Clinical presentation and operative repair of hernia of Morgagni

T P F Loong, H M Kocher

A 77 year old woman who presented with an incarcerated hernia of Morgagni was successfully treated without complications. A Medline search (1996 to date) along with cross referencing was done to quantify the number of acute presentations in adults compared to children. Different investigating modalities—for example, lateral chest and abdominal radiography, contrast studies or, in difficult cases, computed tomography or magnetic resonance imaging—can be used to diagnose hernia of Morgagni. The favoured method of repair—laparotomy or laparoscopy—is also discussed. A total of 47 case reports on children and 93 case reports on adults were found. Fourteen percent of children (seven out of 47) presented acutely compared with 12% of adults (12 out of 93). Repair at laparotomy was the method of choice but if uncertain, laparoscopy would be a useful diagnostic tool before attempted repair. Laparoscopic repair was favoured in adults especially in non-acute cases.

Hernia of Morgagni is the most rare of the four types of congenital diaphragmatic hernia (2%–3% of all cases). In adults, it commonly presents with non-specific symptoms—for example, excess flatulence and indigestion. In severe cases, it might present with symptoms of bowel obstruction or strangulation. In children, the majority present with repeated chest infection; rarely it might present in the neonatal period as acute respiratory distress syndrome. More than half are detected when patients are being investigated for unrelated problems. It is diagnosed with a lateral chest radiograph and confirmed with a barium enema or computed tomogram. Reports in the literature describe repair by the transabdominal or trans-thoracic approach with or without a mesh. In recent years there has been a trend towards repair by laparoscopy.

CASE REPORT

A 77 year old Jehovah’s Witness presented to the accident and emergency department with a 10 day history of worsening abdominal pain, distension, vomiting, and constipation. She had presented to casualty six weeks previously with abdominal pain only and was presumed to have constipation and was treated accordingly.

She had no previous bowel surgery. A barium enema six years before this admission (1996) showed mild diverticular disease. She took aspirin for her previous stroke and lansoprazole for gastritis and severe reflux oesophagitis. She had no family history of bowel malignancy.

On examination, she was dehydrated but stable. Her abdomen was distended with two tender tympanic masses on the right side. Abdominal radiography showed dilated loops of large bowel, measuring about 18 cm in diameter. She was operated on with presumptive diagnosis of caecal volvulus.

At operation, an incarcerated knuckle of the transverse colon was found in the hernia of foramen of Morgagni. It was easily reduced and repaired without a mesh. She recovered uneventfully.

METHODS

A Medline search (1996 to date) along with cross referencing was done to quantify the number of acute presentation in adults compared to children. Patients were subdivided into acute, subacute, chronic, or asymptomatic presentations. Acute presentations were those where patients presented with less than a week of symptoms, subacute where patients presented up to six months, and chronic patients had symptoms for more than six months. We excluded any case reports that had no clear description of surgical repair. Case reports in a foreign language are briefly mentioned and included in the references. The approach for repair was laparotomy, thoracotomy, laparoscopy, or other (as stated in tables 1 and 2).

The results of the Medline search are shown in tables 1 and 2.

LITERATURE REVIEW

Hernia of Morgagni was first described by Giovanni Battista Morgagni, an Italian anatomist and pathologist in 1769, while performing a postmortem examination on a patient who died of a head injury. Hernia of Morgagni is located just posterolateral to the sternum. It has also been called retrosternal, para-sternal, substernal, and substernal. It is caused by a congenital defect in the fusion of septum transverses of the diaphragm and the costal arches. This weakness in the diaphragm later would be stretched by rapid rise in intraperitoneal pressure, giving rise to a hernia. Lev-Chelouche et al mentioned that it is for this reason that hernia of Morgagni is usually not discovered in children. It can occur on either side of the sternum through a muscle-free triangular space called the Larrey space, although it is more common on the right. In rare cases, the hernia can be bilateral.

See end of article for authors’ affiliations

Correspondence to:
Mr Hemant M Kocher,
Tumour Biology Laboratory, Bart’s and the London Queen Mary’s School of Medicine and Dentistry, John Vane Science Centre, Charterhouse Square, London EC1M 6BQ, UK; hemant.kocher@kcl.ac.uk

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Table 1: Case reports on children

<table>
<thead>
<tr>
<th>Author</th>
<th>No of cases</th>
<th>Presentation (No of cases)</th>
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<tr>
<td>Foy et al., 1992</td>
<td>1</td>
<td>Subacute</td>
<td>Laparotomy</td>
</tr>
<tr>
<td>Sinclair and Klein, 1993</td>
<td>1</td>
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<td>Bentley and Lister, 1965</td>
<td>3</td>
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<td>Sanhaji et al., 1994</td>
<td>2</td>
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<tr>
<td>Machnouchi et al., 2000</td>
<td>9</td>
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<td>Sayul et al., 2000</td>
<td>1</td>
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<td>Nurradi et al., 2000</td>
<td>1</td>
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<td>Laparotomy (2)</td>
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<td>Singh et al., 2001</td>
<td>2</td>
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<td>Parmar et al., 2001</td>
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<tr>
<td>Lima et al., 2001</td>
<td>2</td>
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<td>Laparoscopy (2)</td>
</tr>
<tr>
<td>Al-Salem et al., 2002</td>
<td>15</td>
<td>Subacute (13)</td>
<td>Laparotomy (14)</td>
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<td>Kulaylat et al., 2003</td>
<td>1</td>
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<td>Transabdominal</td>
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Total cases in children = 47. Presentation: chronic, 15; subacute, 19; acute, 7; asymptomatic, 6. Repair: laparotomy, 40; laparoscopy, 4; thoracotomy, 3.

Like the first patient described, the majority of hernias of Morgagni are diagnosed late because patients can be asymptomatic or present with non-specific respiratory and gastrointestinal symptoms and signs. Before presenting with acute intestinal obstruction, our patient had been seen for the last three years for symptoms of indigestion and bloating. She may have developed an uncomplicated hernia of Morgagni then or in the interim and this was therefore missed on barium enema at that time. Diagnosis can be difficult and a missed diagnosis can lead to life threatening complications such as obstruction or strangulation. In our literature review, hernia of Morgagni presents itself more acutely (seven cases, 14%) and subacutely in children (19 cases, 40%). In recent years there has been a rise in the number of cases reported, with an approximate total of 200 cases in the last 10 years. This may be due to greater awareness of its diagnosis and because of early treatment to prevent any complications. However, hernia of Morgagni may be more frequent than the literature suggests since most cases are asymptomatic.

Diagnosis is confirmed by plain chest radiographs and contrast films. Hernia of Morgagni usually presents with recurrent chest infections in children (55%) and lateral chest radiographs are usually always conclusive. Screening may apply to children with increased risk associated anomalies and familial forms of congenital diaphragmatic hernias (from 34% to 50%). Patients with Down’s syndrome (five cases) have increased risk of hernia of Morgagni. Obese patients may develop it later in life and sometimes it may follow trauma.

Depending on the contents of the hernia—omentum, stomach, small intestine, or liver—it can appear differently on chest radiography and the diagnosis can be missed. For example, if omentum is present in the sac, a solid paracardiac shadow will appear on the chest radiograph. Differential diagnosis would be an intrathoracic tumour, atelectasis, pneumonia, or pericardial cyst. This might affect the decision to operate and the type of operation carried out—that is, the transabdominal or transthoracic approach. Contrast examination—for example, barium enemases carried out for gastrointestinal symptoms can also be absolutely normal. Computed tomography can be considered to be an accurate, non-invasive method of diagnosing hernia of Morgagni. It can help establish a diagnosis if, as in some cases, the hernia

Table 2: Case reports on adults

<table>
<thead>
<tr>
<th>Author</th>
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<th>Presentation (No of cases)</th>
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<td>Chin and</td>
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<td>Shackelford et al., 1973</td>
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<tr>
<td>Catalano et al., 1972</td>
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<td>Missen, 1973</td>
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<tr>
<td>Paris et al., 1973</td>
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<td>1</td>
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<td>Fernandez and</td>
<td>1</td>
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<td>Oteyza, 1996</td>
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<td>Onita et al., 1997</td>
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<td>Lorasa et al., 1999</td>
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<td>Ramachandran et al., 1999</td>
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<td>and Vijay, 1999</td>
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<td>White et al., 2002</td>
<td>1</td>
<td>Chronic</td>
<td>Laparoscopy</td>
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</table>

Total cases in adults = 93. Presentation: chronic, 13; subacute, 35; acute, 12; asymptomatic, 33. Repair: laparotomy, 21; laparoscopy, 21; thoracotomy, 26; other, 2; not repaired, 23.
sac is empty or contains omentum or part of the liver. But as described by Fagelman et al the computed tomogram did not confirm the diagnosis after the chest radiograph as the presence of gas within the lesion was variable: the bowel was sliding in and out of the defect. This might make diagnosis difficult or confusing. Other investigations such as magnetic resonance imaging (MRI) and radionucleotide liver scan may help with diagnosis but the cost is difficult to justify. In our review, there were 3 cases that were diagnosed with MRI. Collie et al demonstrated with MRI a herniation of liver through hernia of Morgagni on a patient who presented with increasing shortness of breath and exertional angina.11

In our opinion, we feel a simple chest radiograph is most likely to reveal an asymptomatic hernia of Morgagni when done for unrelated problems. However, if suspected clinically, computed tomography would be the preferred imaging modality to confirm the diagnosis in adults and children. Another option, a less expensive one, would be a barium enema for adults. When investigations are non-diagnostic, confirmation by laparoscopy may be needed. Follow up after operative repair can be done with a chest radiograph at three months and one year.

The need for surgery depends on presentation. Although the majority of these hernias are asymptomatic, repair is recommended to avoid future complications. Operation is indicated when the colon is in the sac, as there is a high risk of obstruction. If the hernia is small or if it contains omentum only, operation is indicated when symptoms are recurrent and bothersome. Treatment options include transabdominal or transthoracic repair.12

The transabdominal approach was favoured when the diagnosis was certain as it allows easier reduction of the hernia, especially for bilateral hernias. Furthermore, abdominal viscera within the hernia can be easily pulled down to their normal location in the abdomen. The sac can then be withdrawn and resected along the margins of the defect if need be. In our patient, we left the sac and part of the omentum in situ, closed the defect with interrupted, nylon sutures and reinforced it with a polypropylene mesh. Paris et al suggested a preperitoneal subxiphoid approach because it allows freeing of the pleural adhesions to the sac by an extrapolaparotomy route.13 This avoids the large incision of a laparotomy.

Chin et al advise a transthoracic approach as it provides a wide exposure and easy repair of the hernia sac.14 This is also advocated by Kilic et al who performed thorocotomies on 16 patients, all with uneventful recoveries and no recurrence of symptoms.15 However, Bentley and Lister describe a patient who had to undergo a second operation for intestinal obstruction after the initial thoracic procedure failed to diagnose bilateral hernia of Morgagni.16 Thoracotomy was indicated when the diagnosis was uncertain.

The first laparoscopic repair was reported by Kuster et al in 1992.17 Since then, there have been 25 cases reported: 21 adults (22%) and four children (8%). Laparoscopy is an excellent way to confirm diagnosis and to repair non-complicated hernia of Morgagni. The hernia sac can be easily viewed through the laparoscope. The hernia contents can then be easily reduced once the peritoneum at the perimeter of the defect is incised. The sac is usually not removed and the defect is closed with silk sutures and reinforced with a mesh stapled onto the diaphragm. Other advantages of laparoscopic repair are reduction in trauma, a faster recovery and faster return to normal diet and activity.18 It is also a safe and useful procedure to perform on children, especially when computed tomography is non-diagnostic. Table 3 highlights the complications that were encountered with each approach.

Things to consider during the operation are whether to remove the sac and whether to use a mesh. Almost 90% of cases of hernia of Morgagni have a sac. In our view, in more than half of the cases reported, the sac was not removed. As described in Kuster et al it was recommended not to remove the sac as this may result in massive pneumomediastinum with potential respiratory and circulatory complications.17 Rau et al had a different approach and removed the sac to avoid leaving a loculated space-occupying lesion in the chest that might result in recurrence or cyst formation.18 However there is no available literature to demonstrate the reasons for either procedure. Ramachandran et al left the sac alone and repeat computed tomography a month later showed almost complete disappearance of the sac.19 We feel that removing the sac would depend on the skill of the surgeon and the presentation of the patient. The use of a prosthetic mesh is becoming more popular. If the defect is small, it can be easily sutured as done in our patient. A mesh overlapping the edges of the defect can be easily manipulated with laparoscopic instruments and it provides a good tension-free repair.20 No recurrence or complications have been seen with using a mesh.

CONCLUSION

Hernia of Morgagni is rare in both adults and children. In our literature review, acute presentation occurred more frequently in children. This may be because more cases are being detected due to greater awareness. Most asymptomatic cases were found in adults by chest radiography for unrelated problems. Diagnosis can be confirmed with contrast studies or laparoscopy. In adults presenting more acutely, the transabdominal approach would be the first line method of repair, reducing the hernia, leaving the sac alone, and using a prosthetic mesh. In non-acute cases, laparoscopic repair would be the first choice in children and adults as well being a useful diagnostic tool.

Table 3 Complications and failures

<table>
<thead>
<tr>
<th>Approach</th>
<th>No of cases</th>
<th>Complications (No of cases)</th>
<th>Failure</th>
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<td>Wound infection (1)</td>
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<td>Deep vein thrombosis (1)</td>
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<td></td>
<td></td>
<td>Pulmonary embolism (1)</td>
<td></td>
</tr>
<tr>
<td>Laparoscopy</td>
<td>25</td>
<td>None</td>
<td>Failure to reduce contents: progressed to open surgery</td>
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<tr>
<td>Thoracotomy</td>
<td>30</td>
<td>Pneumonia + sepsis (1)</td>
<td>Bowel obstruction: emergency laparotomy. Death via aspiration</td>
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</table>

References


