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CASE REPORT / *OLGU SUNUMU*

Successful Repair of Congenital Morgagni Hernia and Ventricular Septal Defect via Median Sternotomy in a Patient with Down Syndrome, Anal Atresia, Pectus Carinatum and Congenital Hypothyroidism*

Down Sendromu Anal Atrezi Pectus Carinatum ve Konjenital Hipotiroidinin Eşlik Ettiği Ventriküler Septal Defekt ve Konjenital Morgagni Hernisinin Sternotomi ile Başarılı Onarımı

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ABSTRACT

Here we report a 5-month-old female patient with Down syndrome who was successfully operated on due to congenital Morgagni hernia and ventricular septal defect concomitantly via median sternotomy. Ventricular septal defect was repaired with a polytetrafluoroethylene patch through the right atriotomy using cardiopulmonary bypass, and diaphragmatic hernia was repaired primarily with prolene sutures reinforced with teflon pledgets after excising the diaphragmatic hernia pouch simultaneously. Entire operation was completed via median sternotomy, and transabdominal approach was not required to repair the diaphragmatic hernia. Postoperative course was uneventful.

Keywords: Congenital Morgagni Hernia, Ventricular Septal Defect, Median Sternotomy, Down Syndrome, Anal Atresia, Pectus Carinatum

ÖZ

Olgu sunumumuzda, 5 aylık Down sendromlu kız hastada yapılan başarılı konjenital Morgagni hernisi ve ventrikül septal defekt onarımını sunmaktayız. Median sternotomi yaklaşımıyla aynı seansta yapılan onarımlarda kardiyopulmoner bypass eşliğinde VSD, polytetrafluoroethylene yama ile kapatıldı. Diaframdaki defekt ise primer olarak, transabdominal yaklaşıma gerek olmadan onarıldı. Postoperatif seyir sorunsuzdu.

Anahtar Kelimeler: Morgagni Hernisi, Hipotiroidi, Ventriküler Septal Defekt, Anal Atrezi, Pectus Carinatum

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a congenital defect involving abnormal development of the diaphragm. The hole in the diaphragm causes the abdominal organs to protrude into the mediastinum or thoracic cavity (1). The congenital Morgagni hernia (CMH) is rare in the literature. The incidence is found to be 3-4% of diaphragmatic hernias (2). The association of such a rare hernia with other congenital heart diseases is extremely rare. We report a 5-month-old infant diagnosed with Down syndrome, ventricular septal defect (VSD), CMH in which all defects were repaired concomitantly via median sternotomy approach.

CASE REPORT

A 5-month-old female patient (4,7 kilos in weight) with Down syndrome was referred to our institution for the operation due to the VSD. She was operated on previously for anal atresia in neonatal period, and no additional complication was occurred in the past 5 months. Anal dilatation was made daily. On physical examination, cardiac murmur and a mild

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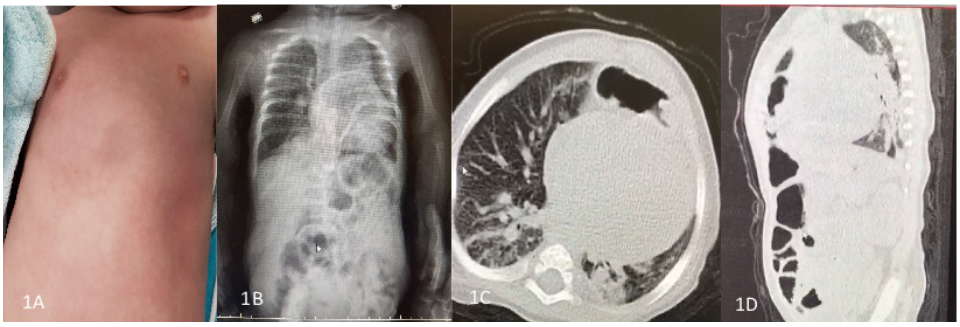

Figure 1A: Pectus carinatum deformity.

Figure 1B: Preoperative X-ray shows gas-filled abdominal organs located in the mediastinal cavity.

Figure 1C, 1D: Preoperative thorax computed tomography imaging indicates gas filled abdominal organs in the anterior mediastinum.

pectus carinatum deformity was present (Figure 1A). She did not have any serious respiratory distress, and therefore she was not investigated with further diagnostic tests except echocardiogram. On routine chest X-ray for preoperative investigation, stomach and intestines were observed in the mediastinal area (Figure 1B). Thorax computed tomography (CT) revealed a diaphragmatic hernia with protrusion of stomach and intestines to the mediastinal cavity (Figures 1C,1D). Echocardiography revealed a 20 mm perimembranous VSD, patent ductus arteriosus (PDA) and a small secundum atrial septal defect (ASD). She was pulmonary hypertensive on echocardiography. She was under thyroid hormone replacement treatment due to congenital hypothyroidism, and preoperative thyroid hormone levels were in normal ranges. Informed consent was taken from the patient's parents.

We decided to repair the VSD and CMH concomitantly. Midline sternotomy was performed carefully. Anterior mediastinum was observed as free of any abdominal organs (Figure 2A). However, anterior and medial part of diaphragm had a cleft, and the hernia sac was detected to be protrude into the defect after the manual compression on the abdominal wall (Figure 2A). Hemidiafragmas were dissected (Figure 2B), reunited (Figure 2C). Using prolene sutures reinforced with teflon pledgets, the defect was repaired (Figure 2C). PDA was ligated. Cardiopulmonary bypass (CPB) was instituted using aortic and bicaval cannulation. The VSD was closed using a Goretex® patch through tricuspid valve. Secundum ASD was closed primarily. The weaning from CPB was uneventful. No rhythm disturbance occurred. In the postoperative period, intravenous iloprost infusion was given for 2 days, and sildenafil was continued following the extubation at the second postoperative day. Her postoperative period was uneventful, and she did not have

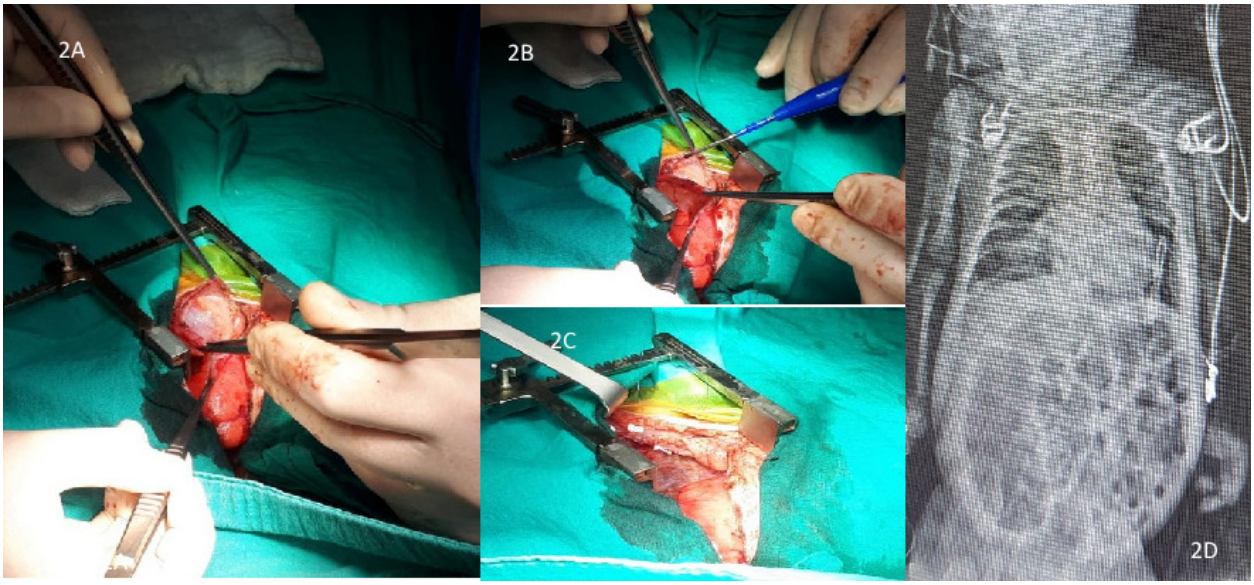

Figure 2A: Intraoperative view of intraabdominal organs protruding into anterior mediastinum.

Figure 2B: Intraoperative view: dissection of the hemidiaphragms.

Figure 2C: Intraoperative view of the primary repair of the Morgagni hernia using prolene sutures.

Figure 2D: Postoperative X-ray, the mediastinal cavity does not have gas-filled abdominal organs.

any problem about intestinal passage due to the repaired CMH or previously repaired anal atresia. Postoperative X-ray was normal regarding abdominal organ locations (Figure 2D). The patient was discharged on postoperative day 10.

DISCUSSION

The CMH is a muscular defect of anterior diaphragm that causes abdominal visceral organs to protrude into the chest cavity, and very rare among congenital diaphragmatic hernias in the ratio of 3-4 % (3). It can be placed on the right, left or bilaterally in the ratio of 90 %, 2 % and 8 %, respectively (3). The content in the hernia sac is colon mostly (80 %) (4). The CMH can be associated with congenital heart diseases ranged from 25 % to 31 % (4). Concomitant congenital heart diseases are as follows: Scimitar syndrome (5), VSD (6), cor triatriatum (7), tetralogy of Fallot (8), coarctation of aorta (8), ASD (8), dextrocardia (6), anomalous pulmonary venous return (6), endocardial cushion defect (6), Williams syndrome and aortic valve stenosis (9), association of VSD and Down syndrome (10). CMH is usually asymptomatic. Respiratory complaints occur due to the compression of the lower lobe of the ipsilateral lung.

In most of the cases, congenital diaphragmatic hernias are repaired via transabdominal approach (11,12,13), and in older patients, the hernia is repaired with a patch due to large defect (11,12,13).

Respiratory distress was not present in our patient. Therefore, CMH remained undiagnosed for the past 5 months. However, the need for the closure of a large VSD required median sternotomy obviously, and a concomitant median laparotomy would complicate the postoperative period after an open-heart operation in a patient with pulmonary hypertension and Down syndrome. Therefore, we repaired the congenital diaphragmatic hernia via median sternotomy simultaneously with VSD closure. Due to the young age, her diaphragmatic tissues were floppy enough to dissect and reunite, thus after excising the hernia sac, it was easy to repair the defect primarily.

CONCLUSION

Median sternotomy may be the optimal choice for the repair of CMH in patients requiring open-heart surgeries especially with associated diseases causing pulmonary hypertension such as Down syndrome and left-to right shunting anomalies. Our patient is a rare example for the association of CMH, VSD, Down syndrome, anal atresia, pectus carinatum and congenital hypothyroidism.

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