

A CASE OF CONGENITAL DIAPHRAGMATIC HERNIA WITH EVENTRATION OF DIAPHRAGM WITH MIDLINE SHIFT AND HYPOXIA

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ABSTRACT

Background: Eventration is a rare presentation seen in 1 in 10,000 live births. This is usually seen as secondary to defective diaphragm formation with weak musculature or defective nerve innervations.

Case Presentation: An 8 day old baby boy with inability to maintain saturation and failure to thrive and chest X ray was suggestive of congenital diaphragmatic hernia. Intra operatively there was evidence of thinned out diaphragm with herniation of small bowel, large bowel, stomach and spleen with midline shift. Evidence of completely collapsed left lung was present. Repair of anomaly done and baby discharged home on breast feed with maintaining oxygen saturation at room air. **Conclusion:** Timely intervention in eventration of diaphragm can affect the outcome of baby.

KEYWORDS : Eventration of diaphragm, CDH, Herniation, Midline shift, Collapsed lung

INTRODUCTION

Eventration of diaphragm is a condition in which there is permanent elevation of hemidiaphragm. It can be due to congenital or acquired causes. Congenital eventration is the result of defect in the migration of myoblasts to the septum transversum during development of diaphragm causing partial or total replacement of the diaphragm muscle with fibroelastic tissue. This creates a thin and weakened hemidiaphragm resulting in a cephalic displacement of the affected side. Symptoms of eventration ranges from asymptomatic state to severe respiratory distress and death if untreated.

Case Presentation

An 8 day old baby boy in delivered in peripheral hospital, referred to tertiary care centre with complaints of inability to maintain saturation and failure to thrive. Baby was delivered via full term normal vaginal delivery with normal antenatal history with birth weight 2.9 kg by 24 year old mother. The mother was screened and found negative for HIV and hepatitis panel and denied any use of alcohol, tobacco, and illicit substances throughout the course of her pregnancy. Baby cried immediately after birth. Baby was admitted in Neonatal ICU and evaluated.

Patient started on symptomatic treatment and evaluated. Surgery opinion was taken in view of chest X ray finding.

Thorough evaluation done by surgeon revealed the baby was tachypneic with respiratory rate of 60 cycles per minute, Spo₂ in room air was 75 % and with non invasive ventilation Spo₂, it was 86 %. On auscultation of chest air entry was reduced on left side of chest with sluggish bowel sounds were audible at the lower lung fields. The abdomen of the baby was scaphoid.

Investigations

Chest X ray was done and it was showing presents of bowel contents within left hemithorax and collapsed left lung. Ultrasonography of abdomen and thorax showed herniation of bowel contents into the left hemithorax.

Diagnosis of congenital diaphragmatic hernia was made decision of emergency exploration and repair of anomaly was taken.

Operative Procedure

Horizontal incision taken over the left subcostal region taken and peritoneal cavity opened. Intra operatively there was no

evidence of defect in the diaphragm instead there was evidence of thinned out diaphragm with herniation of small bowel, large bowel, stomach and spleen with midline shift. Also there was evidence of completely collapsed left lung was present.

Reduction of contents small bowel, large bowel stomach and spleen done. Thinned out eventrated sac excised and repair of congenital diaphragmatic hernia done by taking mattress suture and placcation of thinned out diaphragm. Approximation of right and left crus of diaphragm done. Intercostal drainage tube was put intraoperatively.

Post Operative Care

Post operatively baby was shifted to NICU and was on ventilator support for 2 days. Baby was extubated and was maintaining Spo₂ of 98% with Non Invasive Ventilation with improved air entry over left side. Baby was started on breast feeding from post operative day 3 and baby was tolerating the feed. ICD was removed on post operative day 5 and baby was maintaining saturation with minimal oxygen support. Baby was discharged home on post operative day 9.



Figure 1: Preoperative Chest X Ray Showing Collapsed Left Lung With Herniation Of Bowel Contents To Left Hemithorax With Midline Shift

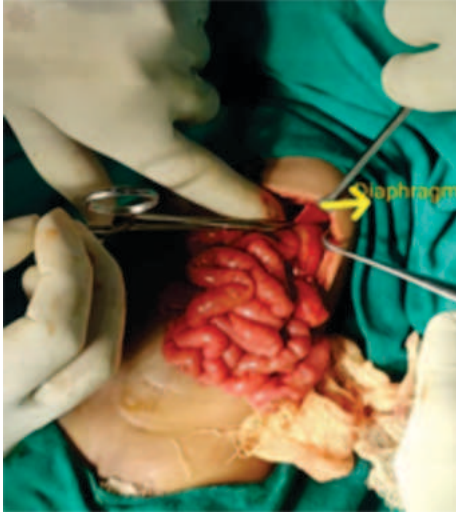


Figure 2: Thinned Out Diaphragm



Figure 3: Empty Left Hemithorax After Reduction Of Hernia Contents



Figure 4: Post Operative Chest X Ray Showing Reduction Of Bowel Contents And Inflation Of Collapsed Left Lung

DISCUSSION

Congenital eventration is due to defect in migration of

myoblasts to the septum transversum causing partial or total replacement of the diaphragm muscle with fibroelastic tissue because of neurogenic muscular aplasia. This creates a thin and weakened hemidiaphragm resulting in a cephalic displacement of the affected side. Eventration can also occur in association with other congenital disorders and infections. Mitochondrial respiratory chain disorder has been linked to diaphragmatic dysfunction in the neonatal period. Infectious associations include fetal rubella and cytomegalovirus infections. [1][2][3]

Eventration can anatomically divide in to complete, partial, or bilateral. Embryological congenital eventration can be classified in to anterior, posterolateral, and medial. Reported incidence is less than 0.05% and most commonly affecting male gender and on left side. Majority of cases are asymptomatic and diagnosed as incidental finding. If the child is symptomatic, they may present with respiratory symptoms. Most of time respiratory complaints are due to midline shift of mediastinal structures. Gastrointestinal involvement is common and may manifest as vomiting, bloating, constipation, poor weight gain, and poor oral intake. [4][5]

Symptomatic and supportive care is needed for asymptomatic and mild cases. If baby is not able to maintain saturation oxygen support supplementation with nasal prongs can be given. Still continuous positive airway pressure (nasal CPAP). Pulmonary rehabilitation and physical therapy are part of treatment. As such patients are having GI complaints nutritional support also to be taken care of. [5][6]

Surgical management should be considered in babies who fail to respond to conservative management. Surgery of choice is Diaphragmatic plication. Thinned out diaphragm is plicated and flattened with U stitches and anchored down. This procedure is to lower down the hemidiaphragm allowing lung to expand by increasing intrathoracic volume. Open thoracotomy, video-assisted thoracoscopic surgery (VATS), laparoscopic, or robotic-assisted surgery are the approaches available for diaphragmatic plication. An intercostal drainage tube should be put for pleural drainage and can be removed once output is less than 200 ml/day. [7][8][9]

CONCLUSION

Congenital diaphragmatic eventration (DE) is a rare condition that can lead to death if left untreated. DE's are difficult to diagnose as they can present without symptoms thus requiring intricate management. Infants dealing with DE are at an increased risk of morbidity as their thoracic cage is underdeveloped leading to life-threatening complications including failure to thrive. This case demonstrates the successful outcome of the patient due to accurate intervention at accurate timing of congenital DE.

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