Bochdalek hernia is a rare condition that usually presents in the neonatal age. Rarely it presents late and is usually misdiagnosed. We came across a Bochdalek hernia, which presented at the age of 15 years and was initially misdiagnosed as pulmonary tuberculosis. The case history, diagnosis and management of this condition alongwith literature review are presented here.

INTRODUCTION

Bochdalek hernia is a rare entity. It usually presents in the neonatal age with respiratory symptoms. It has a high mortality rate due to associated lung hypoplasia despite the newer modalities of treatment. Sometimes it presents late with respiratory symptoms, sub acute intestinal symptoms or an incidental finding. These cases usually have better prognosis.

CASE REPORT

A 15 years old boy presented to the medical out patient department with complaints of cough, vomiting and abdominal pain for one month. He also had constipation and abdominal distension for three days. The vomiting was non bilious and after every meal. Examination showed a thin lean boy with scaphoid abdomen, normal bowel sounds and decreased breath sounds over the left side of the chest. The X-ray showed left pleural effusion. The differential diagnosis included Pulmonary TB, abdominal Koch with sub acute intestinal obstruction. Surgical consultation was made for intestinal obstruction. In the surgical ward X-Rays were viewed with suspicion of diaphragmatic hernia. Detailed history revealed that he had on and off symptoms of sub acute intestinal obstruction since three years of age. He had no respiratory symptoms at all. Examination revealed scaphoid abdomen with bowel sounds in the left hemithorax. Barium meal and follow through confirmed left side diaphragmatic hernia. Peroperatively there was a large postereolateral diaphragmatic defect through which most of the small and large intestine and spleen had herniated into the left chest. There was also malrotation of gut with bands of Ladd giving rise to obstructive symptoms and a large stomach. The contents were reduced and the defect repaired with silk. Malrotation was also corrected at the same time. Although there was some difficulty in closing the abdomen, the patient had an uneventful recovery in ICU. Post-operative X-Ray after one week showed remarkable lung expansion.

DISCUSSION

Bochdalek hernia is a rare entity. McCulley gave its earliest description in 1754. Bochdalek in 1848 described in detail the embryological aspects of the hernia. There are three types of congenital diaphragmatic hernias; Morgagni, Hiatal and Bochdalek Hernia. The most common type (80%) is the posterolateral defect or Bochdalek hernia.

Its incidence in the neonates has been quoted to be between 1:2000–5000. In adults the incidence has variably been reported to be between 0.17% reported by Mullens et al to as high as 6% by Gale. It was calculated on the basis of retrospective CT Scan studies done for some other purpose. The exact cause of Bochdalek hernia is unknown. It has been associated with antenatal use of thalidomide, Quinine, nitrofenide, antiepileptics or deficiency of vitamin A.

The usual presentation is in the neonatal age with respiratory distress, vomiting and cyanosis. The cause of the symptoms is due to pulmonary hypoplasia, malrotation or strangulation of bowel through the narrow hernial orifice. Adult presents in a different way. Presentations include incidental finding at laparotomy performed for some
other purpose or CT and MRI done for another disease. Intermittent sub acute intestinal obstruction, or persistent cough and respiratory problems are other modes of presentation.

The diagnosis in children is based on clinical examination in which there is a scaphoid abdomen and bowel sound in the chest. In good centers it is now diagnosed antenatally by ultrasound in 40–90% of the cases. Postnatally simple X-Ray chest or in cases of doubt barium meal and follow through are usually diagnostic. In the adults diagnosis is usually missed until there is a high index of suspicion. Thomas et al have found that nearly 38% of adults are misdiagnosed in this way. It has been wrongly diagnosed as pleural effusion, empyema, lung cyst and pneumothorax. In asymptomatic adults the diagnosis is usually made by CT or MRI performed for some other condition.

The surgical approach can be done either through abdomen or the chest. The abdominal approach has the advantage of correcting the malrotation at the same time. The contents usually include most of the small intestine and portion of large intestine. The spleen is also nearly always present in the chest. Some times left lobe of the liver, left adrenal gland or left kidney may also be present. The incarcerated loops of bowel should be carefully dealt with. The rent in the diaphragm is closed with non-absorbable interrupted sutures. If the defect is too large mesh can also be inserted. The abdominal wall closure may be problematic, as most of the intestine has never resided in the abdomen. Charles et al. recommend that only skin closure with delayed closure of the muscles should be done in such situations. Laproscopic repair of the hernia has also been reported.

The overall prognosis in the neonatal congenital diaphragmatic hernia has not improved much. Despite the use of latest technique of extra corporeal membrane oxygenation the survival is still between 50–65%. The degree of pulmonary hypoplasia determines the outcome. For this reason it is regarded as physiological emergency rather than surgical emergency. In adults however the prognosis is much better probably because of the absence of pulmonary hypoplasia. The lung on the affected side is nearly always hypoplastic to some extent. Future therapies like intrauterine tracheal plugging or occlusion are under trial.

REFERENCES


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