Morgagni Hernia with Partial A-V Canal Defect; A Rare Condition

Kunal*, Vishal Khante, Saket Agarwal, Deepak Kumar Satsangi

Department of Cardiothoracic & Vascular Surgery, G. B. Pant Hospital, New Delhi, India

ABSTRACT

Morgagni hernia is a rare diaphragmatic hernia usually due to congenital defects in the diaphragm. It is rarely associated with cardiac anomalies, most commonly atrial (ostium secundum) or ventricular septal defects. We report a rare case of Morgagni hernia occurring in association with partial atrio-ventricular septal defect (ostium primum), and its successful surgical correction.

Introduction

Morgagni hernia (MH) is a rare entity, usually due to congenital herniation of abdominal contents into the thoracic cavity through a retrosternal diaphragmatic defect, the Foramen of Morgagni. It is rarely traumatic. Most cases of MH occur as isolated defects, though occasionally, MH has been reported in association with other congenital malformations, including chest wall defects, intestinal malrotation and chromosomal anomalies (1-3). It is very rarely reported with congenital heart disease, most commonly atrial or ventricular septal defects (ostium secundum ASD or VSD) but it has never been reported with atrio-ventricular canal defects (AVCDs). We report one such case of MH associated with partial AVCD.

Case Report

A five-year-old male child presented to us with complaints of fatigue, frequent chest infections and poor weight gain for 2 years. There were no symptoms referable to the gastro-intestinal tract. His general physical examination was within normal limits. His chest was of normal shape and symmetry on both sides. On percussion, dullness was found in the right lower part of the chest. On auscultation, there was widely split and fixed second heart sound and a grade 2/6 systolic murmur over the mitral area. Breath sounds were decreased over the right lower chest.

Morgagni hernia (MH) is a rare diaphragmatic hernia usually due to congenital defects in the diaphragm. It is rarely associated with cardiac anomalies, most commonly atrial (ostium secundum) or ventricular septal defects. We report a rare case of Morgagni hernia occurring in association with partial atrio-ventricular septal defect (ostium primum), and its successful surgical correction.

►Implication for health policy/practice/research/medical education:

Our article is intended for the management of adult window ductus patients with severe pulmonary hypertension, a challenging rare entity for both surgeons and interventional cardiologists, where optimal method of management is controversial.

►Please cite this paper as:

space thereby confirming the diagnosis of Morgagni hernia.

Decision was taken for repair of the AVCD with concomitant repair of the diaphragmatic hernia. The thorax was opened by a vertical midline sternotomy. Pericardium was opened and cardiopulmonary bypass was initiated in routine manner by aortic and bicaval cannulation. The intracardiac repair was performed with repair of the cleft mitral valve through a right atrial incision. Cardiopulmonary bypass was weaned uneventfully. Upon completion of the cardiac procedure, the right pleural space was opened and the hernial sac and the defect were visualized (Figure 4). The sac was dissected from the pericardium, retrosternal space and the herniated part of the right lobe of liver reduced into the abdomen through the defect. Non-absorbable horizontal mattress sutures were sequentially placed through the edge of the diaphragmatic defect and into the retrosternal fascia and periosteum. The defect was obliterated as the sutures were tied. A right pleural tube was placed in addition to routine mediastinal tubes and nasogastric tube. The postoperative course was uneventful with no abdominal complications related to the procedure. Patient was discharged on postoperative day six and is doing well at three months follow-up.

Discussion

Morgagni hernia is herniation of abdominal viscera into the thoracic cavity through the Foramen of Morgagni, which are small intervals just behind the sternum on both sides at which the muscular fibers of diaphragm are deficient and are replaced by areolar tissue. Normally these foramens transmit the superior epigastric branch of internal mammary artery with some lymphatics from the abdominal wall and liver surface. In 1769, Morgagni first described this condition in an autopsy of an Italian stonemason. Other names suggested for this hernia are subcosto-sternal hernia and Larrey’s hernia (4). Embryologically, at the 3mm stage, the muscularization of diaphragm begins from myotomes that invade the mesenchyma from dorsal to ventral direction, along with concomitant fusion of sternum from above. Failure of this fusion leads to MH (5). MH comprises 3-5% of congenital diaphragmatic hernias with Bochdalek hernia constituting the majority of childhood diaphragmatic hernias (87%). Others are evagination of the diaphragm (10-12%) (6). MH occur more commonly in males and the majority of cases are seen on right side (90%), though they can be bilateral in 2% and left sided in 8% (7). The rarity of MH on the left side is attributed to the reinforcing effect of the heart and pericardium. In the paediatric age group, the presentation of MH can be variable. During infancy it can lead to acute respiratory distress indistinguishable from that of Bochdalek hernia (3). At times MH remains asymptomatic or discovered accidentally during evaluation of other unrelated conditions, as in our case. Most commonly, patients present with repeated attacks of pneumonia or vague, unspecific gastrointestinal symptoms.

On chest X-Rays, they present as mass in the anterior right or left cardiophrenic angle. Other radiological investigations that are used include ultrasound, barium enema, barium meal follow-through, CT scan and MRI. On chest X-ray herniation of bowel loops into the chest may be detected which is confirmed by a barium enema or barium meal follow-through. Barium enema is a useful investigation as colon is the most common organ to herniate in MH. However, at times the diagnosis can be difficult or delayed if the hernial sac is empty or contains omentum or part of the liver as it was in our case. In such cases Computerised Tomography scan (CT scan) proves useful in establishing the diagnosis. CT scan is also useful in demonstrating bilateral MH when the sacs are empty.

MH is often associated with other congenital anomalies such as Turner syndrome with coarctation, pectus carinatum, Prader-Willi syndrome, Cantrell’s syndrome, Noonan syndrome, omphalocele, retroperitoneal teratoma, and genitourinary anomalies (8, 9). Cardiac anomalies associated with this are ostium secundum atrial septal defect (ASD) (10), dextrocardia, ventricular septal...
defect (VSD) and anomalous pulmonary venous return, Scimitar syndrome, and coronary heart disease. Cyanosis due to a Morgagni hernia compressing the right ventricle and causing impaired diastolic filling and a right-to-left atrial level shunt, mimicking Tetralogy of Fallot presentation has also been reported. However MH has never been reported before in association with AV canal defects. We believe our case is the first report in English literature describing association of Morgagni hernia with an atrio-ventricular canal defect. This emphasises the need for complete physical evaluation in any patient of Morgagni hernia including echocardiographic examination.

For treatment of Morgagni hernia, a trans-abdominal approach is advocated as both hernial defects can be recognized and repaired. Through the trans-abdominal approach it is not only easy to reduce the hernial contents but also to correct the associated malrotation which is present in up to 25% of patients. However if associated cardiac anomaly is detected, then a trans-sternal approach is recommended.

Acknowledgement

There is no acknowledgment.

Financial Disclosure

The authors declare that they have no conflicts of interest.

Funding/Support

None declared.

References


