



Congenital left hemidiaphragmatic agenesis : a case report

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ABSTRAK

Congenital diaphragmatic hernia is a congenital diaphragmatic disorder characterized by herniation of the abdominal viscera into the thoracic cavity. In rare diaphragmatic developmental abnormality cases, there is complete diaphragmatic agenesis. We report a case of left hemidiaphragmatic agenesis with usual postnatal course. A 13-day old female infant presented respiratory distress for a week. Chest radiograph showed left sided congenital diaphragmatic hernia. We report a left hemidiaphragmatic agenesis case. A 13-day old female infant presented respiratory distress for a week. Chest radiograph showed a left-sided congenital diaphragmatic hernia. The patient undergo surgical exploration and was found to have an unusual and large defect in the left hemidiaphragm. No diaphragm is visible on the anterior and lateral sides, and the anterior sides presented only a small edge of the diaphragm. Agenesis of the left hemidiaphragm defect was identified and well managed by suturing the posterior edge of the diaphragm to the intercostal muscles. This report describes the well-management of hemidiaphragmatic agenesis without incorporating a prosthetic material.

Keywords: *Congenital diaphragmatic hernia, diaphragmatic agenesis, repair*

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a congenital abnormality of the diaphragm, characterized by diaphragm defect accompanied by herniation of the gastrointestinal and other abdominal viscera into the thoracic cavity (Litwinska et al., 2015). There is also complete diaphragmatic agenesis in rare diaphragmatic developmental abnormality cases, which usually involves one side, but sometimes both sides (Mirza et al., 2012). However, it is still a management dilemma to fix these defects. The use of prosthetic materials, abdominal or intercostal muscle flaps, etc., are various techniques that have been tried and proposed to correct this defect. We report a rare case of congenital hemidiaphragmatic agenesis (CHDA), and successful repair without prosthetic materials.

CASE REPORT

A 13-day old female infant presented respiratory distress for a week and was transferred from type C to type B hospital. The child was born by vaginal delivery with respiratory distress at the primary care center.



Figure 1. Operative view of the left hemidiaphragmatic agenesis. No diaphragm is seen on the lateral and anterior sides, whereas a small diaphragm edge can be sutured anteriorly by stay suture to the intercostal muscles.

Oxygen therapy is given through the cannula. Ophthalmic prophylaxis was performed, and then she was referred to a type C hospital, where a nasogastric tube was applied, and antibiotic treatment with meropenem was initiated. KAEN 4B was administered with a metabolic flow of 16 drops per minute. The child was referred to a type B hospital to be managed by General Surgery.

The baby's weight was 2.6 kg during the admission, and a general physical examination revealed tachypnea (respiratory rate >50 min). Pulse was 143/min, and blood pressure was normal. There was an overt sign of respiratory distress on respiratory system examination, such as tachypnea, nasal flaring, intercostal and subcostal retractions. On auscultation, there were bowel sounds in the left chest. Air entry was standard on right chest auscultation. The abdomen was soft and not much scaphoid. A chest radiograph was requested that revealed intestinal gas shadows in the left hemithorax. A diagnosis of left-sided CDH was made and was prepared for elective surgical intervention.

Her preoperative laboratory parameter was average. Treatment was started on parenteral antibiotics, and on the fifth day later, the patient will be planned for an elective operation. Parents were counseled about the surgical procedure and its postoperative complications. The operation was started with the left subcostal incision. Most of the small and large intestines and left liver were found herniating into the left hemithorax. The content was reduced back into the abdominal cavity. The diaphragm was absent on the left hemithorax's lateral and anterior sides, and only a small edge of the diaphragm was presented on the posterior sides [Figure 1].

The lung looked hypoplastic. No hernia sac was found during the operation. The part of the diaphragm present on the posterior aspect was mobilized and sutured to the intercostal muscles using interrupted stitches of PGA 2.0 sutures. The abdomen wound was closed by only skin closure.

The postoperative recovery was good. On the fifth postoperative day, the nasogastric tube was removed. The next day was given oral nutrition, and the patient was discharged on the tenth postoperative day.

METHOD

We report a case of left hemidiaphragmatic agenesis with usual postnatal course. A 13-day old female infant presented respiratory distress for a week. Chest radiograph showed left sided congenital diaphragmatic hernia. We report a left hemidiaphragmatic agenesis case. A 13-day old female infant presented respiratory distress for a week. Chest radiograph showed a left-sided congenital diaphragmatic hernia.

DISCUSSION

Classical CDH named by Bochdalek, the first to identify the defect in 1848, is characterized by a posterolateral defect in the diaphragm (Mirza et al., 2012). The left-sided Bochdalek's CDH is more common and reported at approximately 80%. Left-sided CDH has Better Prognosis as compared to right-sided CDH.(Stolar, 1998)

In the rare maldevelopment of the diaphragm cases, there is also complete diaphragmatic agenesis, which usually involves one side, but sometimes both sides (Litwinska et al., 2015). Of all CDH, only 6% is reported with CHDA, and the left side is more common than the right side (Toran & Emery, 1981). In our case, the abnormality was on the left side, showing the common side of the lesion. Some bilateral CHDA cases have been reported with incredibly high mortality, and cases such as this cannot be treated. The cause of CHDA is based primarily on speculation and assumptions and was not well defined to date (Toran & Emery, 1981). A failure of pleuroperitoneal membrane development to close the pleuroperitoneal canal on the involved side is considered the probable etiology. Various drugs and environmental factors have been proposed to play a crucial role in CDH developmental.(Glavaš et al., 2000)

CDH patients' survival is determined by the severity of pulmonary hypertension and pulmonary hypoplasia in the first 24 hours. The more severe pulmonary hypertension and lung hypoplasia, the early the presentation will be, and the worse will be the outcome and vice versa (Stolar, 1998). The clinical features are usually vomiting, respiratory distress, and cyanosis. In our case, the patient could survive in the first 24 hours. That means the severity grade of pulmonary hypertension and pulmonary hypoplasia is not high.(Glavaš et al., 2000)

To delineate the diaphragm and its abnormalities, CT-Scan and MRI are necessary (Toran & Emery, 1981). In our case, the diaphragmatic abnormality was clearly visible on the chest X-ray. (Sung et al., 2009)

Absent diaphragmatic tissue makes the surgical procedure very difficult. Although diaphragmatic tissue is present, it is still difficult to be repaired (Stolar, 1998). Some of the treatment options are using a prosthesis, abdominal wall, and chest wall muscle flaps, using the pre-renal fascia, suturing the chest margins with the liver, and using the pre-renal fascia (Sung et al., 2009). However, some surgeons even suggest no repair (Glavaš et al., 2000). Approximately 30-50% of CHD patients showed prosthesis-related complications and even hernia recurrence when treated with a mesh union in the defect. (Sheehan et al., 2000)

In contrast to our case, we have a small diaphragm edge. We tried to make it enough to close with the mobilization of the retroperitoneum and suturing it with the intercostal muscles. Although there is a possibility of an abdominal compartment after the liver and most of the small and large intestines return to the abdominal cavity. In our patient, skin closure went well.

CONCLUSION

It can be concluded that left diaphragmatic agenesis is a rare developmental abnormality of the diaphragm. Surgical intervention as early as possible proves to be beneficial for the patient. The patient can be managed surgically without incorporating a prosthetic material by mobilizing and suturing the remnants of the diaphragm with the intercostal muscles.

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