Morgagni Hernia With Down Syndrome And Pectus Carinatum: Case Report

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Abstract

Morgagni hernia is a rare type of congenital diaphragmatic hernia. Numerous congenital defects have been described with it. We report a case of Morgagni hernia with Down syndrome and pectus carinatum. In this case, Morgagni hernia was repaired using thoracic approach but, pectus carinatum was not repaired because of small deformity.

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Introduction

Morgagni hernia is a congenital herniation of the abdominal contents through the retrosternal defect into the thoracic cavity. It is rare, accounting for less than 5 % of all types of congenital diaphragmatic hernia. They are frequently found in association with other congenital defects including Down’s syndrome, mental retardation, omphalocele, tetralogy of Fallot and ventral septal defect. In this report, we submit a case that have Morgagni hernia with Down’s syndrome and pectus carinatum.

CASE REPORT

Six years old a boy was admitted for asymmetry of anterior chest wall in our clinic. In our patient, dyspnea was present symptom. There was mental retardation in patient.

Chest radiograph showed a heterogeneous opacity with contained air, in right paracardiac area (fig.1). This was confirmed by barium meal to be a right Morgagni hernia (fig.2 A,B). Computed tomography demonstrated large and small bowel loop contents in right paracardiac area and, right anterior chest wall is slightly more prominent than left (fig.3). DNA analysis showed a translocation of chromosome 21.

Fig.1 Chest radiograph showed heterogenous opacity with contained air, in right paracardiac area. Right hemidiaphragm was not distinguished from heterogenous opacity.

Right posterolateral thoracotomy was performed in the patient. Exploration through the sixth intercostal space provided a good exposure for the Morgagni hernia. Following the excision of the hernia sac, adhesions were released with blunt and sharp dissection. Hernia contents were pushed into the abdomen. Defect was repaired by interrupted 0 silk stitches. The operation of pectus carinatum was not performed because of small thoracic deformity.
Fig. 2a Barium meal shows herniated bowel loop in right paracardiac area.

Fig. 2b Diaphragm defect is localized anteriorly.

Fig. 3 CT scan demonstrated large and small bowel loop in the right paracardiac area.

Discussion

Morgagni hernia so called substernocostal diaphragmatic hernia is rarely encountered entity. Retrosternal Morgagni hernia is the least common congenital diaphragmatic hernia, accounting for only 2 %- 3 % of all diaphragmatic defects. Embryologically, the foramen of Morgagni represents the junction of the septum transversum, the lateral component of diaphragm, and the anterior thoracic wall\(^3\).

While these hernias are usually established in infancy, they may remain unestablished until later in life. One interesting observation in children with delayed presentation of congenital diaphragmatic hernia in these cases is the high occurrence of a well-defined hernia sac (20 %-40 %)\(^1,4\). There was well-defined hernia sac in our patient.

There is a wide range of associated abnormalities, particularly congenital heart defects and trisomy 21. Several investigators were reported chest wall deformities with Morgagni hernia. Several authors have reported cases of Down syndrome within their series of Morgagni hernia\(^1,2\). However, both pectus carinatum and Down syndrome have not reported with Morgagni hernia patients in the literatures.

The diagnosis of Morgagni hernia is based on chest radiograph with a lateral to show anterior herniation of bowel loops, which can be confirmed by barium enema or barium meal and follow-through. The diagnosis can be difficult or delayed if the hernia sac is empty or contains omentum or part of the liver\(^5,6\).

Morgagni hernia usually contains transverse colon, omentum, liver and sometimes small bowel or stomach. In our patient the hernial sac contained large and small bowel.

Both transabdominal and transthoracic approaches are recommended in the surgical repair of Morgagni hernia. Transabdominal repair of Morgagni hernia has been used by numerous authors with favourable result. Transthoracic approach provides easy repair of the diaphragmatic defect, and pericardial adhesions can be easily released. Some surgeons performed an abdomino-mediastinal approach for diaphragmatic repair\(^5,7\).

Recently, it is claimed that video assisted endoscopic surgery is an effective and safe methot for to repair fix Morgagni hernia\(^8\). We performed right posterolateral thoracotomy in our case.

In conclusion, Morgagnia hernia may find with both pectus carinatum and Down syndrome. Repairing of Morgagnia hernia can be performed on using the thoracic approach.
References

5. Groff DB Diagnosis of a Morgagni hernia complicated by a previous normal chest X-Ray J Pediatr Surg 1990; 25: 556-557