

Case Report

Bochdalek Hernia and Intrathoracic Ectopic Kidney: Presentation of Two Case Reports and Review of the Literature

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ABSTRACT

Bochdalek hernia is a congenital abnormality with high morbidity and mortality characterized by passage of the abdominal organs into the thoracic cavity through a diaphragmatic defect. Intrathoracic location of abdominal organs such as kidneys is very rare, with a reported incidence of only 0.25% in the literature. Herein, we present two cases of Bochdalek hernia with a herniation of intra-abdominal organ such as kidney that was treated in our clinic and compare this rare case with those in the literature. In both cases, the functionally normal kidneys were left *in situ* during diaphragmatic repair. No complications were observed during the postoperative period, and 10- and 1-year follow-ups. In cases with Bochdalek hernia associated with an intrathoracic ectopic kidney, the functionally normal ectopic kidneys were left *in situ* during repair of the diaphragmatic defect without complications.

KEYWORDS: *Bochdalek hernia, intrathoracic ectopic kidney, review of literature, two case reports*

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INTRODUCTION

Developmental anomalies of the diaphragm include various pathological conditions such as eventration, hernias, and hypoplasia, or aplasia at various degrees. Congenital diaphragmatic hernias are the substernal Morgagni's hernia and the posterolateral Bochdalek hernia.^[1] In 1848, Vincent Alexander Bochdalek^[2] first described a congenital abnormality with high morbidity and mortality in which abdominal organs pass into the thoracic cavity through a diaphragmatic defect. In 80%–90% of the cases, the defect develops in the left posterolateral part of the diaphragm.^[2] The posterolateral part of the pleuroperitoneal canal closes last and a closure defect occurring at the end of the embryonic period leads to this rare condition. Most cases are symptomatic during the neonatal period.^[3]

Mainly the small bowel, spleen, stomach, colon, and left lobe of the liver tend to pass through the diaphragmatic defect into the thoracic cavity.^[2,3] Nearly, 5% of all ectopic kidneys are intrathoracic.^[4] An intrathoracic ectopic kidney associated with a Bochdalek hernia is a very rare condition with a reported incidence of 0.25%.^[5]

In this paper, we present two cases of the left Bochdalek hernia combined with intrathoracic ectopic kidney and reviewed the literature on this condition.

CASE REPORTS

Case 1

A 6-month male infant had suffered from intermittent respiratory distress and vomiting since birth. On physical examination, he was tachypneic, and crepitant rales were heard in the basal segment of the left hemithorax. His chest X-ray revealed intrathoracic mass lesion and intestinal gases in the left hemithorax which were consistent with Bochdalek hernia. A computed tomography (CT) image of the thorax revealed that the stomach, transverse colon, and left kidney, had herniated through the left posterolateral diaphragmatic defect into the left hemithorax resulting to the displacement of the heart and major vascular structures toward the right hemithorax.

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Table 1: Clinical features of patients with an intrathoracic ectopic kidney associated with a Bochdalek hernia

Researcher(s)	Age	Gender	Symptom(s)	Herniated part	Herniated organ(s)	Visualization method(s)	Surgical method	Nephropexy
Afşarlar <i>et al.</i> , 2011 ^[2]	45 days	Female	Respiratory distress	Right	Right kidney, small bowel, and colon	Chest X-ray, USG, CT	Laparotomy	Done
Padma <i>et al.</i> , 2014 ^[4]	1 year old	Male	Fever	Left	Left kidney	USG, DPTA	Not specified	Not specified
Juricic <i>et al.</i> ^[5]	1 day	Female	Prenatal diagnosis	Right	Small bowel, transverse colon, right kidney, and adrenal gland	Prenatal USG and MR, postnatal chest X-ray	Laparotomy	Done
Chen <i>et al.</i> , 2015 ^[9]	80 years old	Female	Dyspeptic complaints	Right	Right kidney	Chest X-ray, USG, CT	Undone	Not done
Sesia and Haecker, 2012 ^[10]	5 months	Female	Tachypnea	Left	Left kidney	Chest X-ray, USG	Not specified	Done
Murphy <i>et al.</i> , 2012 ^[11]	4 days	Female	Respiratory distress, malnutrition	Right	Right kidney and small bowel and colon	Chest X-ray	Not specified	Done
Murphy <i>et al.</i> , 2012 ^[11]	4 months	Female	Respiratory distress	Right	Right kidney and small bowel	Chest X-ray, USG	Not specified	Done
Obatake <i>et al.</i> , 2006 ^[6]	1 day	Male	Prenatal diagnosis	Right	Right kidney, small bowel, hepatic right lobe, and adrenal gland	Chest X-ray, USG, CT, IVP	Thoracotomy	Done
Chung <i>et al.</i> , 2010 ^[12]	55 years old	Male	Epigastric burning	Right	Right kidney	Abdominal X-ray, CT	Undone	Not done
Kayıran <i>et al.</i> , 2013 ^[14]	1 day	Erkek	Respiratory distress	Right	Right kidney and hepatic right lobe	Chest X-ray, echocardiography	Not specified	Not done
Noh <i>et al.</i> , 2015 ^[15]	56 years old	Female	Fever, abdominal pain, urinary symptoms	Left	Left kidney	Chest X-ray, CT	Not specified	Not specified
Shah <i>et al.</i> , 2012 ^[16]	20 years old	Female	Cough, fever	Left	Left kidney	Chest X-ray, USG, CT, IVP	Undone	Not done
Panda <i>et al.</i> , 2009 ^[17]	1 day	Male	Prenatal diagnosis	Left	Left kidney, spleen, small bowel, and colon	Chest X-ray, echocardiography, USG, MR	Laparotomy	Done
Lee <i>et al.</i> , 2006 ^[18]	28 years old	Male	Respiratory distress	Right	Right kidney and hepatic right lobe	Chest X-ray, CT	Not specified	Not specified
Jha <i>et al.</i> , 2014 ^[19]	25 years old	Female	Respiratory distress	Right	Right kidney, hepatic right lobe and colon	Chest X-ray, USG, CT	Thoracotomy and laparotomy	Done
Karaoglanoglu <i>et al.</i> , 2006 ^[20]	22 months	Male	Recurrent lung infection, fever, vomiting	Right	Right kidney and colon	Chest X-ray, CT, IVP	Thoracotomy	Done
Masturzo <i>et al.</i> , 2001 ^[21]	1 day	Male	Prenatal diagnosis	Right	Right kidney, hepatic right lobe, small bowel, and colon	USG	Laparotomy	Done
Dingeldein <i>et al.</i> , 2008 ^[22]	18 months	Male	Problems with extubation after tonsillectomy	Bilateral	Bilateral kidneys and adrenal glands	Chest X-ray, CT, MR	Laparotomy	Done
Hidaka <i>et al.</i> , 2012 ^[23]	1 day	Erkek	Prenatal diagnosis	Left	Left kidney, stomach, spleen small bowel, and colon	USG, chest X-ray	Laparotomy	Not specified
Kawashima <i>et al.</i> , 2014 ^[24]	2 years old	Male	Abdominal pain, loss of consciousness	Left	Left kidney, spleen, small and bowels	Chest X-ray, CT	Laparoscopic repair	Done

Contd..

Table 1: Contd...

Researcher(s)	Age	Gender	Symptom(s)	Herniated part	Herniated organ(s)	Visualization method(s)	Surgical method	Nephropexy
Subramanian and Goldfarb, 2008 ^[25]	23 years old	Female	Right-sided pain	Right	Right kidney and adrenal glands	Chest X-ray, USG, MR	Undone	Not done
Fiaschetti <i>et al.</i> , 2010 ^[26]	62 years old	Male	Cough, abdominal pain	Left	Left kidney and colon	Chest X-ray, abdominal X-ray, USG, CT, MR	Not specified	Not specified

USG=Ultrasound sonography; CT=Computed tomography; DPTA=Diethylentriamin pentasetik asit; MR=Magnetic resonance; IVP=Intravenous pyelography

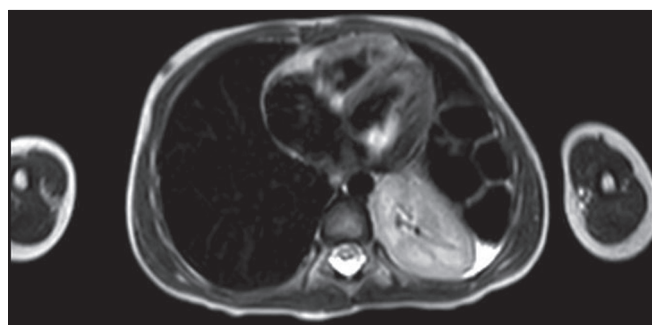


Figure 1: The MRI image of intestinal loop along with the left kidney's being herniated into the left thorax due to the left posterolateral diaphragmatic defect

However, the location of the left adrenal gland could not be evaluated properly. These preoperative findings were suggestive of a Bochdalek hernia. Surgical exploration confirmed herniation of the stomach, small bowel, colon, and left kidney through a diaphragmatic defect into the left hemithorax. After reduction of the bowels and stomach into the abdominal cavity, the hernial sac was opened, which revealed the left extrapleural kidney to the left of the heart and fixed on the posterior wall of the thorax. As was anticipated, the ureter and its vascular pedicle were longer than normal. Since the kidney was covered with pleura, the location of the left adrenal gland could not be determined. Besides, the left adrenal gland was not found in the abdomen. In addition, there was no renal rotation abnormally. The diaphragmatic defect was repaired primarily; the diaphragmatic margin remained nearly 2 cm below the lower pole of the kidney. To avoid inadvertent injury to the ectopic kidney, it was left untouched. During the postoperative period, the patient developed no complications and was discharged on postoperative day 7. Ten years of follow-up revealed no complications.

Case 2

A 7-year-old girl had complained of abdominal pain for 10 days. Her abdominal pain started at the periumbilical region and radiated toward the left lower abdominal quadrant. Her medical history revealed recurrent urinary tract infections. Hemogram, renal function tests, biochemical analysis, and complete urinalysis results

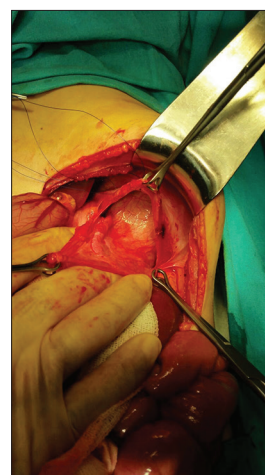


Figure 2: The image of intrathoracic kidney and Bochdalek hernia in posterolateral part of the left diaphragm at laparotomy

were within normal limits. Abdominal ultrasound (US) could not reveal the left kidney while her chest X-ray revealed intrathoracic mass lesion and intestinal gases in the left hemithorax which were consistent with Bochdalek hernia. Magnetic resonance imaging (MRI) showed herniation of the left kidney, and spleen together with intestinal loops through the left posterolateral diaphragmatic defect into the left hemithorax. There was no hydronephrosis. The MRI was taken without contrast material, the location of the left adrenal gland could not be evaluated [Figure 1]. Tc-99 m dimercaptosuccinic acid scanning studies revealed that the function of the medially located dysmorphic looking intrathoracic left kidney were also within normal limits. The right kidney had retained its normal structure, function, and location. Total relative uptake was 54% in the left and 46% in the right kidneys, respectively. Absolute uptake was 15% in the left and 13% in the right kidneys, respectively. Vesicoureteral reflux was not seen in voiding cystourethrography. Laparotomy revealed a Bochdalek hernia measuring nearly 8 cm × 2 cm localized to the posterolateral side of the left diaphragm [Figure 2]. Herniation of the splenic flexure of the transverse colon, part of the small bowel, and the left kidney into the left hemithorax was observed. After reduction of the bowels into the abdominal cavity, the hernial sac was opened;

on abdominothoracic exploration, an extrapleural ectopic kidney was observed to the left of the heart and fixed to the posterior thoracic wall. The left adrenal gland could neither be located in the abdomen nor in the thoracic cavity. The associated ureter and its vascular pedicle were longer than normal. Furthermore, there was no renal rotation abnormality. The diaphragmatic defect was repaired primarily, and the diaphragm margin remained nearly 2 cm lower than the renal tissue. The kidney was left *in situ*. During the postoperative period, there were no complications, and the patient was discharged on postoperative day 9. During 1 year of follow-up, no medical problems were encountered.

DISCUSSION

Bochdalek hernia is the most frequent form of congenital diaphragmatic hernia, with an incidence of one case out of nearly 2000–5000 births. Eighty to ninety percent of Bochdalek hernias occur as a result of extrusion of abdominal organs through a posterolateral defect of the diaphragm into the thoracic cavity.^[2] The intrathoracic ectopic kidney is a very rare congenital anomaly with an incidence of 1/16,000.^[3] It is more frequent in males and constitutes the rarest form of all ectopic kidneys (<5% of cases).^[3] In the literature, an intrathoracic ectopic kidney was reported in the left hemithorax in 62% of cases, and in the right hemithorax or in both hemithoraces in 36% and 2% of cases, respectively.^[6] Similar to reports in the literature, both of our cases, the intrathoracic ectopic kidney was on the same side as the Bochdalek hernia.

Intrathoracic kidneys are divided into four distinct groups based on the status of the diaphragm: Group 1: Intrathoracic ectopic kidney associated with a closed diaphragm; Group 2: Intrathoracic ectopic kidney associated with diaphragmatic eventration; Group 3: Intrathoracic ectopic kidney associated with traumatic diaphragmatic rupture; and Group 4: Intrathoracic ectopic kidney associated with a congenital diaphragmatic hernia. According to this classification, both of our cases were Group 4.^[7]

It is still debatable whether intrathoracic renal ectopy is due to delayed diaphragmatic closure or ascent of the kidney above its normal location before closure of the diaphragm.^[8] Bochdalek hernias are more frequently left-sided owing to the left opening of the posterior diaphragm closes later in fetal life than the right.^[9] Intrathoracic ectopic kidneys are often associated with rotation anomalies, relatively longer and high-lying ureters, and vascular structures arising from their normal origins; renal function is usually normal.^[10] The intrathoracic ectopic kidneys of the two patients presented herein were localized to the left side and had

relatively longer vasculature and ureters. There is no clear information about adrenal gland localization in diaphragmatic herniated patients with the ectopic kidney in the literature. Some researchers have suggested that the developing adrenal gland affects the final anatomic location of the kidneys. However, it has been noted that in the intrathoracic kidney patients, the adrenal gland may be located above or below the kidney. This finding indicates that the kidney may either meet the adrenal gland in a superior localization or carry it along, or the kidney may ascend past the adrenal gland and leaving the adrenal to in its normal localization. Therefore, the mechanism of kidney ascension is independent of adrenal gland development.^[11] However, in our two cases, the location of the adrenal glands could not be evaluated properly.

Intrathoracic ectopic kidneys are visualized on thoracic radiographs as paravertebral mass-like lesions in the posterior mediastinum. Differential diagnoses include an esophageal duplication cyst, bronchogenic cyst, pulmonary sequestration, and aneurysm of the descending aorta. Thoracic US, contrast-enhanced CT, and MRI are useful imaging modalities to aid in diagnosis.^[7] The diagnosis of our first case was made based on chest X-ray and CT findings while our second case was diagnosed using MRI and scintigraphic studies.

The literature review did not indicate the complications of intrathoracic ectopic kidneys such as hypertension, impaired renal functions, and proteinuria. Intrathoracic ectopic kidneys diagnosed during adulthood have been reported to function normally.^[12] In addition, long-term follow-up of the patients with isolated intrathoracic ectopic kidneys has not revealed any complications.^[8] In both of our cases, the preexisting diaphragmatic defect was closed primarily; however, the ectopic kidney was not manipulated to avoid the risk of ureteral folding. No complications were observed during the 10- and 1-year follow-ups. Therefore, attempts to place the intrathoracic kidney in the abdomen is not recommended.^[11] However, as a result of the ectopic nature of the kidney and the risk of complications, these patients would benefit from close monitoring and follow-up.^[13] In contrast, some authors have recommended surgical repair.^[7] Many cases of intrathoracic ectopic kidneys associated with diaphragmatic hernias have been reported to have ureters and vascular structures of adequate length, allowing easy reduction of the ectopic kidney into the abdominal cavity without hemodynamic impairment.^[6] Although, there are studies suggesting surgical repair of diaphragm with lowering ectopic kidney to abdomen in the cases of concomitant Bochdalek herniation or in posttraumatic diaphragm ruptures, diaphragmatic hernia repair without

manipulation of ectopic kidney had been reported by Kayıran *et al.*^[14]

The literature contains case reports that discuss diagnosis, treatment, and follow-up of cases of intrathoracic ectopic kidney associated with Bochdalek hernia. Our literature review revealed only 22 cases of Bochdalek hernia associated with an intrathoracic ectopic kidney exist in 21 articles published in the English and Turkish medical literature. These 22 cases included eight adult and 14 pediatric cases. In the literature, the age of the cases at diagnosis ranged from the prenatal period to 80 years of age. Respiratory system findings were found in 57% ($n = 5$), and gastrointestinal system findings were found in 7.14% ($n = 1$) of 14 patients in child age group. Five patients (36.4%) were prenatal diagnosed. One adult case presented by Noh *et al.*^[15] had urinary symptoms. However, our cases had respiratory and gastrointestinal symptoms, respectively. In addition, patient 2 had recurrent urinary tract infections. However, in this case, recurrent urinary tract infections were not related to structural abnormalities, and it was attributed to poor hygienic conditions. In the literature, while, the kidneys were lowered into the abdomen in 78.5% ($n = 11$) of the children, 7.1% ($n = 1$) of the cases were not manipulated. However, 14.2% ($n = 2$) of the patients were not specified. In the adult age group, the kidneys were lowered into the abdomen in 7.1% ($n = 1$) of the patients, 50% ($n = 4$) of the cases were not manipulated. However, 37.5% ($n = 3$) of the cases were not specified. In our two cases, we opted to leave the kidney in place rather than returning it back into the abdominal cavity. The characteristics of all cases in the literature are presented in Table 1.

CONCLUSION

A Bochdalek hernia associated with an intrathoracic ectopic kidney is a very rare condition. Preoperative diagnosis is possible. In that cases, leaving the ectopic kidneys into the thoracic cavity and repairing the diaphragmatic defect is possible.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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