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journal homepage: www.jpascasereports.comA rare case of Morgagni hernia associated with cor triatriatum[☆]

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ABSTRACT

The Morgagni hernia is a rare congenital disorder that is often associated with other anomalies such as congenital heart disease. We present here a case of Morgagni hernia with concomitant cor triatriatum, both of which were repaired in one operation via a sternotomy approach with satisfactory 6 months follow-up. To the best of our knowledge, this is the first case of its kind reported.

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The rarest congenital diaphragmatic hernia is the Morgagni hernia, which accounts for 3–4% of all diaphragmatic hernias [1]. A Morgagni hernia is believed to occur due to lack of fusion or muscularization of the pars sternalis and pars costalis during diaphragmatic development resulting in a triangular parasternal gap known as the Morgagni gap on the right and the Larrey gap on the left [1,2]. The chance of association with other anomalies in Morgagni hernia is high. It has been reported that 25% of these patients have co-existing congenital heart disease, including atrial septal disease, ventricular septal disease, patent ductus arteriosus and even coronary artery anomaly [3–5]. Nonetheless, to the best of our knowledge, the case we present here is the first one of Morgagni hernia in association with cor triatriatum which *per se* is rare in congenital heart disease [6].

1. Case report

A one year and five months old boy was referred to the Cardiology department at our institution due to secundum atrial septal defect, cor triatriatum sinister, permanent left superior vena cava and pulmonary infection. On physical examination, a II/6 grade gentle murmur at the left border of the sternum, second intercostal space with some fine rales in both lungs were heard. On chest X-ray, a diaphragmatic hernia was accidentally found as air-filled bowel

overlying the heart silhouette (Fig. 1), while there was no gastrointestinal symptoms presented. Barium meal and multislice computed tomography were performed and the hernia was confirmed as a Morgagni hernia which was immediately to the left of and behind the sternum (Figs. 2 and 3). Subsequently, after pneumonitis was controlled, the patient was transferred to our department for surgery.

The operative plan was to repair the hernia first followed by correction of the intracardiac anomalies. During operation, a “soft tissue” pouch was found right behind the sternum. An opening sized 8 cm by 7 cm was made in the hernial sac, and contents which were part of the bowel were replaced back into abdominal cavity and the pouch was reinforced with a polyethylene terephthalate patch. Then cardiopulmonary bypass with bicaval venous cannulation was instituted and cross clamp of the aorta was applied. Intracardiac exploration findings were: typical cor triatriatum sinister as a membranous septum dividing the left atrium into a superior accessory chamber and an inferior true atrial chamber with a foramen in the membrane to allow for blood flow to go through; the secundum atrial septal defect sized 20 mm by 15 mm, as well as a left superior vena cava draining into coronary sinus. The cor triatriatum membrane was excised completely and the atrial septal defect was closed using an autopericardial patch in a routine fashion. Since the permanent left superior vena cava drained into coronary sinus with no change in cardiovascular physiology and hemodynamics, it was left in place. The operation went straightforward however the postoperative course was complicated by a prolonged lung infection. The patient was discharged successfully 2 weeks postoperatively (Figs. 4 and 5).

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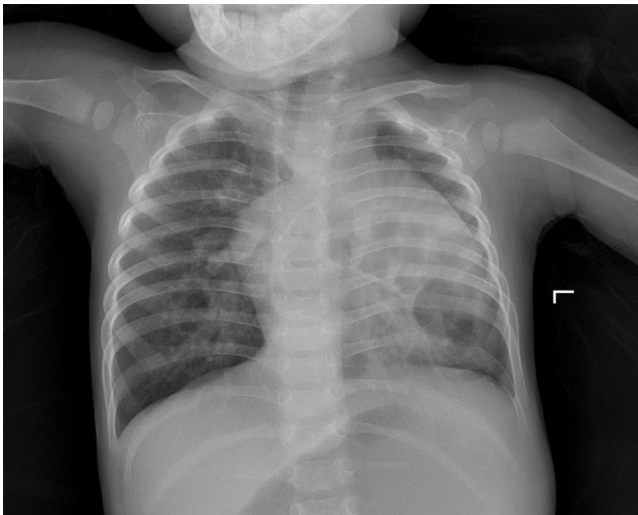


Fig. 1. Air-filling signs overlapping heart silhouette.

At the 6 months of postoperative follow-up the patient had been enjoying normal physical activity and echocardiography demonstrated satisfactory heart function with no residual interatrial pressure gradient.

2. Discussion

The Morgagni hernia, also known as Morgagni-Larrey hernias, anterior parasternal diaphragmatic hernia or subcostosternal

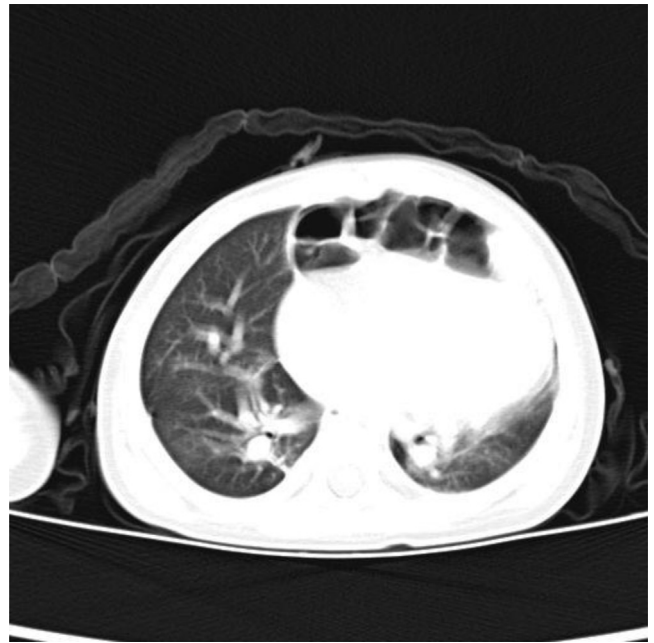


Fig. 3. Axial view of chest CT showing substernal air-filled bowels.

diaphragmatic hernia, comprises 3–4% of all congenital diaphragmatic hernias [1]. It was first described by Giovanni Morgagni in 1761.

It is caused by abdominal viscera protruding up into the chest cavity through a small space between the attachments of the diaphragm to xiphoid process and 7th costal cartilage, the Morgagni gap on the right and the Larrey gap on the left. About 90% of Morgagni hernias occur on the right, 2% on the left, and 8% bilaterally [1,2]; a hernia sac is present in more than 95% of all cases. The most common content of the hernia sac is colon (80%) although the small intestines, part of the liver, and omentum may also reside within [5].

The large proportion of these cases are found incidentally as they are usually asymptomatic. Symptoms, when present, include retrosternal pain, epigastric discomfort and dyspnea, with the most common one being recurrent chest infection, as was the case presented here.

Previous studies have stressed the use of multislice CT scan with coronal and sagittal reformatted images as the most effective and useful imaging technique to detail diaphragmatic hernias including the Morgagni hernia. This imaging modality was also used to diagnose our patient's hernia. Additionally, a barium meal was performed to confirm the presence of the hernia by demonstrating the presence of abdominal content within the thoracic cavity.

Association with other forms of congenital malformation is very common. Cigdem reported 12 patients out of 16 with Morgagni hernia with additional concomitant congenital anomalies. Eight (66.6%) of them had multiple anomalies [4]. In another series, associated anomalies were seen in 14 (70%) of all patients. Five (25%) had congenital heart disease, 4 (20%) had malrotation of bowel, and 3 (15%) had Down's syndrome [5]. Association with CHD ranged from 25% to 31% [4,5] and included atrial septal defect, ventricular septal defect, patent ductus arteriosus, etc. Some also reported concomitant pericardial and pleural defect [3]. To the best of our knowledge, it is the first time that the Morgagni hernia in association with cor triatriatum has been reported in the literature.

Surgical correction is the mainstay of treatment and elective surgery is advised even when the patient is asymptomatic in order



Fig. 2. Sagittal view of chest CT showing substernal air-filled bowels.

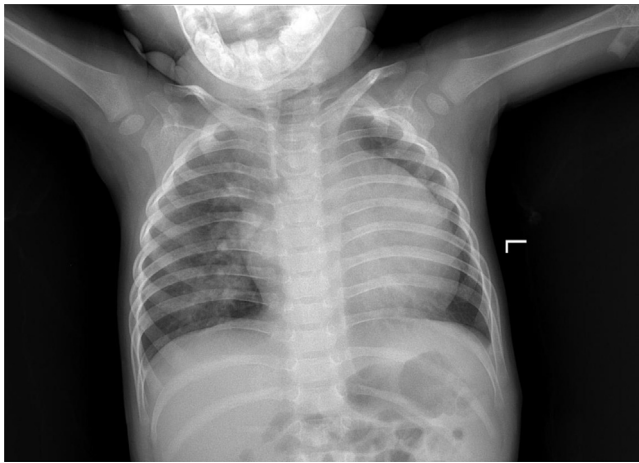


Fig. 4. Postoperative chest X-ray shows a slightly enlarged heart silhouette without overlapping air-filling signs.

to avoid strangulation and incarceration of abdominal viscera. Transabdominal and transthoracic approaches are primarily used to repair the defect. And laparoscopic repair is now considered the procedure of choice [7–9]. However, the transsternal repair of the hernia is preferred in patients undergoing concomitant open heart surgery [3,10,11] because of the necessity of concurrent heart defect repair. In a combined management of aortic valve stenosis and a Morgagni hernia, the author believed the treatment should be in general a two stage procedure, but still chose to perform one-stage repair with satisfactory outcome [12]. Similar one stage management strategy has been reported [3]. Even under the one-stage



Fig. 5. Postoperative barium meal confirms normal position of gastrointestinal tract.

circumstances, discussions of surgical procedures requiring simultaneous operations always bring up arguments about what to do first. Some advocate to perform heart procedures first because of the vulnerability of the heart and hemodynamics as well as an increased potential of infection [3]. We believe the repair of the hernia comes first naturally since the pouch with content inside is in the way of operative field making it reasonable to deal with it first. Since these cases are so rare and the number is too small to make any statistical analysis, we believe the management strategy should be tailored to every individual case associated with varying specific congenital heart disease.

3. Conclusion

To date, this is the first case of Morgagni hernia in association with cor triatriatum. Despite perioperative lung infections, the baby was successfully managed surgically and discharged from hospital with a satisfactory result. The follow-up at 6 month postoperatively showed normal heart function and a return to physical activity.

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Disclosure

The authors have no conflicts of interest to disclose.

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