Clinical Presentation and Radiographic Diagnosis of Non-traumatic Childhood Diaphragmatic Hernia

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Between years 1993 and 2002, a total of 90 patients were diagnosed as diaphragmatic hernia in our hospital. Excluding 2 cases with traumatic diaphragmatic hernia, there were 15 cases (17.0 %, 15/88) with non-traumatic diaphragmatic hernia in childhood (13 Bochdalek hernias and 2 Morgagni hernias) that presented beyond the neonatal period. All of the 15 patients were without major symptoms in the neonatal period. These patients with childhood diaphragmatic hernia might be asymptomatic or present with various gastrointestinal or respiratory symptoms. Nearly half of the patients (46.7%) had hernia sac found at surgery, which might imply a postnatal cause rather than congenital in a substantial proportion of childhood diaphragmatic hernia. Radiography is the most important imaging modality but may not be adequate for accurate diagnosis in some patients. Barium study was performed in 6 patients and always led to accurate diagnosis in our series. The prognosis of childhood diaphragmatic hernia is good since there is little or no pulmonary hypoplasia. However, delayed diagnosis and treatment may result in life-threatening complications. In this article we discuss the clinical and imaging presentation of childhood diaphragmatic hernia.

Key words: Children, Diagnosis, Children, gastrointestinal tract; Hernia, diaphragmatic

Diaphragmatic hernia is the most important disease of the diaphragm, and it usually causes acute severe respiratory distress at birth, including cyanosis and dyspnea [1]. It is seldom problematic obtaining accurate diagnosis by chest roentgenograms in newborn patients with acute respiratory distress. However, misinterpretation is not uncommon among patients with delayed diaphragmatic hernia beyond the neonatal period. This is because not only these patients have healthy or asymptomatic neonatal period but also it causes various nonspecific respiratory or gastrointestinal symptoms late in life. Even though initially silent, childhood diaphragmatic hernia may result in devastating complication [2-7]. Therefore, accurate diagnosis in these cases is based on initial suspicion and familiarity with imaging findings. We reviewed 15 cases of childhood diaphragmatic hernia in our hospital. The clinical presentation and imaging features of these cases were presented in this article.

PATIENTS AND METHODS

We retrospectively collected a total of 90 patients (0 to 19 years of age), who were discharged with associated diagnosis of diaphragmatic hernia during 1993 to 2002. On the basis of detailed review of their clinical history, patients with congenital diaphragmatic hernia (onset within the neonatal period) (n=49), hiatal hernia (n=10), eventration (n=14), and traumatic diaphragmatic hernia (n=2) were excluded. Among them, 15 patients had childhood diaphragmatic hernia (beyond one month old) which was confirmed and treated with surgery. The clinical courses, surgical findings, and radiological assessment were recorded.

RESULTS

The neonatal period was uneventful for these 15 cases (aged 3 months to 12 years; median = 1 year) (Table). Three patients had previous normal chest radi-
Table 1. Summary of clinical features of the fifteen patients

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age*</th>
<th>Sex</th>
<th>Symptom/Sign**</th>
<th>Type of hernia</th>
<th>Ancillary imaging studies</th>
<th>Presence of hernial sac</th>
<th>Associated anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1y</td>
<td>M</td>
<td>Resp</td>
<td>Bochdalek</td>
<td>Chest CT, Barium study</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>12y</td>
<td>F</td>
<td>GI</td>
<td>Bochdalek</td>
<td>—</td>
<td>+</td>
<td>Gastric volvulus</td>
</tr>
<tr>
<td>3</td>
<td>4m</td>
<td>M</td>
<td>Asymptomatic</td>
<td>Bochdalek</td>
<td>Fluoroscopy</td>
<td>+</td>
<td>—</td>
</tr>
<tr>
<td>4</td>
<td>6m</td>
<td>M</td>
<td>Resp</td>
<td>Morgagni</td>
<td>Chest MRI, Barium study</td>
<td>+</td>
<td>Malrotation</td>
</tr>
<tr>
<td>5</td>
<td>3y</td>
<td>M</td>
<td>Resp</td>
<td>Bochdalek</td>
<td>—</td>
<td>+</td>
<td>—</td>
</tr>
<tr>
<td>6</td>
<td>1y</td>
<td>F</td>
<td>GI</td>
<td>Bochdalek</td>
<td>—</td>
<td>+</td>
<td>Gastric volvulus</td>
</tr>
<tr>
<td>7</td>
<td>2y</td>
<td>M</td>
<td>GI</td>
<td>Bochdalek</td>
<td>Barium study</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>1y</td>
<td>M</td>
<td>Asymptomatic</td>
<td>Bochdalek</td>
<td>—</td>
<td>+</td>
<td>Lung hypoplasia, Malrotation</td>
</tr>
<tr>
<td>9</td>
<td>1y</td>
<td>F</td>
<td>Asymptomatic</td>
<td>Bochdalek</td>
<td>Abdominal MRI</td>
<td>+</td>
<td>Lung hypoplasia</td>
</tr>
<tr>
<td>10</td>
<td>10m</td>
<td>F</td>
<td>GI</td>
<td>Bochdalek</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>11</td>
<td>4y</td>
<td>M</td>
<td>Resp and GI</td>
<td>Bochdalek</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>12</td>
<td>10m</td>
<td>M</td>
<td>GI</td>
<td>Bochdalek</td>
<td>—</td>
<td>—</td>
<td>—</td>
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<tr>
<td>13</td>
<td>6m</td>
<td>F</td>
<td>Resp</td>
<td>Bochdalek</td>
<td>Barium study</td>
<td>+</td>
<td>—</td>
</tr>
<tr>
<td>14</td>
<td>9m</td>
<td>M</td>
<td>Asymptomatic</td>
<td>Morgagni</td>
<td>Barium study</td>
<td>—</td>
<td>Lung hypoplasia</td>
</tr>
<tr>
<td>15</td>
<td>8m</td>
<td>M</td>
<td>GI</td>
<td>Bochdalek</td>
<td>Barium study</td>
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</tr>
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</table>

* m = month(s), y = year(s); ** Resp = respiratory, GI = gastrointestinal

Figure 1. Case 1: a. This chest radiography, taken during the neonatal period of the patient, shows no significant finding of the diaphragm. b. Large cystic opacities occupied right hemithorax in this chest radiography which was taken at the age of one year. c. The post-contrast axial chest CT scan reveals peritoneal fat and enhanced vessels in the right thoracic cavity. The diagnosis of right Bochdalek hernia was confirmed by barium study (not shown).
Hernial sac was found in seven patients (46.7%, 7/15) at surgery. The herniated organs included colon, stomach, small intestine, spleen, and kidney. The pulmonary function was normal in the patients with lung hypoplasia in postoperative follow-up for 6 months. One of the cases with Morgagni hernia was with Down’s syndrome and atrial septal defect. All of the patients underwent repair of the diaphragmatic hernia without significant post-operative complication. The post-operative follow-up for at least 12 months period was smooth without recurrence of hernia or any sequelae.

The pre-operative chest radiographs were abnormal in all 15 patients on a retrospective review. The radiographic diagnosis of diaphragmatic hernia was correctly made in 10 cases (66.7%, 10/15) according to the appearance of elevated hemidiaphragm or abnormal intrathoracic mass or gas pattern (Fig 2a, 3a). Five were misinterpreted (33.3%, 5/15), in which two were interpreted as pneumonia of left lower lung (Fig. 4a) and one as left hydropneumothorax (Fig. 5); Furthermore, all of these patients presented with GI symptoms rather than respiratory illness. One chest radiograph was interpreted as necrotizing pneumonia of right lower lung (Fig. 1b) in a 1-year-old boy who presented with cough and rhinorrhea without any respiratory distress. A faint cardiophrenic mass was initially overlooked in one asymptomatic case with Morgagni hernia. The ancillary imaging modalities which had been used in these patients include barium study (Fig. 2b, 4b) in six patients, fluoroscopy in one, chest CT (Fig. 1c) in one, and MR imaging (Fig. 3b) in two. Most of these ancillary studies were used to confirm the diagnosis of diaphragmatic hernia except for the chest CT which was performed under the impression of necrotizing pneumonia of right lung.

**DISCUSSION**

Bochdalek hernias constitute the majority of childhood diaphragmatic hernias (86.7%) in our series. Bochdalek hernia was originally described as herniation through the costolobar triangles. In addition, herniation through a persistent pleuropertoneal canal with a posterior lip to the defect and herniation through larger posterolateral defects with no posterior lip are also included in the definition of Bochdalek hernia. The description of Morgagni hernia now includes a defective attachment of the anterior portion of the diaphragm to the ribs and sternum, true defects in the diaphragm posterior to the sternal and costal insertion, and combinations of the two above [1, 8-11].

Unlike congenital diaphragmatic hernias which are usually present soon after birth with severe respiratory distress, none of the 15 patients with childhood diaphragmatic hernia in our series had symptoms during their neonatal periods. The initial presentation was variable from asymptomatic to chest wall bulging or asymmetry sometimes, resulting in misleading to clinicians. Consequently diaphragmatic hernia was not included in the initial impression of these patients before the radiographic studies. When symptomatic, it may develop gradually or acutely with symptoms and

**Figure 4.** Case 12: **a.** This chest radiography shows faint opacity over left lower lung (arrows) mimicking infiltration of pneumonia. **b.** The upper GI series shows normal position of the stomach and duodenum, but abnormal opacity and gas pattern in left lower lung (arrowheads) connecting with the colon gas in the splenic flexure is noted. Further air enema (not shown) later confirmed diaphragmatic hernia of the colon.
or MR images, but it is usually not needed for diagnosis. However, the CT finding may mimic necrotizing pneumonia or CCAM, and the intrathoracic bowel gas may look similar to pneumatoceles or gas-forming abscesses (Fig. 1c). In such cases, differential diagnosis of diaphragmatic hernia should be made by careful examination the peritoneal fat and enhanced vessels in the thoracic cavity on post-contrast CT study.

Thoracocentesis should be avoided. None of the patients in our series received thoracocentesis, but it has been documented in relation to misdiagnosis of right-sided hernias as pleural effusion [13] or left-sided hernias as pneumothorax [14].

Excluding traumatic diaphragmatic hernia, there was 17.0% (15/88) of the diaphragmatic hernias presented beyond the neonatal period in our current study, which was comparable to 5~25% incidence rate in the literature [3, 4, 15]. Hernia sac was found in nearly half (46.7%) of the patients; nevertheless, the hernia contained in a hernia sac was reported in only 10% of cases with congenital Bochdalek hernia [9]. The severity of lung hypoplasia is the main prognostic factor in the newborns [16]. The overall survival rate is 60% in an aggregated report of 1224 patients with congenital diaphragmatic hernia [17]. However, if affected fetal deaths or still births are included, up to 88% of fetuses antenatally diagnosed with congenital diaphragmatic hernia do not survive [18]. The prognosis of childhood diaphragmatic hernia is good in our experience and literatures [19-21] since there is little or no problem with lung hypoplasia. Mortality or morbidity in childhood diaphragmatic hernia is often secondary to delayed complications of the GI tract, e.g., bowel strangulation/volvulus with ischemia or perforation [2-7]. On the basis of the findings of hernia sac and lack of lung hypoplasia, we suppose that a substantial proportion of childhood diaphragmatic hernia is caused postnatally by weakening or rupture of the diaphragm. Although most of the diaphragmatic hernias are believed to be congenital, such hypothesis may refer to an acquired cause of diaphragmatic hernia [19] rather than congenital in origin.

CONCLUSION

Diaphragmatic hernia beyond the neonatal period is not rare and constitutes 17.0% of non-traumatic diaphragmatic hernias in this series. The clinical presentation is rather different from congenital diaphragmatic hernias. Early suspicion and familiarity with the clinical and imaging findings are most important to accurate diagnosis. We suggest that childhood diaphragmatic hernia be suspected when clinico-radiological mismatch exists between abnormal chest radiographic findings and symptoms/signs presented by the patient. Early diagnosis and treatment is especially important to febrile patients in whom fatal complication may occur if delayed management. Plain radiography is mandatory but may not be adequate for diagnosis in our experience. Ancillary contrast study with barium or air can lead to accurate diagnosis. CT and MRI may be helpful to confirm the diagnosis in some uncertain circumstances.

REFERENCES

非外傷性兒童期橫隔膜疝氣的臨床表現及診斷

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自西元一九九三年至二零零二年間，共有九十位病人於本院診斷為橫隔膜疝氣。其中除了二例為外傷性橫隔膜疝氣外，共有十五例（17.0%，15/88）非外傷性橫隔膜疝氣病例（十三例Bochdalek氏疝氣及二例Morgagni氏疝氣），其臨床表現發生在新生兒時期之外。所有這十五個病例在新生兒時期都沒有臨床症狀。此類兒童期橫隔膜疝氣可以不具症狀，或者表現各種消化道或呼吸道症狀。將近一半的病例（46.7%）於外科手術時發現疝氣囊。此疝囊的表現，可能暗示某個比例上的兒童期橫隔膜疝氣，其實是後天性的，而非先天性的。對於此類病例，X光片檢查是最重要的影像工具，但是在某些病例上尚不足以單靠X光片檢查而獲得正確診斷。本研究當中有六例曾接受攝影，並以此獲得正確診斷。兒童期橫隔膜疝氣的預後良好，主因於病人大多沒有或只有輕微之肺臟發育不良。然延誤診斷及治療的時機仍可能導致致命的併發症。本文主要討論兒童期橫隔膜疝氣的臨床及影像表現。

關鍵詞：兒童，腸胃道；橫隔膜疝氣