Thoracic Lipoma within Morgagni's Hernia

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Morgagni's hernia is a rare subtype of congenital diaphragmatic hernia. It is rarely diagnosed in adulthood. It often presents with non-specific symptoms of obstruction or incarceration or present as cardiopulmonary or gastric symptoms. Usually, the contents include abdominal organs or omentum. The diagnosis could be difficult as they are often confused with different common pathologies.

We report a case of a forty-eight-year-old female who presented with non-specific symptoms of fatigue, cough and dyspnea. Further assessment revealed a mediastinal mass, consistent with a mediastinal lipoma. During thoracotomy, a lipoma was found within a hernial sac located retrosternal, consistent with Morgagni's hernia. The lipoma was removed and the defect was closed. Postoperatively, the patient had an uneventful recovery.

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Lipomas are one of the most common benign tumors of mature adipose tissue¹. They are usually located superficially, in the subcutaneous tissue. Rarely, the tumors are deep-seated, where they could grow to large sizes insidiously². The deep-seated lipomas are usually excised as they may either resemble malignant tumors or transform into malignant liposarcomas^{3,4}.

Congenital diaphragmatic hernia (CDH) is a rare congenital abnormality, which occurs in approximately 1 of 2000 to 5000 live births⁵. They are classified into posterolateral (Bochdalek) hernia, retrosternal (Morgagni) hernia, hiatal hernia and septum transversum defect⁶. Bochdalek hernias are the majority of CDH 95%, the remaining is around 2% each⁷⁻⁸.

The aim of this report is to present a case of a thoracic lipoma within a Morgagni's hernia.

THE CASE

A forty-eight-year-old female presented with a two-day history of dyspnea, fatigue and a mild productive cough of yellow sputum. She had a right-sided homogenous opacity adjacent



Figure 1: Chest X-Ray on Presentation

to the heart on posterior-anterior (PA) chest X-ray, see figure 1. Respiratory examination revealed no abnormalities. ECG was consistent with sinus tachycardia. She had a history of hypothyroidism (currently on Levothyroxine). Family history was negative for any malignancies.

Laboratory investigations including complete blood counts, urea and electrolytes, renal function tests, liver function tests and cardiac enzymes revealed no abnormalities.



Figure 2 (A): CT Scan - Sagittal Reformatted CT Scan, The Fat-Containing Mass is Almost Entirely Within the Mediastinum



Figure 2 (B): Coronal CT Scan

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A high-resolution CT Scan of the chest and upper abdomen revealed a well-defined, irregularly shaped homogenously enhancing hypodense mediastinal lesion. It measured approximately 13x7 cm projecting mainly into the right paracardiac space with no signs of invasion and no lymphadenopathy. The findings were suggestive of a mediastinal lipoma. A limited echocardiograph was performed, which revealed a hyperdynamic left ventricle with no significant regional wall motion abnormalities, with an ejection fraction of 55 percent, good right ventricular function and otherwise no abnormalities.

A three-port video-assisted thoracoscopy (VATS) was performed, which was subsequently converted to a right thoracotomy and excision of a sessile lipoma measuring 10x8x4 cm from within the hernial sac, originating anteromedially from the diaphragm. After removal of the lipoma, it was noted that omentum was protruding from a small diaphragmatic retrosternal hernia, a Morgagni's hernia, which was closed during the procedure. Pathological analysis of the excised mass confirmed the fatty tissue encased in a hernial sac, which tested negative for malignancy. The patient had an uneventful recovery.

To our knowledge, this is the first case of a lipoma within a Morgagni's hernia presented in the literature.

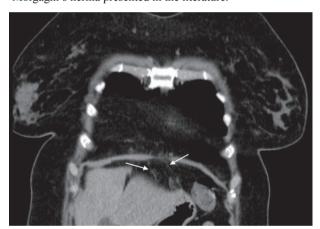


Figure 3: Sagittal Reformatted CT Scan, Curvilinear Densities Originating from the Abdomen, Most Likely Representing Blood Vessels (White Arrows)

DISCUSSION

Retrosternal or Morgagni's hernia is a rare entity first described in 1769 by Giovanni Morgagni⁹. Morgagni's hernia constitutes approximately 2% of all CDH, which is estimated to be 1 in every 2000 to 5000 births^{5,7-8}.

Morgagni's hernias in adults are more common in females with an average age of presentation of 42 to 53 years^{10,11}. It is due to failure of closure of the pars sternalis with the seventh costochondral arch, leading to an anterior medial septum transversum defect, known as Larrey's space^{8,12-13}. About 91% of retrosternal hernias are right-sided, 5% are left-sided and the remaining 4% are bilateral¹¹. A hypothesis proposed for the increased occurrence on the right side is the pericardial attachment on the left side, which strengthens the diaphragm on the left side¹⁴.

Morgagni's hernia predisposing factors include pregnancy, trauma, obesity, chronic constipation, and chronic cough, most likely due to the raised intra-abdominal pressure¹⁵⁻¹⁸. It

is also associated with Down's syndrome, Turner's syndrome, Noonan's syndrome, Prader-Willi syndrome, Teratology of Fallot, connective tissue disorders, omphaloceles, ventricular septal defects, dextrocardia, chest wall deformities and genitourinary abnormalities¹⁹⁻²⁸.

The most striking feature in our case is the presence of a lipoma within the Morgagni's hernial sac. Lipomas are the most common benign tumors of adulthood¹. They tend to be in more superficial tissues and are rarely deep-seated, where they tend to be larger since they are detected late^{2,29}. They are subclassified according to their histologic features, which include conventional lipoma, fibrolipoma, angiolipoma, fusiform cell lipoma, myelolipoma and pleomorphic lipoma³⁰. In our case, histopathological examination revealed conventional lipoma, with mature adipose tissues and no features suggestive of malignancy.

Morgagni's hernias have a wide spectrum of presentation. Pain and pulmonary symptoms such as dyspnea, cough, and shortness of breath were the most commonly reported symptoms¹¹. Other symptoms include bowel obstruction, gastroesophageal reflux symptoms, dysphagia, vomiting, post-prandial distention, constipation and diarrhea^{11,31}. Around 28% of the patients are asymptomatic¹¹. Due to this wide spectrum of non-specific symptoms ranging from asymptomatic to bowel obstruction, most of the cases are diagnosed late³².

The most common contents of the hernia are omentum and colon, however, herniation of the stomach, small bowel and liver have been reported.¹¹

The diagnosis of Morgagni hernias is mainly radiological. Eren et al reviewed the different radiological modalities used to assess diaphragmatic hernias⁶. The initial imaging modality is plain chest radiograph. The hernia may mimic pneumonia, pneumatocele, abscess, lung cancer, liposarcoma, thymoma, hydatid cyst, teratoma or neurofibroma; in our case, it was mediastinal lipoma³³. The presentation is usually right-sided. Imaging may appear normal in cases of intermittent herniation, where the herniated content returns to the abdominal cavity⁶. Moreover, ultrasound (US) may be useful in the assessment of diaphragmatic integrity and herniated organs; Doppler ultrasound could assess vessels within the hernia⁶. It is also useful in discerning the small blood flow in benign lipomas compared to liposarcomas³⁴. A CT scan is useful in demonstrating diaphragmatic discontinuity, and the herniation of intra-abdominal organs into the thoracic cavity⁶. It is particularly useful in assessing the nature of the lesion, where Hounsfield units ranging from -80 to -120 suggest the fatty composition and in defining omental vessels in lipomatous masses which are likely to be represented as curvilinear densities, as seen in figure 3^{35,36}. It also allows for better assessment before surgical interventions³⁷. MRI could be used in select cases for further assessment of soft tissue or if the diagnosis is uncertain⁶. Echocardiography is also potentially useful in assessing a Morgagni's hernia that is adjacent to the heart and possibly affecting cardiac function³⁸⁻³⁹. Eren et al concluded that CT remains the most effective modality in the diagnosis of diaphragmatic hernias due to both its ease of use and short imaging duration6.

The mainstay management of a Morgagni's hernia is surgical. Repair is performed via thoracic approach or the abdominal approach⁵. The abdominal approach can be performed by either laparoscopy or laparotomy and could be used even in incarcerated hernia^{40,41}. Laparoscopy has the benefit of shorter recovery times and better cosmetic results⁴².

The transabdominal approach permits easier reduction of herniated contents, and evaluation and repair of the intra-abdominal pathology or diaphragmatic defects¹¹. The transthoracic approach, which could be performed by thoracoscopy or thoracotomy provides a wider exposure for easier visualization and repair of the hernial sac and allows easier dissection of the hernial sac off the mediastinal and pleural structures^{11,43,44}.

Removal of the hernial sac is disputed in the literature. Some authors support removal to avoid recurrence or cyst formation, while others are against the removal of the hernial sac to minimize the risk of a pneumomediastinum^{15,45-50}.

It is generally agreed that even asymptomatic cases should be treated surgically due to the risk of further herniation and possible incarceration^{20,51}. Conservative treatment is reserved for those who are deemed unfit for the procedure³⁶. However, because the true prevalence of Morgagni's hernia has not been reported, it is difficult to compare surgical and conservative management¹¹.

In our case, a Video Assisted Thoracoscopic Surgery (VATS) was initially attempted. A right thoracotomy was performed after the detection and diagnosis of the size of the lipoma.

CONCLUSION

Morgagni's hernia may be easily misdiagnosed since the differential diagnosis is vast. CT scan is widely accepted as the modality of choice and surgical repair is the gold standard of management, regardless of symptoms. Laparoscopy is a safe approach with shorter hospital stay and better cosmetic results.

Written informed consent was obtained from the patient.

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