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Case Report

Surgery Section

An Ectopic Intrathoracic Kidney in an Adult Female: A Case Report

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ABSTRACT

The kidney is an organ prone to congenital anomalies owing to its complex and sequential development. Ectopia of the kidney is a rare entity with intrathoracic ectopia being an exceptionally uncommon occurrence. Intrathoracic kidneys represent less than 5% of all renal ectopias with a prevalence rate of less than 0.01%. The concurrent association of an intrathoracic kidney with a Bochdalek hernia is extremely infrequent with an incidence of 0.25%. Most of the patients are asymptomatic and have an uneventful clinical course. It is often incidentally detected simulating a posterior mediastinal mass. Thoracic kidney is a condition that shows male predominance. Here, authors present a case of a 43-year-old female patient who presented to the Institution with breathlessness and associated chest pain. Computed Tomography (CT) scan revealed left-sided Bochdalek hernia with the left kidney, adrenal gland, transverse colon, splenic flexure and proximal descending colon within the thorax. The patient underwent elective diagnostic laparoscopy followed by reduction of the contents and mesh repair of the diaphragmatic defect. The patient was discharged and remained asymptomatic on a periodic follow-up of over a year. Awareness regarding this rare entity can obviate the need for a battery of unnecessary investigations and operative procedures.

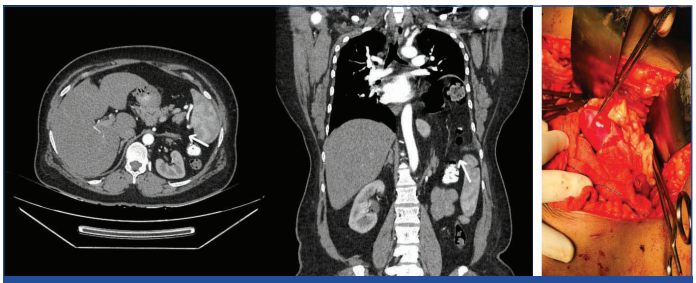
Keywords: Bochdalek hernia, Diaphragmatic hernia, Renal ectopia, Renal agenesis

CASE REPORT

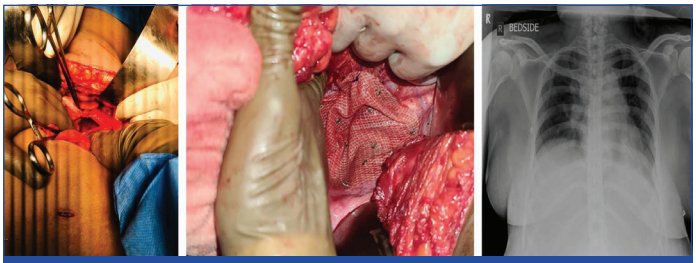
A 43-year-old female presented to the tertiary hospital with complaints of breathlessness since two months which was insidious in onset and progressive in nature, more towards the evening. She gave history of trauma sustained to the chest three years ago following a road traffic accident and was conservatively managed. Patient also gave history of associated chest pain and generalised fatigue. No history of abdominal pain or increase in frequency or burning micturition. Bowel habits were normal. Her abdominal scans dated five years earlier as a part of routine evaluation were reported as normal. Past medical and surgical history was insignificant. On examination, her general condition was fair and vitals were stable with no signs of respiratory distress. Crepitations were elicited in the left lower lung fields associated with gurgling sounds.

Relevant haematological and urine tests showed normal results. The CT scan showed a 7.6×6.3 cm defect in the posterolateral aspect of the left diaphragm with herniation of left adrenal gland, left kidney, transverse colon, splenic flexure and proximal descending colon through this defect as shown in [Table/Fig-1]. Postcontrast images demonstrated normal contrast excretion with a non dilated pelvicalyceal system with homogenously enhancing left kidney excreting into the left ureter. Patient was planned for laparoscopic management of the hernia. About 7×6 cm defect was noted with herniation of the adrenal gland, kidney and bowel. The bowel loops were released laparoscopically. Left kidney was found to be adherent to the thoracic cavity and could not be mobilised.

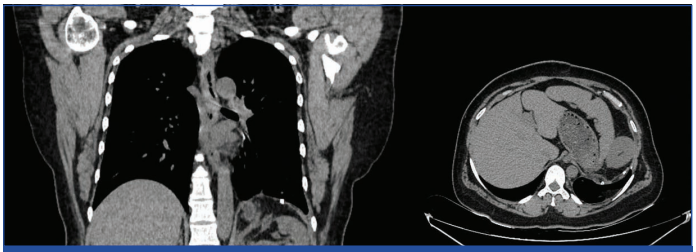
Hence, laparotomy was done by left subcostal incision and kidney was mobilised and returned to the abdominal cavity [Table/Fig-2]. Adhesiolysis was done to delineate the diaphragmatic defect inferiorly [Table/Fig-3] following which the defect was closed using 1 V- LOC sutures and a 12 cm circle composite-polyester mesh with bioabsorbable film was placed centring the defect [Table/Fig-4]. A 24 size intercostal drainage tube was placed in the pleural cavity and a 28 size abdominal drain was placed below the diaphragm and fixed. The procedure was well tolerated by the patient. Postoperative serial chest x-ray showed well expanded lungs with minimal pleural effusion [Table/Fig-5]. The abdominal and intercostal drainage tubes were removed on the third and seventh postoperative day, respectively. The patient was discharged on the 12th postoperative day and was followed-up on outpatient basis for a year. She remained asymptomatic with repeat CT scans showing intact diaphragm [Table/Fig-6].



[Table/Fig-1]: Preoperative axial and coronal sections of the CT images showing diaphragmatic defect with herniation of bowel loops, kidney and adrenals into the thoracic cavity. Arrows indicating the site of diaphragmatic defect.
[Table/Fig-2]: Intraoperative image showing reduction of kidney following its release from the thoracic cavity into the abdominal cavity. (Images from left to right)



[Table/Fig-3]: Diaphragmatic defect after reduction of contents.
[Table/Fig-4]: Composite mesh placement and fixation following closure of the defect.
[Table/Fig-5]: Postoperative chest x-ray showing well expanded lungs. (Images from left to right)



[Table/Fig-6]: Postoperative coronal and axial sections showing repaired defect.

DISCUSSION

In 1848, Bochdalek first described Congenital diaphragmatic hernia as resulting from failure of closure of pleuroperitoneal ducts at eight weeks of gestation [1]. The condition presents with respiratory symptoms in childhood whereas adults predominantly present with gastrointestinal symptoms. Renal ectopy is a congenital anomaly due to disrupted normal embryological migration of the kidneys. Ectopic kidneys occur in approximately 1 in 1000 births, however, only about 1 of 10 is ever diagnosed. Acquired cases in adulthood are rare, with a reported incidence of 0.17-0.6% [2,3]. When the herniation is present from birth, it is termed as "congenital" and when it presents later due to extension of intra-abdominal or perirenal fat into the thorax, it is termed as "acquired." Intrathoracic kidney has the lowest frequency rate among all renal ectopias [4,5]. Pfister-Goedeke and Burnier have classified thoracic kidney into four types: 1) Thoracic renal ectopia with closed diaphragm, 2) Thoracic renal ectopia with eventration of the diaphragm, 3) Thoracic renal ectopia with diaphragmatic hernia, 4) Traumatic rupture of the diaphragm with renal ectopia. This abnormality is reported to occur with higher frequency on the left than the right, with 2% of the patients having bilateral intrathoracic kidneys [5].

The association between intrathoracic kidney with Bochdalek hernia is unique. The intrathoracic kidney here differs from other intrathoracic renal ectopias as it is easily reducible from the thorax to the abdomen. As stated previously, an intrathoracic kidney may be preceded by trauma; however the traumatic intrathoracic kidney is extremely rare and predominantly seen in children due to the increased mobility of the paediatric kidney [6]. There are very few cases reported in adults. Esquis P et al., reported a case of intrathoracic herniation of the left kidney, 20 years after the initial motor vehicle accident. Subsequently, the kidney was reduced into the abdomen and the hiatus was closed using a laparoscopic approach [7]. Pascual Samaniego M et al., reported a case of traumatic intrathoracic kidney and suggested that, the rise in the intra-abdominal pressure causes herniation of kidney through pre-existent congenital pathway [8]. Vazquez-Alonso F et al., reported a case of a 61-year-old male who was incidentally diagnosed with an intrathoracic kidney while he was being worked up for a suspected lung malignancy [9]. Hence, in an adult, an intrathoracic kidney must be differentiated from posterior mediastinal masses and from benign or malignant diaphragmatic, pleural or pulmonary lesions.

The patients are usually asymptomatic as the intrathoracic kidney is almost always functional and does not display signs of dysplasia, contralateral kidney hypertrophy or lower urinary tract obstructive features. Hence, treatment is indicated only in the presence of obstruction or Vesicoureteric Reflex (VUR). Ultrasonographies, Computed Tomography, Magnetic Resonance Imaging are useful diagnostic modalities. Tc-99 m DMSA and Tc-99 m DTPA scintigraphies are used to demonstrate the kidney function precisely [1]. Patients, who necessitate treatment include those with gastrointestinal or respiratory symptoms, as in index case, where the patient presented with the primary complaint of dyspnoea.

CONCLUSION(S)

Intrathoracic renal ectopic is a rare occurrence and is a diagnostic challenge for both clinicians and radiologists akin. A high index of suspicion in patient presenting with breathlessness and history of trauma can result in early diagnosis and timely intervention with reduced morbidity and mortality.

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