

Coexistence of Diaphragm Eventration and Thoracic Ectopic Kidney

O OZTURK, Y YILDIZ, M IGDE, BG OKSUZ

Abstract

Diaphragm eventration is partial or complete elevation of the diaphragm as a result of failure of muscular development. While diaphragm eventration has no symptoms in some cases, it may also cause life threatening problems. Thoracic ectopic kidney is a rare anomaly and usually does not produce symptoms. A 4-month-old male patient who has diaphragm eventration with left ectopic thoracic kidney was described in this case report.

Key words

Diaphragm; Ectopic kidney; Eventration; Thorax

Introduction

Diaphragm is an anatomic structure consisting of aponeurosis and muscle which separates thoracic from abdominal cavity. The highest point of the diaphragm is usually located at the 5th costal level on the right side and 6th costal level on the left side. Diaphragm is the most important muscle of respiration. While volume of thoracic cavity increases by its contractions during inspiration, volume of abdominal cavity decreases simultaneously. Moreover, diaphragm plays a significant role in the mechanism of coughing, sneezing, shouting, laughing, and vomiting. During embryonic development of 4th to 12th weeks of gestation, diaphragm arises gradually from septum transversum and thoracic wall.

Diaphragm eventration is defined as elevation of all or some parts of diaphragm because of defect in the diaphragmatic structure.¹ Eventration can impair ventilation of the lung alveoli leading to recurrent infection with potential life threatening outcome. On the other hand, ectopic kidney within the thoracic cage is an unrelated anomaly which is very rare.² Due to its asymptomatic nature, it is often detected incidentally while one is investigating for other unrelated pathological conditions. In our study, we report an infant with coexistence of diaphragm eventration and thoracic ectopic kidney.

Case Report

A 4-month-old male infant was brought to emergency department because of tachypnoea and pallor. Patient was the 7th lived birth of a 27-year-old mother with 9 pregnancies. All of his siblings were normal except that the 4th pregnancy was terminated because of oligohydramnios. He was delivered at 30th week of pregnancy by normal spontaneous vaginal delivery with a birth weight of 1250 gm. His parents are non-consanguineous and both are healthy. His antenatal and postnatal courses were uneventful. Patient had no cyanosis at birth but then he was admitted to the newborn intensive care unit with the diagnosis of "respiratory distress syndrome" shortly after till 20-day-old. He was eventually discharged from hospital when he reached 2000 gram. The

Department of Family Medicine, Samsun Education and Research Hospital, Samsun, Turkey

O OZTURK MD

Department of Pediatrics, Samsun Education and Research Hospital, Samsun, Turkey

Y YILDIZ MD

M IGDE MD

BG OKSUZ MD

Correspondence to: Dr O Ozturk

Received March 5, 2014

baby's development was satisfactory and after discharge, he did not have any problem until this acute respiratory distress episode.

On physical examination his general condition was poor. Heart rate was 156 beats/min, respiratory rate 46 beat/min, oxygen saturation was 85% in room air with sign of cyanosis. His oropharynx, nose and ears were normal. There was no choanal atresia or thoracic deformity. But heart sounds were heard on the right side of the thorax with no murmur detected. His thorax did not move equally during respiration. No breath sound could be heard on the left side and it was mixed with the heart sounds on the right side. There were no rales or rhonchi. Femoral arterial pulses were present. He had a scaphoid abdomen and the contents could not be evaluated fully. The other findings were within normal limit.

Laboratory investigations showed that haemoglobin was 11.1 gm/dL, white blood cell was $15.39 \times 10^9/L$, platelet was $609 \times 10^9/l$. Serum electrolytes, kidney functions and blood glucose were normal and C-reactive protein count was 20.9 mg/dL. Blood gas analysis showed respiratory acidosis of pH: 7.19, pO_2 : 40.4 mmHg, pCO_2 : 79.3 mmHg, HCO_3^- : 29.8 mEq/l, BE: 0.3 mmol/L and oxygen saturations were 60.8%. There was no growth in blood culture.

In posteroanterior chest radiography, intestinal

segmental gas pattern simulated large bowel haustration was found filling up the left hemithorax up to the hilus level. This caused displacement of heart and mediastinum to the right. A continuity between left upper abdominal quadrant with the left hemithorax was apparent (Figure 1). Contrast study with radiopaque agent revealed that displaced intestinal segments filled with localised opaque agent within the left hemithorax. Such displacement pushed the heart and mediastinum to the right. Appearance was consistent with diaphragm eventration (Figure 2).

Patient was referred to paediatric surgery for operation and during the procedure, it was found that pancreas, intestines, colon, stomach and left kidney were displaced to the left hemithorax. There was no hernia because diaphragm was intact. Pancreas, intestines, colon and stomach were pulled back to the abdomen. However, the normal size left kidney was impossible to relocate to the retroperitoneal area due to inadequate space available. The left lung was not hypoplastic. Primary repair was performed by plication of the diaphragm. Because of its structure similar to diaphragm eventration and there was no posterior rim, 2-0 prolene steered through the costal periosteum and the anterior rim with 4 pieces of U suture was performed. No postoperative complication was noted.

In postoperative abdominal ultrasonography, right

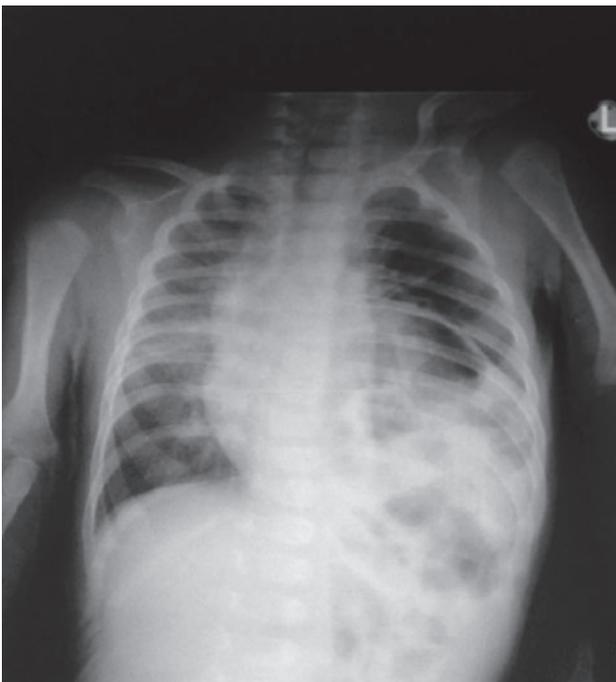


Figure 1 Diaphragm eventration, mediasten shift and bowel loops.



Figure 2 Intestinal segments filled with opaque agent on left hemithorax.

kidney was 48 mm, left kidney was 37 mm in vertical length. Left kidney was located above the spleen in the thorax and could only be evaluated between ribs with limited assessment. Parenchyma echos of both kidneys were normal. In kidney scintigraphy, the right kidney was within normal limits and the left kidney was located within the thorax.

Postoperative chest radiograph showed residual eventration of diaphragm but the patient's general condition was good, respiration and oxygen saturations were normal and there was no need for an urgent surgery. He was followed up 2 weekly at outpatient clinic and after 2 months, second operation would be arranged.

Discussion

Diaphragm eventration is defined as abnormal elevation of all or some parts of diaphragm towards the thoracic cavity¹ with a prevalence of about 5/10,000 live births. It is important but difficult to differentiate diaphragm eventration from diaphragmatic paralysis. Eventration is a congenital condition but paralysis is mostly acquired.³ There is no clear agreement for the diagnosis criteria of diaphragm eventration. Developmentally, it is a consequence of deficient or insufficient development of pleuroperitoneal membrane in fetal life (8 to 10 weeks). It accounts for 5% of all diaphragm anomalies.⁴ It is more common in male and involvement of the left hemidiaphragm is relatively more than the right.⁵ There is a significant decrease in the strength of the diaphragm muscle fiber. Eventration has 3 anatomic forms: partial, complete and bilateral.⁶ Partial form is usually seen at right hemidiaphragm anteromedially; on the other hand complete form is usually localised at left hemidiaphragm. Acquired diaphragm eventration is related to phrenic nerve injury. Neuromuscular disorders or adjacent organ lesions may cause acquired diaphragm eventration by altering diaphragm levels.

A thorough examination to search for congenital anomalies had not been performed after delivery for this patient. But by the imaging studies, thoracic ectopic left kidney was spotted. This location is the rarest form of ectopic kidney. Chung et al found 30 cases of ectopic kidney among 132,686 healthy school children by using ultrasound screening to investigate the frequency of renal anomalies. Only one of these cases had ectopic kidney in the thoracic cavity.⁷ Embryology of intrathoracic ectopic kidney is not clear. Normally, it is described as a consequence of

excessive migration of kidneys from sacral to lumbar region which must be completed by eighth week of pregnancy. There are various postulations on the pathogenesis but the most accepted hypothesis is that it is a consequence of a developmental disorder of pleuroperitoneal membrane.⁸ Usually it is asymptomatic and cases were often diagnosed incidentally by chest radiograph performed for other causes. Therefore, its actual incidence remains unknown. It is seen mostly on the left side (61%) as in this index case but it can also be found on the right side (36%) or even both sides (2%).⁹

Symptomatic diaphragm eventration seen in newborns period may lead to life threatening consequence such as severe dyspnoea and hypoxia, decrease in lung volumes, atelectasis, ventilation/perfusion mismatch, or mediastinal shift as seen in this patient. Exclusion of other physiologic and pathologic conditions are essential before considering eventration. In normal condition, because of the bigger size liver is located at the right side, diaphragm is therefore 1-2 cm higher than the left. On the left side, because of being adjacent to stomach fundus and spleen, diaphragm may be elevated when abdominal distension occurs.

Pathological conditions such as iatrogenic damaged of phrenic nerve; phrenic nerve paralysis as a sequels of poliomyelitis, diphtheria or syphilis; aortic aneurysm compression; substernal thyroid compression; or diaphragmatic hernia; paraesophageal hernia; hiatus hernia; bronchogenic cyst; pericardial cyst; epidiaphragmatic abscess; infected haematoma or teratoma; cardiac sarcoma; lymphosarcoma; and schwannoma. For diagnosis, postero-anterior chest radiography and fluoroscopy, computed thorax tomography, magnetic resonance imaging, respiratory function tests and ultrasonography can be used. The diagnosis of this case is established by bilateral lung radiographs and colon radiographs with barium. In cases where paradoxical movement of diaphragm is observed due to eventration and causing respiration problem, surgical treatment is indicated and the best approach is diaphragm plication. In this method, hemidiaphragm is strained, lung compliance is increased and mediastinum is stabilised.¹⁰ Being congenital or acquired eventration does not affect the surgical approach.

Conclusion

In patients who have laboured breathing and tachypnoea during infancy, one should consider diaphragm eventration as one of the differential diagnosis and radiological

investigations can help to establish the diagnosis. In patients with eventration, the presence of mediastinal masses found in imaging should prompt the consideration of a thoracic kidney. Furthermore, organ dysfunction may occur because of atypical locations. Operation may be the only curative option when necessary.

Conflict of Interest

None

References

1. Arensman RM, Bambini DA. Congenital diaphragmatic hernia and eventration. In: Ashcraft KW, Murphy JP, Sharp RJ, Sigalet DL, Snyder CL, editors. *Pediatric Surgery*. 3rd ed. Philadelphia: W. B. Saunders; 2000;300-17.
2. Yalcinbas K, Sasmaz H, Canbaz S. Thoracic left kidney: a differential diagnostic dilemma for thoracic surgeons. *Ann Thorac Surg* 2001;72:281-3.
3. Efthimiou J, Butler J, Woodham C, Benson MK, Westaby S. Diaphragm paralysis following cardiac surgery: Role of phrenic nerve cold injury. *Ann Thorac Surg* 1991;52:1005-8.
4. Tsugawa C, Kimura K, Nishijima E, Muraji T, Yamaguchi M. Diaphragmatic eventration in infants and children: is conservative treatment justified? *J Ped Surg* 1997;32:1643-4.
5. McNamara JJ, Paulson DL, Urschel HC, Razzuk MA. Eventration of the diaphragm. *Surgery* 1968;64:1013-21.
6. Jawad AJ, al-Sammarai AY, al-Rabeeah A. Eventration of the diaphragm in children. *J R Coll Surg Edinb* 1991;36:222-4.
7. Sheih CP, Liu MB, Hung CS, Yang KH, Chen WY, Lin CY. Renal abnormalities in schoolchildren. *Pediatrics* 1989;84:1086-90.
8. Eren S, Polat B. Congenital intrathoracic ectopic kidney. November 2003. <http://www.eurorad.org/eurorad/case.php?id=1895> DOI: 10.1594/EURORAD/CASE.1895
9. Liddell RM, Rosenbaum DM, Blumhagen JD. Delayed radiologic appearance of bilateral thoracic ectopic kidneys. *Am J Roentgenol* 1989;152:120-2.
10. Oguzkaya F, Bilgin M, Akcali Y, Kahraman C, Sahin A. Eriskinlerin tek taraflı diyafragma paralizisi ve evantrasyonunda diyafragma plikasyonu. *Turk Gogus Kalp Damar Cerrahisi Dergisi* 1998;6:163-7.