A rare Case of Right Sided Bochdalek Hernia Co-existing with Intrathoracic Kidney
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ABSTRACT

Background: Congenital diaphragmatic hernias are developmental defects involving the diaphragm. Three major types have been identified; Bochdalek hernia, Morgagni hernia, and hiatus/hiatal hernia, with the posterolateral Bochdalek type being the most common. Rare concomitant existence with intrathoracic kidney was reported. Cases of left sided Bochdalek hernia were previously reported in the literature but none is associated with intrathoracic kidney. A successfully surgically treated rare case of right sided Bochdalek hernia co-existing with intrathoracic kidney in a neonate is therefore presented.

Keywords: Bochdalek hernia, Intrathoracic kidney, Neonate

Introduction
Congenital diaphragmatic hernias (CDHs) are developmental defects of the diaphragm that allows abdominal viscera to herniate into the chest. Three types have been identified and includes the most common posterolateral Bochdalek hernia, anterior Morgagni hernia and hiatal hernia. The reported incidence of CDH is between 1 in 2000 to 5000 births. They comprised 8% of all major congenital anomalies and are generally more common on the left side, with approximately 80% being left sided and 20% right sided. There is a male predominance with a ratio of 3:2. The etiology of congenital diaphragmatic hernia remains unclear. It is thought to be multifactorial with genetic, environmental, and/or nutritional factors playing a role. The septum transversum and pleuro-peritoneal membranes are the main components in the development of the diaphragm, which is completed by the 12th week of gestation. Any disturbance in the formation of the pleuro-peritoneal membranes can result in diaphragmatic discontinuity and congenital diaphragmatic hernia.

The incidence of Bochdalek hernia associated with intrathoracic kidney is reported to be less than 0.25% with only six case reports of concomitant, right-sided intrathoracic kidney and Bochdalek hernia published in the medical literature thus far. Many cases of left sided Bochdalek hernia have been reported in Nigeria in the literature but none is associated with intrathoracic kidney. A rare coexistence of intrathoracic kidney with right sided Bochdalek hernia in a neonate is therefore presented.

Case Summary
KU is a 2-weeks old male neonate delivered via spontaneous vaginal delivery at term to a
A 27-year-old mother of three in an unsupervised pregnancy. The delivery was also not supervised. He presented as a referral to paediatrics emergency unit of Aminu Kano Teaching Hospital (AKTH), Kano, Nigeria on account of difficulty in breathing since birth with associated on and off episodes of cough. Patient had no gastrointestinal symptoms. On examination there was normal chest symmetry and equal chest movement on both sides. He was however tachypneic with respiratory rate of 60 cycles/min and had mild intercostal retraction and decreased air entry on the right side. Percussion notes were dull on same side with gurgling sound heard on auscultation. Auscultation revealed crepitations on the right mid and lower lung zones. He was diagnosed to have failure to thrive secondary to chest infection.

Postero-anterior chest radiograph showed slightly elevated right hemidiaphragm associated with inhomogeneous area of opacity with tubular appearance and air-fluid levels in the right hemithorax mimicking a cystic structure. (Fig. 1).

A supplementary barium meal done to further clarify the nature of the mass demonstrated barium coated bowel loops in the thoracic cavity which were localized to the postero-inferior aspect of the right hemithorax above the hemidiaphragm (Figs 2 and 3). So, a diagnosis of right-sided congenital diaphragmatic hernia of the Bochdalek type was made.

Abdominal ultrasound scan revealed absent right kidney. The left kidney was seen in its normal position. Extended ultrasound scan to the chest revealed the right kidney to be located in the right hemithorax adjacent to the heart (fig. 4). No hydronephrosis was seen.

The patient was referred to the paediatric surgeon who admitted him for surgery. At surgery, he was found to have a posterior diaphragmatic defect with herniating loops of bowel into the posterior mediastinum. The herniating loops of bowel and the right kidney were reduced into the abdominal cavity with subsequent repair of the diaphragmatic defect.

The patient has been stable with no post-op complications and has been on follow-up for 8 months and was found to be doing well. The parents were then advised on the importance of antenatal care/visits in the mother’s subsequent pregnancies and deliveries.

Figure 1: Frontal chest radiograph showing an ill-definition of right hemidiaphragm with multiple cyst-like structures occupying the entire right hemithorax and associated mediastinal shift to the right.

Figure 2: Barium meal and follow through, antero-posterior projection, demonstrating contrast filled bowel loops in the right hemithorax
Discussion

Bochdalek hernia is a congenital posterior lateral diaphragmatic defect that allows abdominal viscera to herniate into the thorax, resulting from failed closure at 8 weeks of gestation of the pleuro-peritoneal ducts, primitive communications between the pleural and abdominal cavities. It is more common in infants (90%) with an incidence of 1/2500 live births; however, it occurs most frequently on the left side with approximately 80% being left-sided and 20% right-sided. This is presumably due to the pleuro-peritoneal canal closes earlier on the right side, or to narrowing of the right pleuro-peritoneal canal by the caudate lobe of the liver.

Herniated organs are frequently the omentum, bowel loops, spleen, stomach, kidney, and pancreas on the left, and part of the liver on the right. As in the present case in which bowel loops and right kidney herniated. Intrathoracic kidney is a rare developmental anomaly representing <5% of all renal ectopias. Ectopic intrathoracic kidney is slightly common on the left side, in our case it was on right side. The incidence of Bochdalek hernia associated with intrathoracic kidney is reported to be <0.25% with most of the cases occurring on the left side. The incidence of concomitant, right-sided intrathoracic kidney and Bochdalek hernia is even rarer with only two case reported in adults, three cases in children and two prenatal case thus far. In all cases, the kidney is located within the thoracic cavity and not in the pleural space. In spite of these abnormalities, intrathoracic kidney is usually fully functional and does not exhibit dysplasia, contralateral hypertrophy, or obstruction of the lower urinary tract. Because of the pulmonary hypoplasia due to the compression of the lungs by the adjacent hernia, these patients are frequently symptomatic at birth. Thus early- and late-presenting Bochdalek hernia may lead to different clinical presentations. Respiratory symptoms seem to be associated more with early Bochdalek hernia and gastrointestinal symptoms with late-presenting Bochdalek hernia. In our patient, tachypnoea associated with failure to thrive was reported. In majority of individuals, the thoracic kidney is benign and asymptomatic as in our case. Surgery is only necessary in case of ureteral obstruction or vesicoureteral reflux.

Figure 3: Barium meal and follow through, lateral projection demonstrating the herniated bowel loops in the posterior aspect of the thorax.

Figure 4: Longitudinal B-mode sonogram showing the right kidney (vertical arrows) being adjacent to the heart (horizontal arrows) in the thorax.
Conclusion:
Chest radiography, barium studies of the upper gastrointestinal tract and abdominal ultrasonography are very important imaging modalities in the evaluation of patients with suspected diaphragmatic anomalies as this saved the life of our patient.

References

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