Surgical techniques used to repair sternal clefts range from primary closure to grafts to the use of metal plates.[1,7,9]

Corresponding author: Barbra Charina V. Cavan, MD
Department of Pediatrics,
Cebu Doctors' University Hospital,
Osmena Blvd., Cebu City, Philippines
Telephone: (+63)(32)414-1108
E-mail: cavanbarbie@yahoo.com

We describe below the first reported case in Cebu of a Filipino female newborn with complete congenital sternal cleft and patent ductus arteriosus.

Case Report

The patient was a female born term at 38 weeks age of gestation, by primary caesarian section (secondary to variable deceleration) to non-consanguineous Filipino parents. The pregnancy course was unremarkable except for thin meconium staining during labor. Her birth weight was 2.693 kg and she had feeble cry and marked chest indrawing.

On closer examination, there was a marked abnormality on the midline chest characterized by thin, transparent skin and an absent sternum. The apex beat was evident through the skin and noted to be at the 5th intercostal space. The patient's umbilicus was at the level of the 12th rib. A thin ligament connected the base of the umbilicus to the thin skin (Figure 1). There were no cutaneous hemangiomas, no dysmorphic features, and no median raphe on the sternal area. The thin skin became less transparent and developed central atrophic scar later on. Chest radiographs showed no other abnormalities of the rib cage. Two-dimensional echocardiography revealed a small patent ductus arteriosus (PDA), a patent foramen ovale, and an intact pericardium. Abdominal ultrasound showed a laterally placed esophagus.

At 7 hours old, the baby had an episode of generalized tonic-clonic seizures. Metabolic investigations (blood sugar, arterial blood gases, and electrolytes) were normal. A cranial ultrasound revealed mild cerebral edema. Recurrence of the convulsions prompted Phenobarbital treatment which provided good control in the succeeding days. Hypoxemic ischemic encephalopathy (HIE) stage 1 due to intrapartum asphyxia was considered as the cause of the seizures.

Primary repair of the complete sternal cleft was done on the *18th* day of life. The surgical technique involved the trimming of the parallel unfused sternal bars and closure of the sternum with wires. The vertical wound was closed subcutaneously. Chest tube thoracostomy was also performed. The patient was discharged on the 27th hospital day improved.

On subsequent clinic visits, the patient was noted to have normal milestones, normal repeat 2-D echo at the age of 3

years, but her growth curve has been following the 5th centile curve (FNRI-PPS, 1992). Phenobarbital was discontinued 2 weeks after discharge since the patient remained seizure free.

The family was counseled regarding the sporadic occurrence of these clefts making the recurrence risk in subsequent pregnancies negligible.

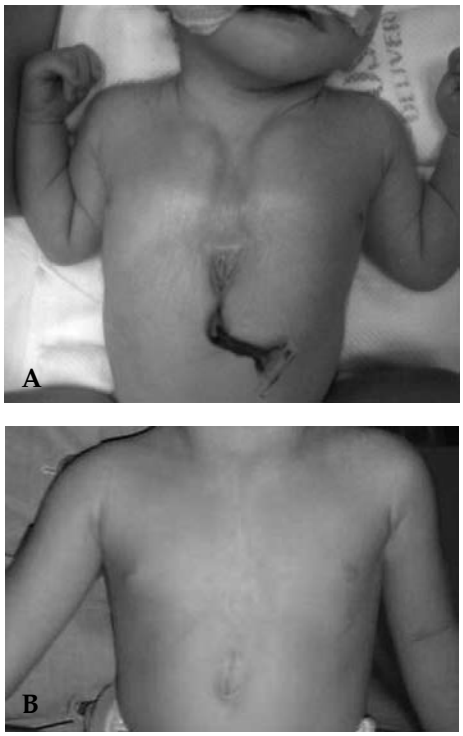

Figure 1. A) At 1 day old - Thin skin over the sternal area with displacement of the umbilicus superiorly and a ligament from the sternal area to the umbilical stump. B) At 1 year of age.

Discussion

This is the first case of complete sternal cleft reported in the Philippines after a careful literature review.

The presence of the sternal cleft prompted a careful search for other malformations in this reported patient. Ectopia cordis, where the heart is displaced outside the chest wall, is one common malformation associated with sternal defects.[5] Other reported associated malformations include congenital heart defects, vascular abnormalities, hemangiomas, and skeletal defects.[4,12] Reported syndromes known to have sternal anomalies include Posterior fossa malformations, Haemangiomas, Arterial anomalies, Coarctation of the aorta and cardiac defects, Eye anomalies, and Sternal clefts or supraumbilical abdominal raphe (PHACES) syndrome, Cantrell's pentalogy, Goltz syndrome, and sternal malformation-vascular dysplasia syndrome.[10,12] The index case did not fully meet the classical criteria of these enumerated syndromes. She only had a small PDA. The PDA is not reported as a common major finding associated with sternal clefts, having been mentioned in only a few reports.[4,9] Unfortunately, angiography that would exclude other vascular malformations was not done on the patient.

There are many hypotheses on the formation of sternal clefts. They include the absence of the pre-sternum or secondary splitting and intrauterine rupture of the chorion or yolk sac.[1,7] Early yolk sac rupture might affect the descent of the heart, heart development and failure of fusion of the sternum, while late rupture may just affect the sternum alone. Sternal clefts associated with vascular dysplasia may be due to persistent midline angioblastic tissue.

The absence of a complete sternum evidently leads to an unstable chest wall. Most surgeons prefer a surgical repair in the neonatal period, with primary direct closure.[1,7,9] Repair during this period is favorable because of the compliant chest wall. Other advantages are protection of the vital structures within the thoracic cavity and correction of the visual deformity of the chest. The timing of the repair has to be considered in accordance with the stability of the patient's condition. Fortunately for this patient, there was no major cardio-respiratory compromise that would have complicated the contemplated procedure. It was likewise important to make sure that after the surgical approximation, the cardio-respiratory status remained stable.

The lateral displacement of the esophagus noted on the patient's ultrasound may be a normal variant. Such variants were studied by Smith et al using computed tomography with the cricoid as the reference point.[8] There is no mention of a similar displacement of the esophagus in other case reports of complete sternal clefts. Her seizures were considered to be due to hypoxic ischemic encephalopathy (HIE) stage 1 secondary to antepartum and intrapartum asphyxia, as evidenced by the thinly meconium stained amniotic fluid. The neurologic sequelae of perinatal asphyxia include seizures, hypotonia and coma[13] although this stage of HIE usually has a good prognosis.

Complete sternal cleft is reported as a sporadic event, whether associated with other anomalies or isolated.[1,2] Counseling on recurrence risks being low in such cases was done with the couple, especially as this patient was their first child.

Conclusion

Complete sternal clefts are rare types of birth defects. Repair of this malformation during the neonatal period, especially in the absence of other major congenital abnormalities leads to a good outcome as seen in this case.

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References

1. Hazari A, Mercer N, Pawade A, Hayes A. Superior sternal cleft: construction with a titanium plate. Plast Reconstr Surg. 1998; 101: 167-170.
2. Jones KL. Smith's Recognizable Patterns of Human Malformation, 5th ed. Pennsylvania: W.B.Saunders Company, 1997.
3. Alphonso N, Venugopal PS, Deshpande R, Anderson D. Complete thoracic ectopia cordis. Eur J Cardiothorac Surg. 2003; 23: 426-428.
4. Shalak L, Kaddoura I, Obeid M, Hashem H, Haidar R, Bitar F. Complete cleft sternum and congenital heart disease: Review of Literature. Pediatr Int. 2002; 44(3): 314-216.
5. Sarper A, Oz N, Arslan G, Demircan A. Complete congenital sternal cleft associated with pectus excavatum. Tex Heart Inst J. 2002; 29: 206-209.
6. Sawin RS. Pediatric Chest Lesions. Ped Clinics N Amer. 1998; 45(4): 861-873.
7. Yavuzer S, Kara M. Primary repair of a sternal cleft in an infant with autogenous tissues. Interact Cardiovasc Thorac Surg. 2003; 2: 541-543.
8. Smith KJ, Ladak S, Choi PTL, Dobranowski J. The cricoid cartilage and the esophagus are not aligned in close to half of adult patients. Can J Anesth. 2002; 49(5): 503-507.
9. Singal A, Balamourougane P, Bhatnagar V. Primary closure of thoraco-abdominal ectopia cordis. J Indian Assoc Pediatr Surg. Oct-Dec 2006; 11:4.
10. Loguercio Leite JC, Faermann R, Rodrigues Stein N, Zandona D, Maegawa G. Focal dermal hypoplasia assodiated with split sternum – Goltz syndrome. Clin Dysmorphol. 2005; 14(1): 37-39.
11. Khoury J, Jerushalmi J, Said R, Loberant N. U shaped sternum: a rare malformation demonstrated on bone scintigraphy in an adult patient. Clin Nucl Med. 2003; 28(3): 224-225.
12. Vermeer S, van Oostrom C, Boetes C, Verrips A, Knoers N. A unique case of PHACES syndrome confirming the assumption that PHACES syndrome and the sternal malformation-vascular dysplasia association are part of the same spectrum of malformations. Clin Dysmorphol. 2005; 14(4): 204-206.
13. Section of Neonatology of the Philippine Pediatric Society, Inc. Standards of newborn care, 2nd ed. Manila: PPS, 2001.