http:// ijp.mums.ac.ir
Case Report (Pages: 489-492)

Silent Tachypnoea in a Neonate: A Rare Presentation of Right Side Bochdalek Hernia with Intrathoracic Kidney

*Shilpa Kalane [1], Umesh Vaidya[1], Pradeep Suryawanshi[2], Shashank Shrotriya[3]

[1]Consultant Neonatologist, Sahyadri Speciality Hospital, Nagar road, Pune, India.
[2]Senior Consultant Neonatologist, Sahyadri Speciality Hospital, Nagar road, Pune, India.
[3]Consultant Pediatric Surgeon, Sahyadri Speciality Hospital, Nagar road, Pune, India.

Abstract

Congenital Diaphragmatic Hernia (CDH) is a rare condition. The reported incidence of intrathoracic renal ectopia due to CDH is also rare. A right-sided thoracic kidney is much less common due to the location of the liver. Isolated intrathoracic kidney is usually asymptomatic and diagnosed incidentally on chest imaging. The authors report on a 21-day- old- female infant with late-presenting right sided congenital diaphragmatic hernia associated with intrathoracic ectopic kidney and adrenal gland. Prenatal ultrasound showed no pathology. A female baby was investigated for silent tachypnoea, a chest X-ray confirmed the diagnosis of congenital diaphragmatic hernia, the postnatal ultrasound revealed a right-sided kidney herniation. On Computed Tomography (CT) scan intrathoracic ectopic kidney was diagnosed. CDH is a delivery room emergency. This case had complex anatomy of right side CDH and intrathoracic kidney and presentation was only tachypnoea. Hence we are reporting this case.

Key Words: Diaphragmatic-hernia, Kidney, Neonate, Thoracic.

***Corresponding Author*:**

Dr. Shilpa Kalane, Flat no. 202, G Building, Wondercity, Katraz-S.N.76, Pune-46, Maharashtra, India.
Email: drshilpakalane@gmail.com

Received date: March 11, 2015 ; Accepted date: March 22, 2015

Introduction

Bochdalek hernia is a congenital posterior lateral diaphragmatic defect, which allows abdominal viscera to herniate into the thorax. Intrathoracic ectopic kidney accounts for 5% of all renal ectopias and its association with congenital diaphragmatic hernia has been reported to have an incidence of only 0.25%. The reported incidence of intrathoracic renal ectopia due to CDH is 0.25%.

A right-sided thoracic kidney is much less common due to the location of the liver. This condition is common in males and in 80–90% on the left side (1, 2). We report a case of a female infant who had a right thoracic kidney associated with right Bochdalek hernia.

Case Report

Our female infant was born at full-term by uneventful vaginal delivery with a birth weight of 2,400 g (Head circumference 35 cm, total length 50 cm). The prenatal ultrasound showed no pathological result. Baby had uneventful postnatal history and was with mother on breast feeding. However, the newborn attracted attention by mild tachypnoea and failure to thrive noticed on day 20 of life.

Baby was referred to our hospital for further management on day 21 of life. Clinical examination revealed mild tachypnoea with 90% saturations on room air. Baby needed oxygen, had decreased air entry on right side.

Conventional chest X-ray showed mediastinal shift to the left side and a raised left dome of the diaphragm with gas shadows in the right lung field, suggestive of Congenital Diaphragmatic Hernia (CDH) (Figure.1).

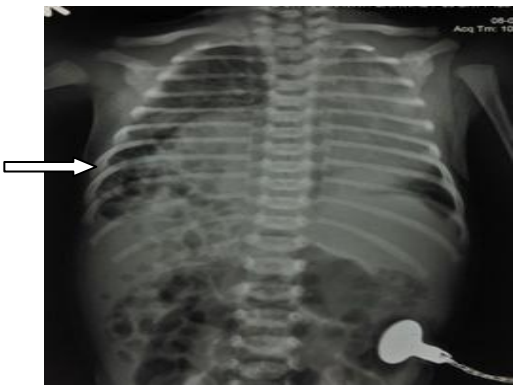

Fig. 1: Conventional anteroposterior chest X-ray demonstrating air-filled loops of the bowel in the right hemithorax and a paucity of gas in the abdomen. The right diaphragm is not discernable and there is an extensive mediastinal shift to the left site.

Echocardiography was normal. Preoperative sonography abdomen revealed herniation of right kidney. To confirm CT with CT angiography to look for vascular supply of both kidneys was done which revealed herniation of small bowel loops along with right kidney in right posterior hemithorax through defect in posterior part of diaphragm. Right renal artery and vein appear stretched (Figure.2).

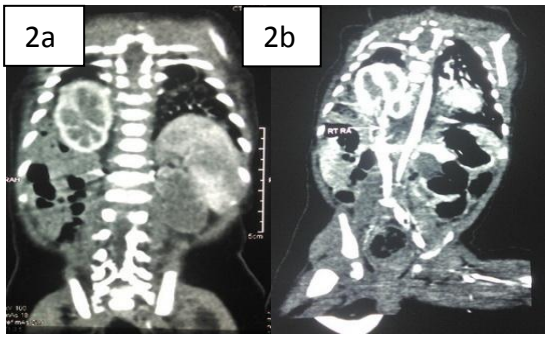

Fig. 2: CT scan showing Herniation of small bowel loops along with right kidney in right posterior hemithorax through defect in posterior part of diaphragm (a). Right renal artery and vein appear stretched on CT angiogram (b)

Immediately performed surgery demonstrated a true right side posterolateral congenital diaphragmatic hernia (Bochdalek type) without hernial

sac. The right kidney and adrenal gland, the majority of small intestine were found to be protruding into the right chest cavity (Figure.3). Intestinal malrotation was excluded. After replacement of the organs into the abdominal cavity, the diaphragmatic defect was closed by primary repair. The right kidney was mobilized without difficulty in a near to normal site. Opening the renal fascia, the kidney showed no deformed shape and no abnormal rotation, as well as a normal age-related size and a normal abdominal origin of renal vessels.

After primary closure of the abdominal wall, the postoperative course was uneventful, and the patient was discharged on the day 30 of exclusive breast feeding. Postoperative ultrasound demonstrated persistent correct position of the right kidney and a normal renal perfusion without any signs of urinary obstruction.

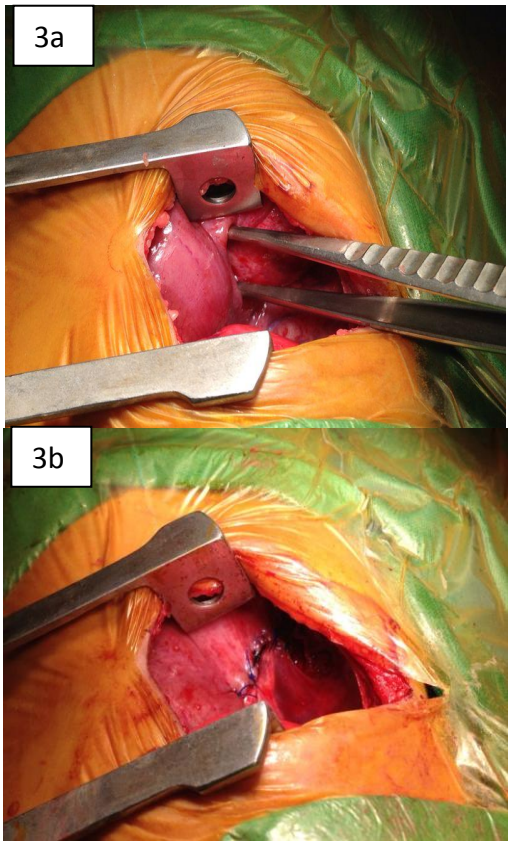

Fig.3: 3a and 3 b reveal intraoperative closure of diaphragmatic hernia by primary repair

Discussion

The posterolateral defect in the diaphragm through which abdominal organs might herniate into the thorax was first described by Vincent Alexander Bochdalek, a Czech anatomist in 1848, hence bears his name Bochdalek's hernia. It occurs more frequently on the left side. This is presumably due to the fact that the pleuroperitoneal canal closes earlier on the right side, or due to the narrowing of the right pleuroperitoneal canal by the caudate lobe of the liver (1, 3). It is common in males than in females in the ratio 2:1 (4). In our case it was a female infant with right side CDH. The ectopic kidney is usually pelvic, iliac, abdominal, thoracic, contra lateral, or crossed (4). Ectopic kidney is slightly common on the left side, in our case it was on right side (2, 5). The intrathoracic location of an ectopic kidney is the rarest with a reported incidence of 5% of all ectopic kidneys (1). The association of a Bochdalek hernia and intrathoracic renal ectopia is even rarer at 0.25%.The ectopic kidney has been known to be associated with many other defects ranging from acromelic frontonasal dysplasia to Williams syndrome (5). However there were no other deformities in this patient.

In majority of individuals, the thoracic kidney is benign and asymptomatic as in our case. It is most often detected incidentally on chest imaging or at the time of thoracotomy for a suspected mediastinal tumor (6). In such patients the symptoms and X-ray picture might be easily confused for pleuritis, pulmonary tuberculosis, or pneumothorax (1, 7). Chest radiographs are not appropriate, CT scan and Magnetic Resonance Imaging (MRI) would be better for diagnosing diaphragm defects (8). Intra-venous pyelography or renal scintigraphy usually clarifies the diagnosis. As such an incidentally detected intrathoracic kidney which is asymptomatic does not require

any active intervention. Its clinical significance lies in its potential to be confused as a pathological thoracic mass leading to its erroneous excision (9). Thus early and late-presenting CDH may lead to different clinical presentation. Respiratory symptoms seem to be associated more with early CDH and gastrointestinal symptoms with late-presenting CDH (10). In our patient, tachypnoea associated with failure to thrive was reported. Diagnosis of both these conditions thoracic ectopic kidney and Bochdalek hernia can be reached if and only if there is a high degree of suspicion on the part of the treating physician.

In conclusion, CDH is an emergency in delivery room. Right side CDH has poor prognosis compared to left. In present case, silent tachypnoea was a presentation of this rare anatomy of right side CDH with intrathoracic kidney.

Conflict of interest: None.

References

1. Fiaschetti V, Velari L, Gaspari E, Mastrangeli R, Simonetti G. Adult intra-thoracic kidney: A case report of bochdalek hernia. Case Report Med 2010; 2010:pii 975168.
2. Motiani PD, Gupta PK, Aseri KC, Agarwal KC. Bochdalek hernia on the right side in an adult - case report and review. Lung India 1991;9(2):63–5.
3. Tubbs RS, Gribben WB, Loukas M, Shoja MM, Tubbs KO, Oakes WJ. Franz Kaspar Hesselbach ,1759-1816: Anatomist and surgeon. World J Surg 2008; 32(11):2527–9.
4. Mensah YB, Forson C. Left thoracic kidney: A rare finding at intravenous urography. Ghana Med J. 2010; 44(1):39–40.
5. Woodward M, Evans K. Abnormalities of migration and fusion of the kidneys. Oceanan Therapeutics Ltd. 2011. Available at: http://www.Pediatricutricularologybook.com/kidney fusion.html. Accessed on 2011 Dec 8.
6. Drop A, Czekajska-Chehab E, Maciejewski R, Staśkiewicz GJ, Torres K. Thoracic ectopic kidney in adults. A report of 2 cases. Folia Morphol (Warsz) 2003; 62(3): 313–6.
7. Ramaprasad G, Vidyasagar B. Bochdaleks hernia simulating pleural effusion. Lung India 1989; 7(2): 95–6.
8. Mirza B, Bashir Z, Sheikh A. Congenital right hemidiaphragmatic agenesis. Lung India 2012; 29(1): 53–5.
9. Hithaishi C, Padmanabhan S, Kumar BS, Kalawat TC, Venkatarammappa M, Siva Kumar V. Ectopic thoracic kidney- a case report. Indian J Nephrol 2003;13: 79–80.
10. Chang SW, Lee HC, Yeung CY, Chan WT, Hsu CH, Kao HA, et al. A twenty-year review of early and late-presenting congenital Bochdalek diaphragmatic hernia: are they different clinical spectra? Pediatr Neonatol 2010; 51(1):26–30.