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Case Report

Peri-Operative Management of a Neonate Undergoing Sternal Cleft Repair: Case Report

Archana S Ramesh MD, Jacob P Bray MD*, Khoa N Nguyen MD

Department of Anesthesiology, UPMC Children's Hospital, Pittsburgh, USA

***Corresponding author:** Dr. Jacob P Bray, Department of Anesthesiology UPMC Children's Hospital 4401 Penn Ave. Pittsburgh, PA 15224 USA. Tel: 412-692-5939; Email: brayjp@upmc.edu

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Abstract

Sternal clefts are a rare congenital malformation where there is a partial or complete defect in the sternum. The condition leaves the vital organs in the mediastinum exposed for trauma. Surgical repair is required and should be completed soon after the diagnosis is made. We present the peri-operative management of a neonatal that was transferred to our hospital for management of a partial sternal cleft.

Introduction

The term sternal cleft describes a congenital condition ranging from an incomplete or partial sternal cleft to complete sternal agenesis. Though sternal clefts are described as the most common sternal malformations, it is still quite rare with an estimated incidence of 1 in 50,000 to 100,000 live births [1]. Complete sternal clefts are much more rare than partial clefts. Sternal clefts and sternal agenesis places vital organs in the thoracic cavity at risk for external trauma. Sternal clefts are usually not associated with major cardiac anomalies and patients are usually not in distress, while patients with sternal agenesis can present in respiratory distress upon presentation. Patients diagnosed with sternal malformations should undergo testing to determine if they have further congenital defects. Some patients may have other defects associated with PHACES syndrome or Cantrell's Pentalogy [2]. Prior to publication written consent for this case report was obtained from the patient's parents.

Case Report

Our patient was a 6-day old term female weighing 3.27 kg. She was born at a gestational age of 40 weeks and 5 days by spontaneous vaginal delivery with a birth weight of 3.47 kg. Maternal medical history was significant for gestational diabetes. The pre-natal course was otherwise unremarkable. The patient had Apgar scores of 9 and 10 at 1 and 5 minutes, respectively. She was not in any respiratory distress but was found to have some mild sternal retractions. Post-delivery she was noted to have a hypopigmented lesion over her mid-chest indicative of a possible sternal malformation. (Figure 1) The diagnosis of a sternal cleft was then confirmed with computed tomography (CT) of the chest.

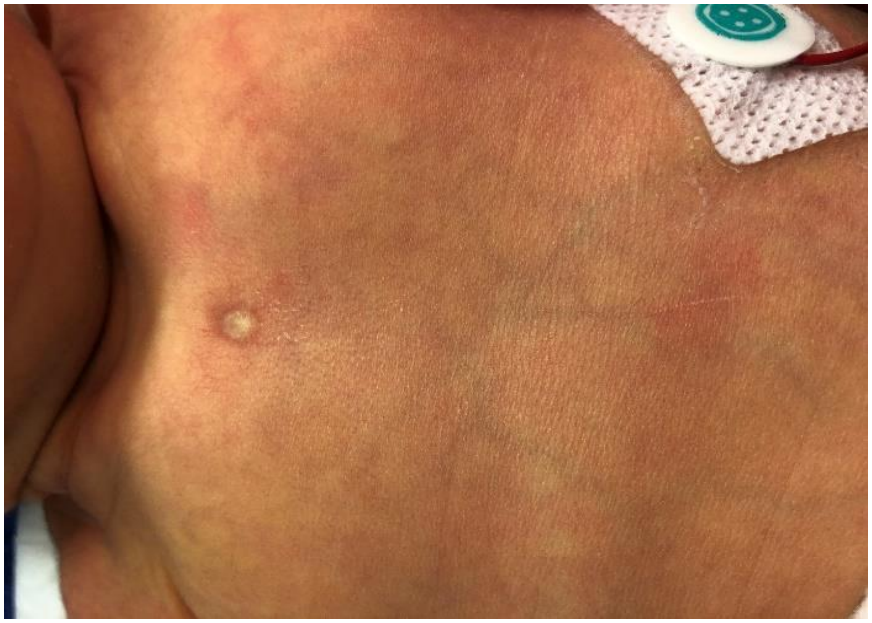

Figure 1: Hypopigmented lesion and superior aspect of sternum.

(Figure 2) The CT impression reported "congenital absence of the majority of the ossified sternum with transverse width of the sternal cleft measuring up to 16.3mm." No definite evidence of visceral herniation through the sternal cleft defect was noted. She underwent additional evaluations for other congenital defects which were negative. Ultrasound scans of her head, spine, and abdomen were unremarkable. Transthoracic echocardiogram revealed a patent foramen ovale and otherwise normal anatomy. Her admission bloodwork was also unremarkable. She was transferred to our facility for management of the sternal cleft and was admitted to the neonatal intensive care unit.

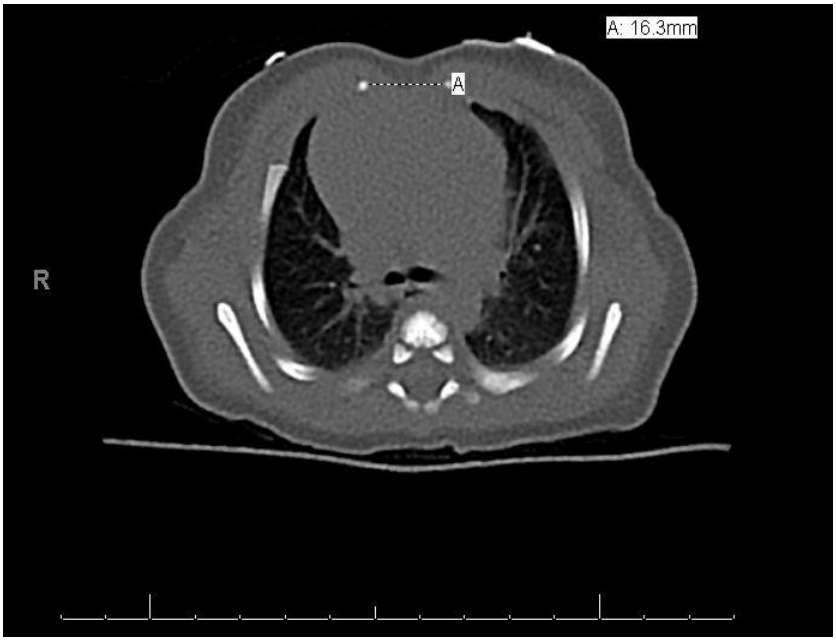

Figure 2: CT of chest showing 16.3mm sternal cleft.

On the morning of surgery, she was hemodynamically stable and spontaneously breathing room air with no signs or symptoms of respiratory distress. Inhalational induction of anesthesia was initiated with sevoflurane and oxygen after standard ASA monitors were placed. After induction, a peripheral intravenous line (PIV) was placed in the left arm and rocuronium was administered to facilitate tracheal intubation. A 3.0 cuffed endotracheal tube was placed without issue. The ventilator was set to pressure control mode and tidal volumes of 7-8 cc/kg were achieved with a

peak inspiratory pressure of 17 cm H2O. An additional 24-gauge PIV was placed in the saphenous vein, and a 24 gauge right radial arterial line was placed for close hemodynamic monitoring and potential need for frequent laboratory checks. Anesthesia was maintained with sevoflurane in oxygen and air mixture. Intermittent fentanyl was used for analgesia.

The operative findings included a partial sternal cleft with a hypopigmented area over the lesion. The cleft was found on the superior aspect of the sternum and represented about 60% of the sternum in the shape of a V. Using a median sternotomy incision, the skin lesion was excised and extended down to the subcutaneous tissue. The edges of the sternal cleft were identified and carefully dissected down to the inferior portion of the sternum where it was complete. The right lobe of the thymus was excised to provide more room for sternal closure. The edges of the sternal cleft were reinforced using horizonal sternal wires. The edges of the sternum were then approximated with suture and stainless-steel wires. During sternal approximation we closely monitored the patient's ventilation and hemodynamic parameters. She tolerated sternal closure well without a significant change in the required peak inspiratory pressure or mean arterial pressure. At the end of the surgery the trachea was extubated in the operating room and supplemental oxygen was provided via facemask for goal O2 > 90%. The patient was transferred to the cardiac intensive care unit for close monitoring. Post-operative analgesic regimen included one dose of intravenous morphine and oral acetaminophen. There were no cardiopulmonary complications post-operatively. She was transitioned to room air and transferred out of the intensive care unit to the post-surgical unit on POD1. She was discharged to home on POD 2. She was noted to be doing well during a three month follow up clinic visit.

Discussion

Sternal malformations make up a broad spectrum of deformities ranging from a simple sternal cleft to complete sternal agenesis. Shamberger and Welsh classified them as cervical ectopia cordis (incompatible with life), thoracic ectopia cordis, thoracoabdominal ectopia cordis, and sternal clefts based on their anatomic differences although there may be considerable overlap in their clinical features and presentation [3].

Sternal cleft as in our patient, though said to be the most common subtype, is still a rare diagnosis. A previous report of over 5000 patients found that only 0.15% of all anterior chest wall deformities were due to sternal clefts [4]. Sternal clefts can be partial or complete. Partial sternal clefts may be superiorly or inferiorly located. Superior sternal clefts are usually isolated. Rarely it may be associated with PHACES syndrome which is a neurocutaneous syndrome that includes- Posterior fossa malformations of the central nervous system; Hemangiomas, Arterial lesions, Cardiac anomalies; Eye abnormalities and Sternal cleft. The inferior sternal clefts on the other hand may be associated with Cantrell's Pentalogy (defect of the anterior abdominal wall, diaphragm, pericardium, lower sternum and heart).

Patients with sternal clefts usually present during infancy. Rare cases of asymptomatic sternal clefts repaired well into adulthood have been reported. It is best to repair sternal clefts early in life when the thoracic cavity is still pliable. Chest wall rigidity and growth of thoracic organs may hinder primary closure in the older patients necessitating the need for prosthetic or tissue interposition. Although most patients with sternal clefts may have an isolated defect, other associated lesions may be subtle and should be ruled out before proceeding with surgery. A detailed history, comprehensive physical exam and targeted imaging studies should be performed to evaluate for possible co-existing defects as these may not only have significant anesthetic implications but also aid in planning the appropriate surgical procedure. Following a thorough assessment, pre-operative anxiolysis may be given in the older patients if needed prior to transporting to the operative room. General anesthesia can be safely induced by inhalational or intravenous technique. Adequate and reliable intravenous access should be established especially since the volume of allowable blood loss in young infants is limited. Invasive blood pressure monitoring may be considered given the concern for compression of the great vessels with sternal closure. Close monitoring of airway pressures is also highly essential if primary closure is intended. It may be prudent to minimize opioid administration with the use of non-opioid analgesics including acetaminophen and local anesthetic infiltration by the surgeon. Successful use of neuraxial analgesia has been reported in older patients. These children are especially at risk for hemodynamic and respiratory complications post-operatively since there exists a concern for compromised cardiopulmonary development due to the absence of structural scaffolding in-utero. Hence close monitoring in the intensive care unit for 24 hours is recommended.

In summary, our patient was fortunate to have an isolated partial sternal cleft. She was repaired within the first week of life, tolerated the procedure well, and was noted to be thriving at her three month follow up appointment. This case highlights the need to closely inspect benign appearing hypopigmented areas on the sternum of otherwise health appearing neonates as they may be associated with an occult sternal cleft. Despite how well our patient did peri-operatively, hemodynamic and physiologic changes which occur during sternal closure can precipitate cardiorespiratory instability and the anesthesia team should be prepared to act accordingly.

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