Unusual presentation of congenital diaphragmatic hernia - Anaesthetic consideration.

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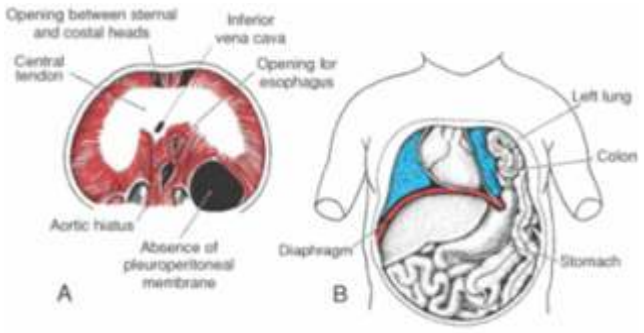
Abstract :

Congenital diaphragmatic hernia (CDH) is herniation of bowels and abdominal solid organs in thoracic cavity, causing pulmonary hypoplasia with decreased pulmonary vasculature and dysfunction of the surfactant system. In severe cases, left ventricular hypoplasia is also observed. Delayed herniation of the abdominal contents through a congenital diaphragmatic hernia may occur beyond the neonatal period. CDH occurs in about 1 in 2000 births. The most common defect is the posterolateral (Bochdalek) type. Over 90% of the patients will be diagnosed either antenatally or will present with respiratory distress in the first few hours of life. In these cases there are few diagnostic problems. There is a significant mortality associated with this group. However, about 5% to 30% of diaphragmatic hernias present beyond the neonatal period. Although the mortality in this group is low, the morbidity may be significant. The late presenting congenital diaphragmatic hernia poses considerable diagnostic and anaesthetic challenges because of its varied presentation often resulting in diagnostic delay, inappropriate treatment, and potential fatal outcome. We report the successful anaesthetic management of a 9 month old infant who was planned to be operated for tension pneumothorax and finally diagnosed as CDH intraoperatively.

Key words : Congenital diaphragmatic hernia, Pneumothorax.



CASE REPORT

An 9-month-old male infant weighing 8 kgs referred to our hospital with severe respiratory distress. No h/o trauma, No loss of consciousness, nausea or vomiting was reported .

O/E : Infant was restless and agitated. Pt. was a febrile, HR :150/min, RR :62/min with nasal flaring, subcostal and intercostal retraction and grunting. Breath sounds: diminished on left side. Blood pressure : normal, heart sounds : normal with no murmur.

Investigations: Complete blood count and other investigations revealed no abnormality. Arterial blood gas on room air revealed PCO2=55 mm Hg, PH=7.25, PO2=35 mm Hg, and co2 content 19 mEq suggesting metabolic acidosis.

Chest X-Ray (CXR) revealed radiolucent left hemithorax with mediastinal shift to right side.(Fig-1A). There was no referral note with the patient; hence further details of clinical findings and treatment were not available.

Nasal O2 was administered however SpO2 of patient remained less than 90%. Surgical consultation was done for placement of chest tube with impression of patient having left sided tension pneumothorax.

The baby [ASA grade III E] was shifted to Operation Theatre and intravenous access secured with 24-G i.v. cannula. Baby was premedicated with Inj. Atropine 0.01mg/kg i.v., Inj. Midazolam 0.03mg/kg i.v., Inj. Ondansetron 0.08mg/kg i.v. Precautionary measures taken and monitors attached. Baby was administered inj .ketamine 15 mg i.v. and sevoflurane in 100% oxygen using JR Circuit maintaining spontaneous ventilation.

A left-sided intercostal drain (ICD) was inserted which drained serosanguinous fluid, A second ICD was inserted which began to drain blood stained fluid. After ICD insertion there was temporary relief of symptoms. However paediatric surgeon was not convinced with the diagnosis and scheduled the patient for thoracotomy with diagnosis of congenital lobar emphysema (CLE) or large cystic lung disease. Radiologist confirmed that the CXR finding is not compatible with pneumothorax; and the large cystic lesion may be due to a gas filled stomach herniated into the left hemithorax because of absence of gastric fundus gas under diaphragm. Nasogastric tube (NGT) was placed and follow up CXR revealed reduction in size of the radiolucent lesion in left hemithorax and position of NGT in left hemithorax.(Fig-1B).

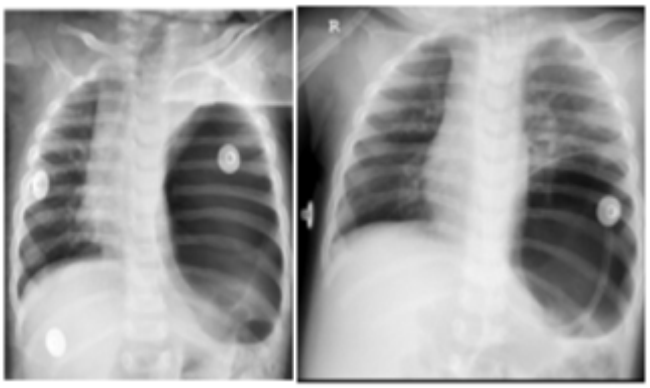


Figure findings are suggestive of herniated stomach to left hemithorax 1A: CXR reveals cystic lesion in left hemithorax with mediastinal shift and pushes left hemidiaphragm downward. Gastric gas shadow isn't seen in normal position.

Figure 1B : CXR after placement of NGT reveals position of NGT in left hemithorax and reduce size of mentioned cystic lesion in Fig-1A.

As soon as the decision of thoracotomy was done, pt. intubated with uncuffed , portex ETT no 4 and maintained on oxygen, and sevoflurane along with muscle relaxant vecuronium bromide. Defect was noted in the posterolateral part of the left dome of the diaphragm. There was no peritoneal sac over the stomach which signed the congenital nature of the hernia. The two ICDs were found to have traumatized the stomach and were draining blood and gastric contents. The tubes were disconnected, the hernial contents reduced, perforation in the stomach was closed. An ICD was placed in the pleural cavity and the diaphragmatic defect was repaired by primary closure. The ipsilateral lung was found to be hypoplastic and collapsed. The patient was shifted to the intensive care unit post operatively for ventilatory and hemodynamic support without reversal of neuromuscular blockade. Immediate postoperative chest radiograph revealed mild pleural effusion in the left hemithorax. Over the next three to five days, both lobes of the lung were found to have re-expanded on serial chest radiographs. No findings suggestive of lung hypoplasia seen and pt. extubated on the fifth postoperative day . Pt. remained hemodynamically stable during the postoperative course. Chest expansion techniques and physiotherapy helped to aid in his recovery. Subsequently he was transferred to the general ward on the seventh postoperative day and he recovered without further complications.

Discussion : Congenital diaphragmatic hernia(CDH) can present with various clinical presentations.

Misdiagnosis is the most important things in management of late presenting CDH which led to delayed diagnosis of patient and appropriate treatment [1-4]. In study by Baglaj more common wrong diagnosis according to CXR were, pneumothorax, pleural effusion with or without pneumonia, lung cyst, hydro pneumothorax or pyopneumothorax, pneumatocele and even mediastinal or paravertebral mass when the lesion was opaque[5].

There are a number of case reports in the literature where late presenting diaphragmatic hernias have been misdiagnosed as pneumothorax and patients have undergone unnecessary emergency thoraco-centesis[6-9].

In our patient although the clinical presentation was suggestive of left sided pneumothorax, a differential diagnosis of left CDH was considered, confirmed by chest radiograph after naso- gastric tube insertion, which showed stomach on the left side of thorax, and also relieved his respiratory symptoms. The inappropriate insertion of a chest drain, although relieving the symptoms temporarily, may result in serious consequences by damaging intrathoracic abdominal viscera. The other diagnostic tools available for the accurate diagnosis of late presenting CDH include CT scan, MRI, and upper or lower GI contrast studies9.In conclusion, the diagnostic possibility of late presenting CDH should be considered in unusual cases of pneumothorax, especially in the absence of trauma so that thoracocentesis can be avoided.

Respiratory support is most important in the emergency management for the patient with CDH. Blow-by oxygen or bag-masking can lead to gastric and abdominal distension and compression of the lung and should therefore be avoided. Patients presenting with severe respiratory distress should be immediately intubated and ventilated with low peak inspiratory pressure to minimise lung injury. A NG Tube connected to continuous suction should be placed in the stomach for decompression of the abdominal contents and decreasing respiratory compromise. Extracorporeal membrane oxygenation is generally reserved for patients who continue to have hypoxia and hypercarbia refractory to conventional ventilation.

Surgery was performed via thoracotomy in our patient. Chest drain insertion was usually performed at the end of the operation for drainage to help the affected lung to re-expand. The thorascopic or laparoscopic approach for the repair of late-presentating CDH has been described recently[10]. However, it is not recommended

in unstable children with mediastinal shift or acute incarceration of CDH because the insufflation pressure can contribute to circulatory collapse and cardiac arrest.

CONCLUSION

Late presenting congenital diaphragmatic hernia may have varying presentation and may be misleading. Anesthetic management of such patients with CDH poses a significant challenge to anesthesiologists especially juniors.

Understanding of the related pathophysiology, clinical picture, anesthetic problems and the specific considerations for perioperative anesthetic management are important for proper management of such cases especially diagnosed on table with subsequent better outcome.

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