Delayed Presentation of Congenital Diaphragmatic Hernia (Bochdalek Type)

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ABSTRACT

Objective: To document various clinical manifestations of Bochdalek congenital diaphragmatic hernia presenting late and identify reasons of late presentation.

Design: Prospective study.

Place and duration of study: The department of pediatric surgery, The Children’s Hospital and The Institute of Child Health, Lahore, from July 2001 to July 2005.

Patients and methods: 40 children with Bochdalek congenital diaphragmatic hernia (CDH) who presented after one month of age were studied. Detailed history, examination and results of required investigations were recorded. Data was processed regarding manifestations, reasons for late diagnosis and outcome of these cases.

Results: 45 children with Bochdalek type of CDH met the inclusion criteria. Three main modes of presentation were observed. Thirty five (77.78%) children presented with respiratory symptoms, nine (20%) had gastrointestinal symptoms and one (2.2%) patient manifested combined symptoms. Diagnosis was made on chest x-ray (68.88%), chest ultrasound (11.11%), upper GIT barium contrast study (15.56%) and chest CT scan (4.45%). Four (8.89%) patients died preoperatively. Surgical complications included wound infection in two (4.88%), burst abdomen in four (9.75%), adhesion obstruction in two (4.88%) and recurrence of hernia in two (4.88%) patients. Four patients died postoperatively. The overall prognosis was favorable.

Conclusion: Delayed CDH has a variable presentation. Early diagnosis can be made if it is included in the differential diagnosis of recurrent chest infection and gastrointestinal tract symptoms. Chest radiograph with nasogastric tube and chest ultrasound in suspected cases are useful tools to prevent delay in final diagnosis.

Key words: Congenital diaphragmatic hernia, Bochdalek type, late presentation.

INTRODUCTION

Embryologically diaphragm is composite of four components i.e. septum transversum, pleuroperitoneal membranes, oesophageal mesentery and musculature of body wall. The congenital diaphragmatic hernia (CDH) results either due to incomplete closure of diaphragm or early migration of gut from umbilical coelomic into abdominal cavity before complete formation of diaphragm (around 10 weeks). The most common defect is posterolateral defect being found in 60-85% of cases. It is often referred to as Bochdalek’s foramen. This defect is established due to failure of formation or fusion of pleuroperitoneal membranes with septum transversum.

Thus pericardioperitoneal canal is not sealed permitting the peritoneal and pleural cavities to remain continous with one another and allowing abdominal viscera to herniate into thorax. Herniation through this defect is called Bochdalek hernia. Most of these defects occur on left side. The right side is protected slightly by early development of right hemidiaphragm and by presence of liver. If pleuroperitoneal canal closes but fail to become muscularized, a hernial sac results which is present in 10 to 15% of cases. Most CDH cases present in neonatal period and even diagnosed antenatally. The neonates present uniformly with respiratory symptoms, cause little difficulty in diagnosis but have high mortality rate. If CDH presents outside the neonatal period, it is called congenital diaphragmatic hernia with delayed presentation. It accounts for 5-25% of total CDH cases. Because its clinical picture is chronic, variable, atypical, and inconsistent, its diagnosis becomes more difficult, complex and challenging. The radiologic features are also troublesome, so delay in treatment is common. Young children manifest with respiratory or gastrointestinal symptoms. Children presenting with gastrointestinal symptoms are significantly older than those presenting with respiratory symptoms. Respiratory symptoms include persistent cough, fever (repeated chest infection), dyspnea, wheezing, cyanotic spells,
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Grunting respiration and retrosternal discomfort. Gastrointestinal manifestations consist of abdominal pain, recurrent vomiting and nausea. In addition anorexia, diarrhea, constipation and failure to thrive may be present. Some patients may remain asymptomatic and are detected incidentally. Physical examination may reveal ipsilateral over distention of chest, increased anteroposterior diameter, intercostal and subcostal in drawing. The breath sounds are decreased over affected side. The heart tones are muffled with displaced apex beat and may be heard on right side in left sided hernia. The late presenting CDH represents a considerable diagnostic challenge. Cases have been misdiagnosed as pneumothorax, pleural effusion, lung cyst and bullae. The radiologic evaluation includes chest radiograph which is mandatory but may be non contributory or even misleading in some of cases. Barium contrast study may be added for confirmation. Chest ultrasound is also a supportive tool in suspected cases. CT scan chest with oral contrast confirms the diagnosis. Laparoscopy is also a useful diagnostic tool. MRI is rarely indicated. After reduction of contents the defect is repaired preferably by open abdominal approach. Most thoracic surgeons prefer thoracic approach. Patch repair is rarely indicated. Thoracoscopic and laparoscopic approach for repair has been successfully done in some institution claiming it feasible and safe technique, with reasonable functional and cosmetic results and a very quick recovery.

MATERIAL AND METHODS

This analytical study was conducted in the department of pediatric surgery, the children hospital and the institute of child health, Lahore, from July 2001 to July 2005. In this study those children were included who presented with CDH after one month of age and those presented before one month of age or with recurrent hernia were excluded from the study. A detailed history and complete physical examination was performed. Routine blood and urine examination was done in all cases. Chest radiograph, posteroanterior view was mandatory and chest ultrasound was performed in some doubtful cases who presented in emergency room. Special investigations including barium contrast study and CT scan chest was done in few cases for confirmation of diagnosis. In selected cases echocardiography was also done. Those patients who landed in emergency room were operated after resuscitation and optimizing the condition of the patient. Elective surgery was offered to cases who were admitted through out patient department. A detailed record of all patients was maintained. Complete clinical examination and required investigations were obtained on regular follow up visits. Each case was followed up for six months to one year of duration.

RESULTS

During the study period cases of Bochdalek type of CDH were admitted from different areas of Pakistan with no significant concentration from any area. The total number of patients was 45. Out of 45 patients 36 were, male contributing 80% to the total and 9 were female making 20% of the total. The male to female ratio was 4:1. The age range was from one month to four year with mean age of 10.16 month. The maximum numbers of 30 patients (66.67%) manifested within first year of life. Respiratory problems (cough, respiratory distress) were the most common mode of presentation (Table.1). Out of 45 patients 35 (77.78%) manifested with respiratory symptoms while 9 patients (20%) had symptoms related to gastrointestinal system (vomiting, abdominal pain).
Only one patient (2.22%) had combined symptoms of both systems (cough and abdominal pain). Considering the duration, only six patients (13.34%) had symptoms since birth but presented after neonatal period. As regards mode of admission, 22 patients (48.49%) landed in emergency ward with acute symptoms, 12 patients (26.87%) were shifted from medical section and 11 patients (24.44%) were admitted from out patient department. All those patients manifested acutely, had history of previous recurrent chest infection.

Chest radiograph was the most useful modality to diagnose CDH (Fig.1). It was performed in all cases including 10 suspected cases, in which diagnosis was established with placement of nasogastric tube. So overall, in 31 patients (68.88%) correct diagnosis was made with the help of chest radiograph. Barium contrast study (Fig.2) was required in 5 cases (11.11%) and in two cases (4.45%) CT scan was needed (Fig.3). Chest ultrasound was carried out in emergency in 7 (15.56%) doubtful cases.

Echocardiography was done in five (11.11%) selected cases. Four patients (8.88%), who appeared marasmic and developed sepsis, did not respond to stabilization and died before surgery (Table-III).

Table I. Various Clinical Manifestations (n=45)

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>No. Of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Respiratory</td>
<td>35 (77.78%)</td>
</tr>
<tr>
<td>GIT</td>
<td>09 (20%)</td>
</tr>
<tr>
<td>Combined</td>
<td>01(02.22%)</td>
</tr>
</tbody>
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All patients were operated via an abdominal approach except one case of right sided defect which was approached through thoracotomy. The defect was more common on left side affecting 39 patients (86.67%) while right side was involved in six cases (13.33%). Sac was found in 10 cases (24.39%). Primary repair without patch was carried out in all cases.

During early postoperative period four patients (9.75%) died. Complications (Table-II) included, two patients (4.88%) showed wound infection. Burst abdomen was observed in four cases (9.75%). After preoperative death of four patients the remaining 41 cases were followed up for period ranged from six month to one year. Ten patients (24.35%) showed mild chest infection. Adhesion obstruction was noticed in two patients (4.88%). Two patients (4.88%) were readmitted with recurrence of hernia which necessitated second surgery.

DISCUSSION

The Bochdalek hernia is the most common type of CDH. Majority of cases present in neonatal life uniformly with respiratory symptoms but a considerable number of patients of 5-25% had delayed presentation, outside neonatal period. These patients have vague and nonspecific respiratory, bowel or combined symptoms. Fewer than 450 patients with late presenting CDH have been reported in the literature as of December 2006.

In the present study 35 cases (77.78%) presented with respiratory symptoms, of which only 5 cases (14.3%) having duration of respiratory symptoms within a week, were identified as CDH. The remaining 30 cases (85.7%) were mistreated somehow or the other way as recurrent chest infection before final diagnosis. All of these cases lacked chest radiograph during early course of symptoms. In these cases first radiograph of chest was obtained minimally three weeks after initiation of symptoms, which contributed to erroneous assessment of radiological findings and undesired delay in diagnosis. Three patients received antitubercular therapy on trial basis (before being diagnosed as case of CDH) because previously they had multiple antibiotics for recurrent chest infection with no improvement. Two patients misdiagnosed as pleural effusion and two patients with herniated stomach simulated pneumothorax, were mistakenly chest intubated. The thoracostomy tube perforated the stomach and diagnosis of CDH was established when oral fluid feed started coming through chest
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In our study misdiagnosis of CDH as chest infection was the most common reason of delay diagnosis. The same reason of delay is also mostly documented in world literature. 3-6,7,14 The long time elapsed between appearance of symptoms and getting first x-ray chest, was another cause of delaying diagnosis. This made the radiologic assessment cloudy, leading to misinterpretation of findings, thus further delaying the diagnosis. Similarly paucity of knowledge of uncharacteristic and varied presentation also contributed to undesired delay. Technical explanation of delayed presentation can be justified on the basis of non significant pulmonary hypoplasia in all cases of the study. This observation is also supported by Javier Anaya in a report 18. Other logical explanation such as plugging of defect by liver or spleen, presence of sac and well formed posterior rim of diaphragm has been described in literature 12. We have found good quality posterior rim in 15 cases (36.58%) and sac in 10 patients (24.39%), but these are not constant features of delayed presentation. Mechanical or pressure change in the thoracoabdominal cavities has also been postulated as contributory factor 6.

All patients presented with respiratory symptoms were regarded and treated for recurrent chest infection leading to delay in final diagnosis. So delay in diagnosis can be prevented by high suspicious of CDH in all such cases which do not respond to multiple antibiotics. Secondly a clinical clue of usually distended abdomen is present in most patients of pneumonia in contrast to CDH cases with flat or scaphoid abdomen 6. Thirdly, lung consolidation on chest radiograph has pulling tendency rather than pushing effect as in CDH. Fourthly, placement of nasogastric tube followed by chest film usually diminishes the confusion of pneumonia or pneumothorax and provides diagnosis. Fifthly, ultrasound should be used supportively in all suspected diseases of the diaphragm 19. Usually ultrasound in chest infection is not widely in practice. With limited experience we have found it very informative and supportive modality in suspected cases. Moreover, its importance lies in its easily availability, noninvasiveness (no radiation as in C T scan), and rapidity (less time consumption compare to Barium contrast study) and is bed side manoeuvre. The ultrasonic findings such as absence of free pleural fluid and presence of an air containing cavity with peristaltic activity suggesting bowl, aids information to diagnosis. In fact, Forter et al has suggested that ultrasound should be the complementary imaging modality of choice in all suspected cases 4. Similarly, there should be suspicious of CDH in those patients having recurrent abdominal pain unresponsive to medication.

The overall prognosis was good. During early postoperative period four patients (9.75%) died. These patients developed chest infection followed by sepsis and death. Wound infection was observed in two patients (4.88%), who were successfully treated with daily dressing. Four patients (9.75%) who appeared weak and marasmic at presentation, got burst abdomen. This event can be justified by increased intra-abdominal pressure after reduction of contents in comparatively low abdominal capacity, especially in malnourished patients with thin and weak abdominal muscles. Burst abdomen was repaired successfully. During early follow up period chest infection was found in ten patients (24.35%), who responded to oral antibiotics. Four patients were readmitted, two (4.88%) with adhesion obstruction and two (4.88%) with recurrence of hernia. Adhesion obstruction may occur after any abdominal surgery. One patient was managed conservatively while in other laparotomy with adhesiolysis was needed to relieve the obstruction. Recurrence of hernia necessitated second surgery four month after first operation. The undesired occurrence of recurrence has been reported in literature 13.

CONCLUSION

In conclusion, late revealing CDH has very uncharacteristic and inconsistent respiratory and less frequently gastrointestinal symptoms. Delay in diagnosis can be prevented if high index of suspicious of this condition is kept in mind in differential diagnosis of recurrent non specific respiratory or gastrointestinal symptoms. A plain radiograph of chest with nasogastric tube might help in establishing diagnosis. Before going for contrast study or C T scan, chest ultrasound is investigation of choice in all suspected cases especially in emergency room. The overall outcome of late presenting CDH is favorable.

REFERENCES


