A report on postero-lateral type of congenital diaphragmatic hernia

Dr. Arun Kumar S. Bilodi[1]
Dr. B.N. Sinha[2]
Dr. Neeraj Jain[3]
Dr. Vibha Jain[4]

Abstract

A male child aged 9 months came to the Paediatric OPD of Nepalgunj Medical College Teaching Hospital, with difficulty in breathing, cyanosis, clubbing and cervical lymphadenopathy. Congenital diaphragmatic hernia was detected after radiological examination. The case is reported.

Keywords: Diaphragmatic hernia; Septum Transversum; Foramen of Bochdalek.

Introduction

Diaphragm is a musculo tendinous partition separating the thoracic cavity from the abdominal cavity. This complete separation of two cavities (ie. thoracic & abdominal) is the characteristic feature of mammals.[8] It has a central tendon on which the heart is situated with dome-shaped cupolae on each side. The right cupola is at higher level due to the presence of liver.

Development

Definitive musculo-tendinous diaphragm is derivative of 4 embryonic structures.

a. Septum transversum

b. Pleuro peritoneal membrane

c. Paraxial mesoderm of the body wall

d. Esophageal mesenchyme

Some of the myoblast that arise in the septum trasversum emigrate into pleuro peritoneal membrane pulling the phrenic nerve along with them. Most of the Septum transversum gives rise to non-muscular central tendon of the diaphragm.[3]

Congenital diaphragmatic hernia

One of the pericardio peritoneal canals fails to close and allows developing abdominal visceras to bulge into pleural cavity. It occurs in about 1 in 2500 livebirth. The left side of diapharagm is involved 4 to 8 times more often than the right because the left pericardio-peritoneal canal is larger and closes later than the right. All the congenital diaphragmatic hernia should be corrected at birth. If the hernia has resulted in severe pulmonary hypoplasia, the newborn may die of pulmonary insufficinecy even if the hernia is repaired.

Case Report

A male child of 9 months came to the OPD of paediatrics of Nepalgunj Medical Teaching Hospital with the history of difficulty in breathing, frequent attacks of cyanosis associated with fever, cervical lymphadenopathy since 6 months of age. The child was the second son of his parents and was born at home. There was no antenatal checkup. On clinical examination, this child had cyanosis, clubbing and cervical lymphadenopathy with no edema. Systemic examinations were normal, no organomegaly was found.

The X-ray of the chest, PA View, showed hernia of the stomach as evidenced by the presence of gas in the fundus on the left side of the chest (Fig. 1). The hernia occurred most probably through a gap in diaphragm due to failure of development of pleuro-peritoneal septum on the left side. The result is - no closure of pleuro-peritoneal canal, which has permitted herniation of abdominal contents into the chest. The left lung shadow has been deeply intended on its medial side. The shadow of the stomach has overlapped cardiac shadow, which has been pushed up and to the right. The cardiac shadow has extended to a larger extent to the right of the mid line. The shadow of the right lung appears to be overlapped and pressed by the heart.

The nature of the sequence will be mal development of the lung resulting into dyspnoea as well as dextro cardiac position of the heart may affect the functioning of the lung. The shadow of trachea is in the mid line. The right dome is higher as usual due to the presence of liver.

Discussion

• Moore and Persaud (1998) said, "Postereo lateral defect of diaphragm is the only relatively common congenital anomaly of the diaphragm occurring in 1 in 2200 newborns associated with herniation of the abdominal contents into the thoracic cavity. Life threatening breathing difficulties maybe associated with this anomaly because of inhibition of development and inflation of the lung."[5]

• Bailey & Love (1995) pointed out that "Hernia through foramen of Bochdalek is really the persistence of pleuro-peritoneal canal and opening is in the dome of the diaphragm posteriorly. It is more common (Diaphragmatic hernia) in children and present with severe respiratory distress."[1] "There is a classical triad of respiratory distress, apparent dextrocardia & scaphoid abdomen" (Bailey & Love).[1] In the present study, there is H/o difficulty in breathing, but no H/o dextrocardia, and Scaphoid abdomen.

• Mc. Gregor (1986) had simlar view, "Postero-lateral hernia occurs in 1 in 2000 livebirths leading to herniation of abdominal contents into the thoracic cavity, which compresses the lung and displaces th heart. The hernia results from defective formation and fusion of pleuro-peritoneal membrane, which closes the pleuro-peritoneal canal in the foetus. There is larger defect in
the postero-lateral portion of the diaphragm."[2] Hernia do occur most commonly on the left side through Left hemi diaphragm.

• Last (1995) gave the following reason of this problem – "If the lumbar and costal elements fail to fuse at all, there will be a gap known as Bochdalek's foramen, usual site for congenital diaphragmatic hernia. This type is common on the left side, possibly because of pressure from the liver on the right encourages earlier closer there and large defect present at birth may result in stomach and abdominal visceras herniating into thoracic cavity.[4]"

• Gray (1995) said "postero-lateral hernia occurs as a result of defect in posterior diaphragm in the region of 10th and 11th ribs. It is more common on the left side, presents with abdominal contents in the left hemithorax at birth. Respiratory distress often results and condition is LIFE threatening."[10]

• Saddler (1995) said "it occurs in newborns, 1 in 2000 and is most frequently caused by failure as one or both pleuro-peritoneal membrane to close the pericardio-peritoneal canal. The peritoneal and pleural cavities are then continuous with one another
along the posterior body wall. Such defects allow abdominal contents into pleural cavity contents in 80-90% of cases; hernia is on the left side.
The heart is pushed anteriorly while the lungs are compressed and often hyphoplasic. A large defect is often associated with high mortality rate (75%) due to pulmonary ***hypoplesia and pulmonary dysfunction***.[6] The organs like stomach, intestine, spleen and part of liver may enter into the thoracic cavity.[7]

• Nelson (1995) also mentioned that congenital diaphragmatic hernia is present where there is herniation of abdomen contents through Postero lateral foramen of Bochdalek. These lesion presents with profound respiratory distress in neonatal period with significant 30 to 40% mortality rate.[6]

• Snell (1995) said congenital diaphragmatic hernia occur through pleuro-peritoneal canal (More common on the left side due to failure of fusion of septum transversum and pleuro peritoneal membrane).[9]

• Sahana (1993) says that as a result of error in the development gap known as Bochdalek's gap at Vertebro Cortal angle fail to close through which herniation (of abdominal viscera) occurs known as Congenital diaphragmatic hernia. Usually in this type of hernia, there is no hernial sac.[8]

• Rege M Vivek (1995) associated Anomalies like malrotation of gut and patent ductus arteriosus occurs from 40 to 57%. Only 5-7% of these patients survive with multiple anomalies like anencephaly, myelomeningocele, hydrocephalus & CVS anomalies like VSD, triosomies[13,18], Coactation, GIT anomalies like atresia of oesophagus and cleft palate.

But in the present study no such above anomalies were found.[11]

Thus all the above authors have described postero-lateral diaphragmatic hernia with herniation of abdominal contents into the pleural cavity and consequent lung hypoplasia and displacement of the hearts.

The present case has shown ideal textbook description where there is herniation of the fundus of the stomach in the left pleural cavity (by the presence of gas) and also indentation of the left lung on its medial side. The shadow of the stomach is overlapped by the cardiac shadow, which has been pushed up and to the right.

Conclusion

In the present study, the child is suffering from cogenital diaphragmatic hernia (Bochdalek) reported at Paediatric OPD of Nepalgunj Medical College Teaching Hospital with history of difficulty in breathing. The X-ray confirmed congenital diaphragmatic hernia, stomach lying in the pleural cavity with the consequence of hyphoplasia of the lung. This case shows the absence of antenatal checkup and diagnosed by ultrasonography. The child is now showing congenital hypoplasia of the lung, which should have been relieved at birth by surgical intervention. It is acute emergency surgery for congenital diaphragmatic hernia and impairment of development of the lung, which in turn will relieve respiratory distress and dysponea.

This is a serious condition that requires surgery in order to allow the lung to expand. In advanced countries, surgeries are done on foetuses in utero by opening the uterus restoring the herniated organs back into abdominal cavity and repairing the foetal diaphragm.[3]

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