



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

ARTICLE INFO

Article Type:
Case Report

Article History:
Received: 3 Mar 2012
Revised: 27 Mar 2012
Accepted: 15 Apr 2012

Keywords:
Congenital Diaphragmatic Hernia
Morgagni Hernia
Congenital Heart Diseases
Partial A-V Canal Defect

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.

► 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:

Betigeri VM, Betigeri AV, Vasudevan A, Kasturi VA, SubbaRao KSVK. Circulatory Arrest – A Surgical option For Adult Window Ductus Closure. *Int cardiovasc Res J.* 2012; 6(2):62-64.

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 with equal movements 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.

Chest X-Ray postero-anterior view (Fig. 1) showed a soft tissue mass in the right cardio-phrenic angle silhouetting the lower right cardiac border. Two dimensional echocardiography confirmed the diagnosis of partial AVCD with moderate mitral regurgitation (fig.2a, 2b, 2c). Further evaluation by ultrasound abdomen revealed upward displacement of the liver into the thoracic cavity but the diaphragm was found to be intact. A computerised tomography scan of the abdomen (Fig. 3) showed the herniation of right lobe of liver through the retrosternal

* Corresponding author: Kunal, Department of CTVS, G. B. Pant Hospital, New Delhi, India, E-mail: drkunal97@gmail.com



Figure 1. Chest x ray PA views showing a soft tissue mass in the right cardio-phrenic angle silhouetting the lower right cardiac border.

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 stonecutter. 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 eventration 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

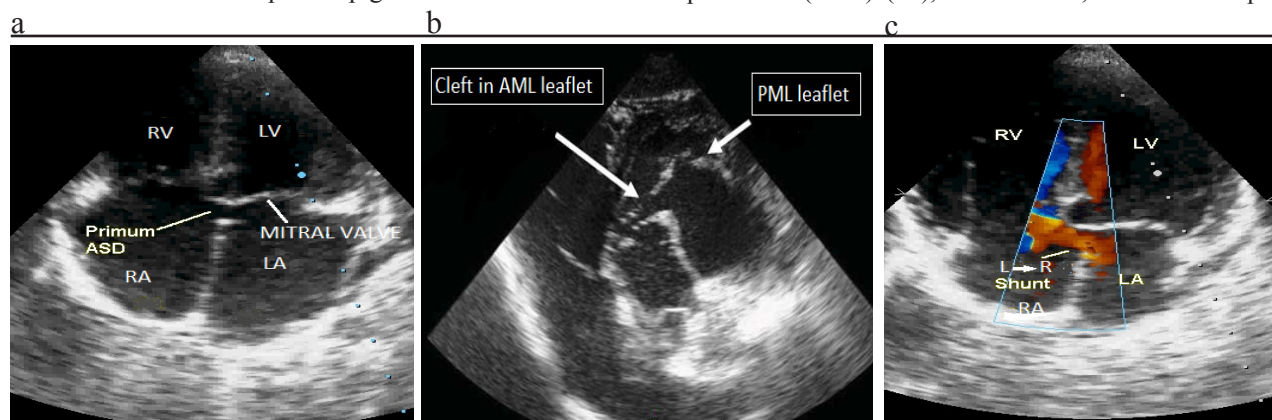


Figure 2. Echocardiography findings in partial AVCD. (a: apical four chamber view showing defect; b: cleft in mitral valve; c: colour Doppler showing shunt)

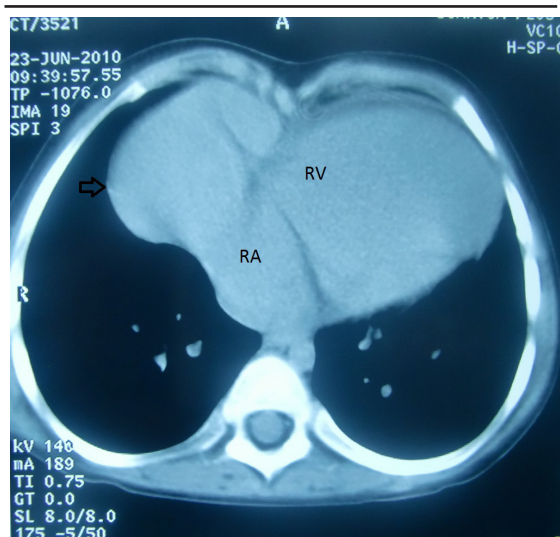


Figure 3. Non contrast axial CT sections at the level of upper xiphisternum showing herniation of right lobe of liver in the thorax (arrow) abutting the adjacent cardiac chambers [right atrium (RA) and right ventricle (RV)].

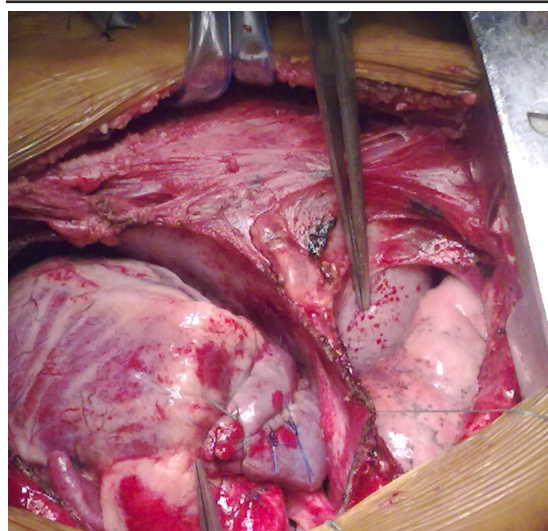


Figure 4. Operative photograph showing the Morgagni hernia visible through the right pleural space (tip of forceps). Right atrial suture line is also seen.

defect (VSD) and anomalous pulmonary venous return, Scimitar syndrome (11), and coronary heart disease (12). 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 (13). 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 the association of Morgagni hernia with an atrio-ventricular canal defect. This emphasizes 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 (2) 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 an associated cardiac anomaly is detected, then a trans-sternal approach is recommended (14).

Acknowledgement

There is no acknowledgment.

Financial Disclosure

The authors declare that they have no conflicts of interest.

Funding/Support

None declared.

References

1. Al-Salem AH, Nawaz A, Matta H, Jacobsz A. Herniation through the foramen of Morgagni: early diagnosis and treatment. *Pediatr Surg Int.* 2002;18(2-3):93-7.
2. Berman L, Stringer D, Ein SH, Shandling B. The late-presenting pediatric Morgagni hernia: a benign condition. *J Pediatr Surg.* 1989;24(10):970-2.
3. Pokorny WJ, McGill CW, Harberg FJ. Morgagni hernias during infancy: presentation and associated anomalies. *J Pediatr Surg.* 1984;19(4):394-7.
4. Larrey DJ. Clinique chirurgicale: exercée particulièrement dans les camps et les hôpitaux militaires, depuis 1792 jusqu'en 1829. *Gabon;* 1829.
5. Brown RW. A case of bilateral parasternal diaphragmatic hernia. *Thorax.* 1952;7(3):265-9.
6. Wang C, Lo WC. Clinical Presentation and Radiographic Diagnosis of Non-traumatic Childhood Diaphragmatic Hernia. 2004
7. Comer TP, Clagett OT. Surgical treatment of hernia of the foramen of Morgagni. *J Thorac Cardiovasc Surg.* 1966;52(4):461-8.
8. Elawad ME. Diaphragmatic hernia in Down's syndrome. *Ann Trop Paediatr.* 1989;9(1):43-4.
9. Honore LH, Torfs CP, Curry CJ. Possible association between the hernia of Morgagni and trisomy 21. *Am J Med Genet.* 1993;47(2):255-6.
10. Nouri NM, Rajae Sh, Marajie MS. Pediatric Morgagni hernia with secundum atrial septal defect (report of two cases). *Journal of Shahid Sadoughi University Of Medical Sciences.* 2002;1(2):91-94.
11. Adebo OA, Osinowo O, Adebajo SA, Grillo IA. Scimitar syndrome with associated Morgagni hernia in a Nigerian infant. *J Natl Med Assoc.* 1979;71(10):965-7.
12. Tuygun AK, Balci AY, Tuygun A, Gunay R, Sensoz Y, Yurtseven N, et al. Simultaneous operation in a patient with coronary heart disease, abnormal orifice of coronary arteries, Morgagni hernia, atrial septal defect, and pericardial and pleural agenesis. *Heart Surg Forum.* 2010;13(4):E260-2.
13. Etheridge SP, Ruttenberg HD, Williams RV. An unusual cause of severe cyanosis in infancy. *Ann Thorac Surg.* 2001;71(3):1016-8.
14. Mert M, Gunay L. Transsternal repair of Morgagni hernia in a patient with coexistent ventricular septal defect and Down syndrome. *Acta Chir Belg.* 2006;106(6):739-40.