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Diaphragmatic herniation diagnosed at a late stage: an evaluation of eight cases

Son evrede tanılanan diyafragmatik herniasyon: Sekiz olgu değerlendirmesi

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Background: In this study, we aimed to analyze patients with diaphragmatic hernias who had specific clinical features and interesting clinical, radiologic presentations, and had operated following late diagnosis.

Methods: A total of eight diaphragmatic hernia cases, of whom three were adults and five were children (4 males, 4 females; mean age 12.4±24 years; range 9 months to 28 years), who were diagnosed in the late period and who were operated on in our clinic between January 2006 and August 2010 were evaluated retrospectively. In all patients, primary repair of the diaphragm was performed following laparotomy and thoracotomy.

Results: The children were diagnosed following the neonatal period. Three patients were diagnosed in adulthood. Five patients had right-sided congenital Morgagni hernias, one patient had a hiatal hernia, and two patients had left-sided Bochdalek hernia. All patients were discharged between the 5th and 19th postoperative days. The average duration of hospital stay was 9.8 days (range 7 to 23 days). The follow-up time were between six months and two years, and no significant respiratory and gastrointestinal complaints were recorded. No morbidity was detected in three cases with Morgagni hernia.

Conclusion: Congenital diaphragmatic hernias are pathologies which are seen in the neonatal period, and the diagnosis of these hernias can be difficult after this period. Mortality and morbidity rates are low in cases who are followed and treated in clinics by dedicated thoracic surgeon.

Key words: Acquired; congenital; diaphragm; herniation.

Amaç: Bu çalışmada, spesifik klinik özellikleri ve ilginç klinik ve radyolojik tabloları olan ve son evrede tanı konulduktan sonra ameliyat edilen diyafragmatik hernili hastalar analiz edildi.

Çalışma planı: Ocak 2006 ve Ağustos 2010 tarihleri arasında son evrede tanı konan ve kliniğimizde ameliyat edilen üçü erişkin ve beşi çocuk toplam sekiz diyafragmatik hernili olgu (4 erkek, 4 bayan; ort. yaş 12.4±24 yaş; dağılım 9 ay-28 yıl) retrospektif olarak değerlendirildi. Tüm hastalarda laparotomi ve torakotomi sonrasında herni redüksiyonu ve primer onarımı yapıldı.

Bulgular: Çocukların tanısı neonatal dönem sonrasında konuldu. Üç olguya erişkin dönemde tanı kondu. Beş hastada sağ taraflı doğuştan Morgagni hernisi; bir hastada hiatal herni ve iki hastada sol taraflı Bochdalek hernisi mevcuttu. Ameliyattan sonra 5 ila 19. günler arasında tüm hastalar taburcu edildi. Hastanede ortalama kalış süresi 9.8 gündü (dağılım 7-23 gün). Hastalar altı ay ila iki yıl süreyle takip edildi ve anlamlı bir solunum veya gastrointestinal yakınma kaydedilmedi. Morgagni hernisi olan üç olguda da herhangi bir sorun tespit edilmedi.

Sonuç: Doğuştan olan diyafram hernileri, çoğunlukla neonatal dönemde görülen patolojiler olup, sonrasında bu hernilerin tanısı güçleşebilir. Takip edilen ve deneyimli uzmanlar tarafından klinik olarak tedavi edilen olgularda mortalite ve morbidite oranları düşüktür.

Anahtar sözcükler: Edinilmiş; doğuştan; diyafram; herniasyon.

Congenital diaphragmatic hernias are seen in every 2000-3000 births and constitute 8% of all major congenital anomalies.[1]

The shifting of abdominal organs into the thorax can lead to life-threatening respiratory problems. This shift can also cause significant problems in lung development if it begins in utero.[2] Bochdalek hernias comprise approximately 85% of all congenital diaphragmatic hernias. Morgagni hernias develop from a defect in Larrey's space and are seen with the same prevalence (5%) as hiatal hernias in large series.

Mortality rates are high in congenital diaphragmatic hernia cases presenting with severe respiratory insufficiency in the early neonatal period.[3] Diagnosis

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made after the neonatal period is rare, and complaints on admission are different from those of early stage cases. The prognosis of late-presenting cases is better.[4,5] These patients are usually diagnosed as the result of clinical investigations performed because of recurrent respiratory complaints and obscure abdominal pain. Severe respiratory and circulatory problems due to the mediastinal shift sometimes accompany these hernias, and severe symptoms related to strangulation or trapping of the intraabdominal organs are also seen.

In our study, we aimed to analyze lately diagnosed patients with diaphragmatic hernias who had specific clinical features, and interesting clinical and radiological manifestations.

PATIENTS AND METHODS

A total of eight diaphragmatic hernia patients, of whom three were adults and five were children (4 males, 4 females; mean age 12.4±24 years; range 9 months to 28 years), who were diagnosed in the late period and who were operated on in the Thoracic Surgery Clinics of the Van Training and Research Hospital and the Yüzüncü Yıl University Medical Faculty Hospital between January 2006 and August 2010 were evaluated retrospectively.

The results of the preoperative and postoperative first and sixth-month respiratory function tests of the three patients diagnosed in adulthood were compared.

The demographic features of the patients are shown in Table 1. Plain and lateral chest X-rays, thoracic and upper abdominal tomography, barium esophagogastrography, abdominal ultrasonography, magnetic resonance imaging (MRI), and posteroanterior abdominal X-rays were used for the preoperative diagnosis.

Our pediatric cases were diagnosed late after the neonatal period. The diagnosis is made in three patients in adulthood. Five patients had right-sided congenital Morgagni hernias, one patient had a hiatal hernia, and two patients had left-sided Bochdalek hernias (Table 1).

Frequent cough, fever, wheezing, recurrent pneumonia episodes, abdominal distention, constipation, abdominal pain, vomiting, and palpitations constituted the main complaints. The percentage of pediatric patients was low, and they had growth retardation (Table 1). Three children were referred to our clinic due to recurrent pulmonary infections and suspicious images on chest X-rays. A 28-year-old obese patient was referred to our clinic due to detection of intestines reaching the medial-upper zones of the right hemithorax on chest X-ray taken for gradually increasing respiratory distress.

RESULTS

On physical examination of the patients, there was abdominal distention, audible intestinal sounds in hemithorax, and loss or decrease of respiratory sounds in related regions on auscultation. Pallor related to anemia was also present along with growth retardation in the pediatric patients.

The average duration of hospital stay was 9.8±2.5 days (range 7 to 23 days). Diagnosis was supported with preoperative chest X-rays, thoracic CT, barium radiographs, and thoracic MRI.

The diameters of the diaphragm defects varied between 3 and 5 cm based on intraoperative assessment. The most commonly herniated organs were the colon, spleen, small intestine, liver, stomach, omentum, and appendix.

The left lower lobe was completely hypoplastic in one of the patients with Bochdalek hernia (Figure 1). The large and small intestines, the stomach, the omentum, and the appendix were herniated in this case (Figure 2). This subject underwent a left lower lobectomy along with an intrathoracic splenectomy and an appendectomy. We performed a left lower lobectomy in one patient who had a Bochdalek hernia based on the decision that the hypoplastic lung could not be expanded. We performed a splenectomy in the same patient because of hemorrhaging which occurred while separating intense intraoperative adhesions. The colon, the stomach, and part of the peritoneum were herniated in the other Bochdalek hernia case. Atelectasis was present in the left lower lobe in this case. Lung tissue was observed to have expanded completely after ventilation was provided following the placement of the intrathoracic organs into the abdomen. Resection was not performed for this subject.

The diaphragm was closed with interrupted non-absorbable sutures after reduction of the herniated organs by a laparotomy through a supra-umbilical median incision in four out of five of the cases with Morgagni hernias (Figure 3). In their long-term (two-year) follow-up, no complaints or radiological abnormalities were detected in our patients.

The hernia sac was extremely adherent to the intrathoracic organs in three out of the five Morgagni hernia cases. Thus, the sac could not be removed, and primary diaphragmatic repair was performed. The sac was removed subtotally in another Morgagni hernia case. Lacerations in the lower lobe of the lung were

Table 1. Demographic and clinical features of the patients

Patients	Age	Sex	Hernia type	Hernia location	Symptoms	Herniated organs	Surgical intervention
1	9 months	Female	Morgagni	Right anterolateral	Frequent pulmonary infection Constipation Vomiting Cyanosis	Omentum Colon	Laparotomy Reduction Diaphragm repair
2	18 months	Female	Morgagni	Right anterolateral	Constipation Vomiting Frequent pulmonary infection	Omentum Liver Colon Peritoneum	Laparotomy Reduction Diaphragm repair
3	2 years	Male	Morgagni	Right anterolateral	Frequent pulmonary infection Vomiting Distention Constipation	Colon Peritoneum Omentum	Laparotomy Reduction Hernia repair
4	5 years	Male	Morgagni	Right anterolateral	Frequent pulmonary infection Constipation Vomiting Abdominal pain Distention	Colon Stomach Small intestine Peritoneum Omentum	Laparotomy Reduction Removal of hernia sac Defect repair
5	13 years	Male	Hiatal	Hiatal	Vomiting Growth retardation Dyspnea Chest pain	Stomach Omentum Peritoneum	Laparotomy Right thoracotomy Reduction Diaphragm repair
6	16 years	Female	Bochdalek	Left posterolateral	Abdominal pain Distention Vomiting (projectile) Dyspnea Palpitations	Spleen Colon Small intestine Appendix Stomach Omentum Peritoneum	Left thoracotomy Splenectomy Appendectomy Partial peritoneum resection Left lower lobectomy
7	22 years	Male	Bochdalek	Left posterolateral	Abdominal pain Chest pain Dyspnea	Spleen Small intestine Colon Peritoneum Omentum	Left thoracotomy Reduction Diaphragm repair
8	28 years	Female	Morgagni	Left posterolateral	Dyspnea Distention Abdominal pain Chest pain Vomiting (Projectile)	Colon Small intestine Liver Stomach Peritoneum	Right thoracotomy Reduction Diaphragm repair

primarily repaired. The postoperative air leak ceased after six days of follow-up. The control chest X-rays of these patients were evaluated as normal in the postoperative follow-up period.

In a case with a hiatal hernia and in the adult patients with Morgagni hernia who underwent a thoracotomy, the diaphragm was sutured and closed after reduction of the herniated organs, and adherences were lysed following complete exploration, including a laparotomy, to prevent organ injury due to intensive organ adherences. The omentum majus was excised due to the development of necrosis in one subject with a Morgagni hernia.

The patient with the hiatal hernia was a 12-year-old elementary school student. The patient had a failure to thrive, hypoproteinemia, and anemia and was referred to our clinic due to a hydatid cyst. This patient had continuously received symptomatic treatments during the four to five years prior to being referred to our clinic,

Figure 1. Hypoplasia of the left lower lobe (Bochdalek hernia).

and no further investigations were performed. The well-shaped, crescent image consistent with a perforated hydatid cyst on his first chest X-ray performed in our clinic preoperatively was interpreted as a cystic mass lesion. However, on subsequent plain and lateral chest X-rays, the image was seen to have a leveled appearance, and the lesion was determined to be empty. The patient did not have a history of cyst perforation when he was hospitalized in the clinic or before he was referred to our clinic. Tomography was planned, and an upper gastrointestinal tract X-ray with oral contrast agent was obtained since a diaphragmatic hernia was suspected (Figure 4). Intraoperatively, the stomach was found to have completely herniated through the hiatus and had become localized in the posterior mediastinum. The stomach could not be reduced by opening the hernia sac because of extensive adhesions on the abdominal side, so it was reduced through a laparotomy. The dilated hiatal area was repaired from the thorax. In this patient, hematemesis and severe abdominal pain occurred in the fourth postoperative day. Ulcers in the stomach and lower esophagus and an erosive luminal structure were detected on endoscopy, and the condition was diagnosed to have gastric ulcer and esophagitis. The patient immediately began to have anti-ulcer treatment and gastrokinetics. The complaints improved after this medical treatment. The patient did not experience a relapse of these problems during follow-up.

Our three adult patients had dyspnea preoperatively. The 28-year-old obese patient was orthopneic. Respiratory function tests and partial blood gas pressure were found to have significantly improved at the first and sixth postoperative months compared to the preoperative values. Our first case's predicted preoperative forced expiratory volume in one second (FEV1) was 45%, and the postoperative first- and sixth-month predicted FEV1s in this patient were 55% and 75%, respectively. In the second case, the preoperative FEV1 was 47%, and the predicted postoperative FEV1 first and sixth month values were 50% and 52%, of predicted respectively. In the third case, the preoperative predicted FEV1 was 46%, and the postoperative preticted FEV1s measured in the first and sixth months were 56% and 58%, respectively.

Our patients were discharged between the 5th and 19th postoperative days, with the patient with the hiatal hernia being discharged in the 19th postoperative day. The patients were followed up from six months to two years

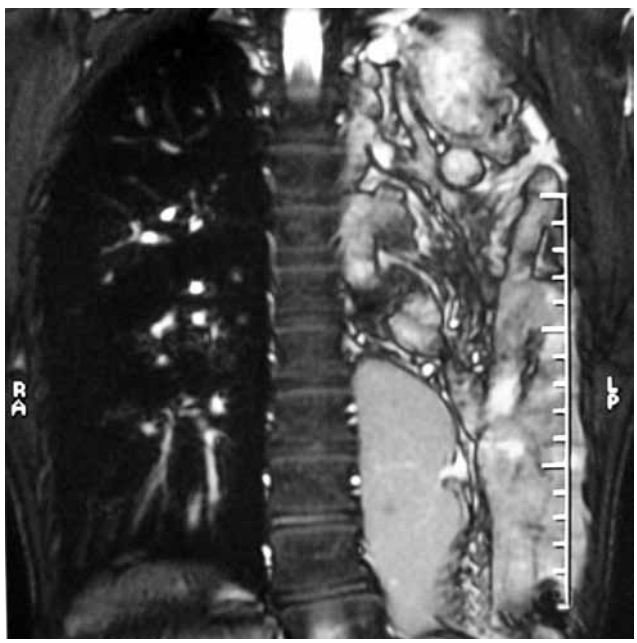

Figure 2. Colon and small intestine, stomach, omentum, and appendix herniation (Bochdalek hernia).

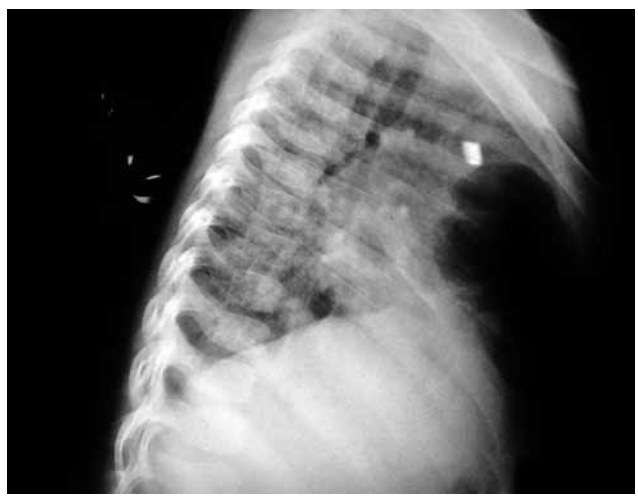

Figure 3. Morgagni hernia (lateral X-ray).

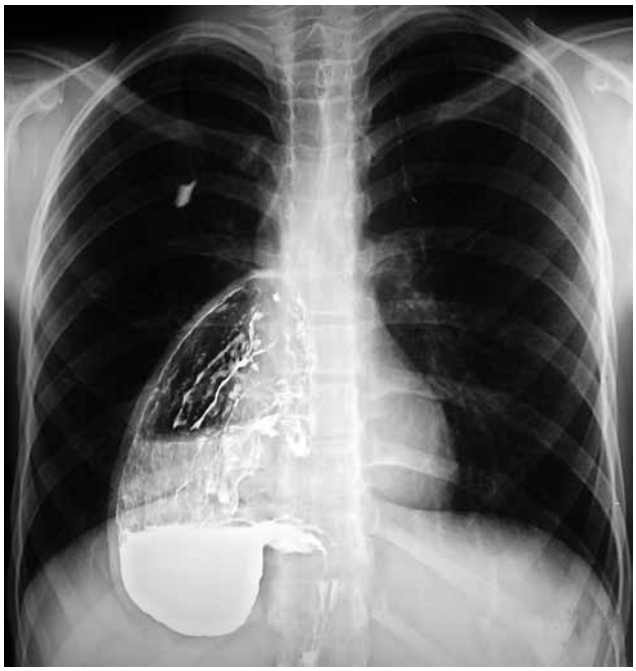

Figure 4. Herniation of the stomach (barium radiography).

after being discharged, and no significant respiratory or gastrointestinal complaints were recorded. No problems were detected in the three patients with Morgagni hernias whose hernia sacs had not been removed, and no deaths occurred in any of our patients.

DISCUSSION

Although Bochdalek hernias are more prevalent than Morgagni hernias among congenital diaphragmatic hernias, Morgagni hernias were found to be more prevalent in terms of long-term occurrence in our study (62.5%). Severe hypoplasia in the left lower lobe was observed in one of the two patients who were operated on with a diagnosis of a Bochdalek hernia, and a lower lobectomy was performed. Compression atelectasis was present in the lower lobe in one of other patients. This patient did not undergo resection, and no postoperative pulmonary problems were observed.

The prevalence of diagnosis in the neonatal period is higher, and mortality rates are high. The number of cases diagnosed after the neonatal period is low, and the prognosis for patients with late diagnosis is significantly better. Approximately 10% of all congenital diaphragmatic hernias are diagnosed after the neonatal period, and these cases can be encountered in any period of life.[4-6]

The prevalence has been reported to be equal in both sexes in previous studies.[6] The female-male ratio was found to be one in our study as well.

In patients who are diagnosed in the late period, the diagnosis can be troublesome and delayed.[4,5,7] Respiratory symptoms are usually predominant in early childhood whereas gastrointestinal complaints are predominant in late childhood in subjects with congenital diaphragmatic hernias diagnosed in the late period.[8,9] Gastrointestinal complaints were also present in our study, and recurrent pulmonary infections were predominant in five patients. Although palpitations and dyspnea were predominant in the remaining three cases, abdominal symptoms were also present. Cardiorespiratory complaints, such as palpitations and dyspnea, were also present along with projectile vomiting, severe abdominal pain, abdominal distention, and constipation in the other subject.

Herniation was located in the anterior diaphragm in five cases and posterolaterally in two cases; a hiatal hernia was present in one case. This distribution is consistent with the percentages reported in the literature which show that diaphragmatic hernias are predominantly located retrosternally and on the right. The prognosis has been reported to be better in these cases.[9-11] One of the complaints on admission was dyspnea in three late-diagnosed congenital hernia patients. Preoperative respiratory function tests and blood gas analysis were done preoperatively in these cases. These parameters were studied in the controls at the first and sixth postoperative months, and respiratory problems improved significantly.

Conditions such as bronchogenic cysts, cystic adenomatoid malformation, and pneumothorax should be excluded in the differential diagnosis of late-onset congenital diaphragmatic hernias. Chest X-rays performed following insertion of a nasogastric tube are beneficial in making the diagnosis when gastrothorax is suspected. Thoracic tomography, MRI, and ultrasonography in the neonatal and prenatal period can also be beneficial in these cases.[6] A hernia was suspected in the 12-year-old child with a hiatal hernia during the course of the preoperative work-up, and thereafter, tomography and barium X-rays were utilized for a definitive diagnosis.

Morgagni hernias constituted the majority of our cases. All pediatric patients with this diagnosis and symptomatic adults should undergo surgery. Surgical treatment is controversial for asymptomatic adult patients. Surgery should not be delayed if the symptoms (abdominal pain, constipation) are frequent in this group of patients.[12] A hernia sac is present in 90% of Morgagni hernia patients. In the literature, it has been reported that the sac was not removed in more than half of the evaluated 140 cases.[13] Kuster et al.[14]

recommended leaving the hernia sac in place due to the risk of massive pneumomediastinum which could lead to respiratory and circulatory complications. In one of our Morgagni hernia cases, the sac could be partially separated from the neighboring organs, and prolonged air leak occurred in this patient in the postoperative period. The sac was not removed in the other cases, and in two of these cases, 2x1 cm of asymptomatic pouch was left in the paracardiac area. No pathologies were done in this particular case, and no significant pathologies were observed during the long-term follow-up of the other cases.

Diaphragmatic hernia defects can be repaired by either the transthoracic or the abdominal approach. The thoracoscopic approach, however, gives better access to the sac, allowing for safer dissection of pericardial and pleural adhesions.[15,16] According to Kılıç et al.,[16] the transthoracic approach provides sufficient exposure, easy repair of hernia sacs, and an acceptable morbidity when compared with the transabdominal approach. The transthoracic approach facilitates the release of pericardial adhesions. Yavuz et al.[17] suggested that the hernial diaphragmatic defect should be repaired either by primary suturing or by the use of prosthetic mesh. In large diaphragmatic hernia cases, using mesh is not only facilitates to reconstruct the dome of the diaphragm but also prevents undue tension on the repair and recurrences. In adults, prosthetic mesh repair is preferred. In our patients, the diaphragm was closed with interrupted non-absorbable sutures after reduction of the herniated organs by a laparotomy through a supra-umbilical median incision in four out of the five cases with Morgagni hernias. Because primary diaphragmatic repair could be completed in all cases, we did not use mesh in any patient. In the series, no complications were observed postoperatively. During the long-term (two-year) follow-up, no complaints were reported, and no significant radiological abnormalities were detected.

Recently, laparoscopic repair of Morgagni hernias has been introduced and has gained wide acceptance. According to Yavuz et al.,[17] this type of repair is a safe, simple and reliable procedure that has all of the advantages of minimally invasive surgery. Laparoscopic repair reduces postoperative pain, shortens recuperation, and reduces the severity of wound complications in these patients.[18]

We favored the transthoracic procedure in our pediatric cases that had hernia sacs; however, we performed laparotomies as a general rule.

In conclusion, congenital diaphragmatic hernias are pathologies that can be seen in the neonatal period, and the diagnosis may be difficult. Recurrent pulmonary infections, persistent cardiorespiratory problems, and frequent abdominal complaints are findings that can lead to a diagnosis. Mortality and morbidity rates can be quite low in cases that are followed and treated in clinics with experienced and concerned specialists.

Declaration of conflicting interests

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