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Article / Autopsy Case Report

Thoracoabdominal compartment syndrome complicating right-sided diaphragmatic eventration with co-existent unilateral renal agenesis

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ABSTRACT

Diaphragmatic eventration (DE) associated with intestinal malrotation and renal agenesis is a rare entity. The authors report a case of a 69-year-old man who had symptoms of heart failure. He had a previous imaging diagnosis of right diaphragmatic eventration and dilated cardiomyopathy. He died on the second day after the hospital admission and had a post mortem examination that confirmed complete right diaphragmatic eventration, intestinal malrotation, left renal agenesis, dilated cardiomyopathy, and anteriorly rotated right kidney and had findings suggestive of a thoracoabdominal compartment syndrome. Thoracoabdominal compartment syndrome is described as transmission of abdominal pressure through a defective diaphragm causing compression of the hemithorax viscera and mediastinal shift with a hemodynamic alteration. The association of these anomalies is rare, and the possibility of this finding in a patient with eventration should always be considered.

Keywords
Diaphragmatic Eventration; Compartment Syndrome; Hereditary renal agenesis.

INTRODUCTION

The diaphragm is a significant respiratory muscle that also serves as a barrier between the thoracic cavity and the abdominal cavity. It can be affected by different congenital or acquired entities.[1] Diaphragmatic eventration (DE) is an infrequent entity. It can be a result of the intrinsic weakness of the diaphragmatic muscles or secondary to phrenic nerve palsy, pulmonary, or subphrenic diseases.[2] Congenital DE can be either complete agenesis of the diaphragm or partial loss with a persistent thin fibrous tissue usually at the posterior wall.[3,4] DE can either be unilateral (hemidiaphragmatic) or bilateral. However, right DE seems to be the most common presentation.[4-8] DE is usually accompanied by poor lung expansion, reduced blood flow, reduced oxygen and carbon monoxide exchanges, and upward displacement of the abdominal contents. A few cases have been associated with other malformations such as intestinal malrotation, dextrocardia, renal agenesis, and ectopia.[2,5,8] Protrusion of colon into the thorax resulting in unilateral lung collapse, mediastinal shift, and features similar to tension pneumothorax have also been described and referred to as the thoraco-abdominal compartment syndrome.[9] This

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syndrome is believed to be a result of the transmission of the high abdominal pressure into the thoracic cavity ending up in physiologic and anatomic disruption.[9]

Most adult patients with DE complain of respiratory symptoms, while others with associated mal-rotation of the gut also present subtle gastrointestinal symptoms. Renal agenesis and renal ectopia are sporadic findings in association with DE.[10] Sharma et al.,[8] described a case of renal agenesis and intestinal mal-rotation but without a thoracoabdominal compartment syndrome.[8]

We report a case of DE associated with other rare malformations complicated by thoracoabdominal compartment syndrome.

CASE REPORT

A 69-year-old man has been attending several medical appointments, at different hospitals, over the last year complaining of easy fatiguability, orthopnea, paroxysmal nocturnal dyspnea, pedal swelling, and worsening breathlessness. He presented to the emergency room of University College Hospital – Ibadan, with worsening of these symptoms, including early satiety and abdominal pain over the last three weeks. His past medical history included the diagnosis of a right DE and atelectasis of the right lung and heart failure with reduced ejection fraction, much probably due to dilated cardiomyopathy. He was not previously known with hypertension, diabetes mellitus; neither was he aware of any family history of such diseases. He drank sparingly alcoholic beverages in the past, and never smoked tobacco. An echocardiogram done at diagnosis showed eccentric left ventricular hypertrophy, dilated left atrium 5.0 cm (RR 3.0-4.0 cm), and left ventricle 6.5 cm (RR 4.5-5.9 cm), impaired LV systolic function (Ejection fraction of 26.8%) with a restrictive pattern of diastolic dysfunction and marked tricuspid regurgitation. ECG report at diagnosis showed left ventricular hypertrophy (LVH), Left atrial enlargement, and inverted T waves in the inferolateral leads.

His cardiovascular examination showed normal volume and regular pulse of 118/min, blood pressure of 120/80mmhg, engorged neck veins, diffuse apex, with a pansystolic murmur in the tricuspid area that increased in intensity on inspiration. Fine basal crackles were heard in the left hemithorax, and tender hepatomegaly was palpable in the abdomen. He had dusky lower limbs with bilateral pitting pedal edema. The respiratory rate was 48/min, SaO2 (84-89%) in room air. A tracheal deviation to the left was noted, as well as a reduced chest expansion on the right hemithorax. There was dull percussion, and diminished breath sounds on the right hemithorax, while resonant percussion with basal crepitations was detected in the left hemithorax. A limited bedside echocardiography done at presentation showed dilated cardiac chambers with an estimated ejection fraction of 20-30% by eyeballing, flattened interventricular septum, eccentric tricuspid regurgitation and dilated inferior vena cava with less than 50% collapse on inspiration. No mural thrombi were seen. He was managed for heart failure with reduced ejection fraction secondary to dilated cardiomyopathy/idiopathic heart disease complicated with a respiratory infection and pulmonary hypertension with the provisional working diagnosis of pulmonary thromboembolism. He was placed on low dose ACE-Inhibitors, diuretics, mineralocorticoid antagonist and prophylactic anticoagulation. Broad-spectrum antibiotics were commenced on account of neutrophilic leukocytosis (white blood cell count of 12,400/mm^3 (Reference range [RR] 4 -11,000/mm^3) on complete blood count. Hematocrit and hemoglobin, as well as the renal function, and electrolytes, were within the normal range. His clinical status deteriorated with severe respiratory distress. The patient died within 36 hours of admission.

Autopsy Findings

The postmortem examinations showed an asthenic looking older man measuring 175 cm in length. He was not pale, anicteric; he had mild peripheral cyanosis and bilateral pitting pedal edema up to the mid-third of the legs. His abdomen was scaphoid. At the opening of the cavities, the right hemidiaphragm eventration was depicted. The liver, segments of the small intestine, and transverse colon were within the right thoracic cavity (Figure 1).

The heart was enlarged and weighed 465g (reference range [RR] 300-350g) with biventricular hypertrophy and chambers dilation; the tricuspid valve measured 13.8 cm (RR; 10-12 cm) and the mitral valve measured 11 cm (RR; 8-10.5 cm). The pulmonary valve

and aortic valve were within normal ranges. The right ventricular wall thickness measure 0.7 cm (RR; 0.2-0.5) and the left ventricular wall thickness measure 1.8 cm (RR; 1-1.5 cm). The histology of the heart showed myocyte hypertrophy with marked interstitial fibrosis (Figure 2).

The left and right coronary arteries and the branches were patents. The right lung was found to be atelectatic weighing 200grams (RR; 360-570g) with mediastinal shift to the left (Figure 3). The right main bronchus contained thick mucoid secretions, and cut surfaces of the right lung showed fibrosis with significant atherosclerosis of the intraparenchymal vessels consistent with pulmonary hypertension (Figure 4). The left lung showed moderate edema and congestion, as well as features suggestive of pulmonary hypertension. Both lungs showed marked anthracosis. There was no embolus in the pulmonary trunk or its intimal branches. There was malrotation of the gut with the caecum lying in the midline 3 cm away from the pubic symphysis, with most of the small intestine and transverse colon located over the right hemidiaphragm (Figure 5A).

Only one kidney was found in the right lumbar region. The right kidney weighed 245g (RR; 115-220g) and was anteriorly rotated with prominent superficial lobulations (Figure 5B). The capsule stripped with ease to reveal a smooth subcapsular surface. The kidney's cut section showed accentuated cortico-medullary differentiation. The pelvicalyceal system and ureters showed no gross lesion.

The cause of death from the autopsy findings was the respiratory and hemodynamic repercussion of thoracoabdominal compartment syndrome due to the right hemidiaphragm eventration in a patient with coexistent dilated cardiomyopathy.

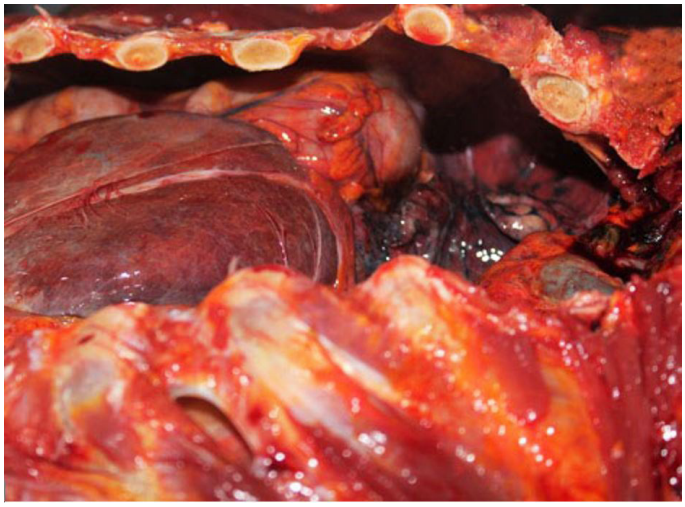

Figure 1. Gross view of the right thoracic cavity showing intrathoracic abdominal contents. Note part of the gut and the liver within the thoracic cavity.

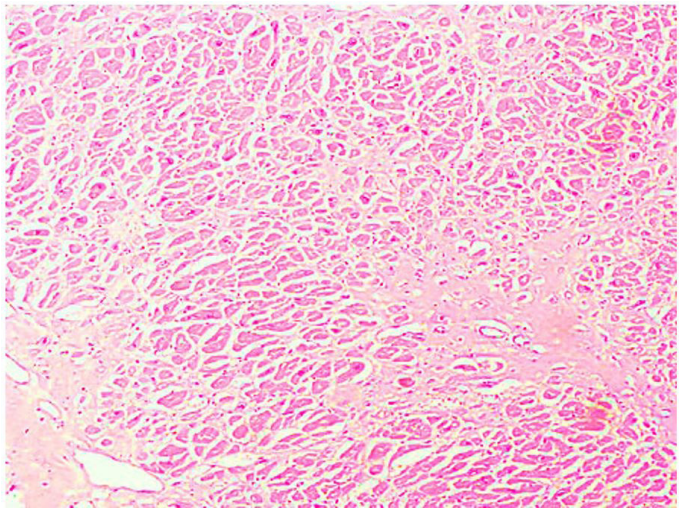

Figure 2. Photomicrograph of the myocardium (right ventricle) showing cardiomyocytes hypertrophy and interstitial fibrosis (H&E x10 objective).

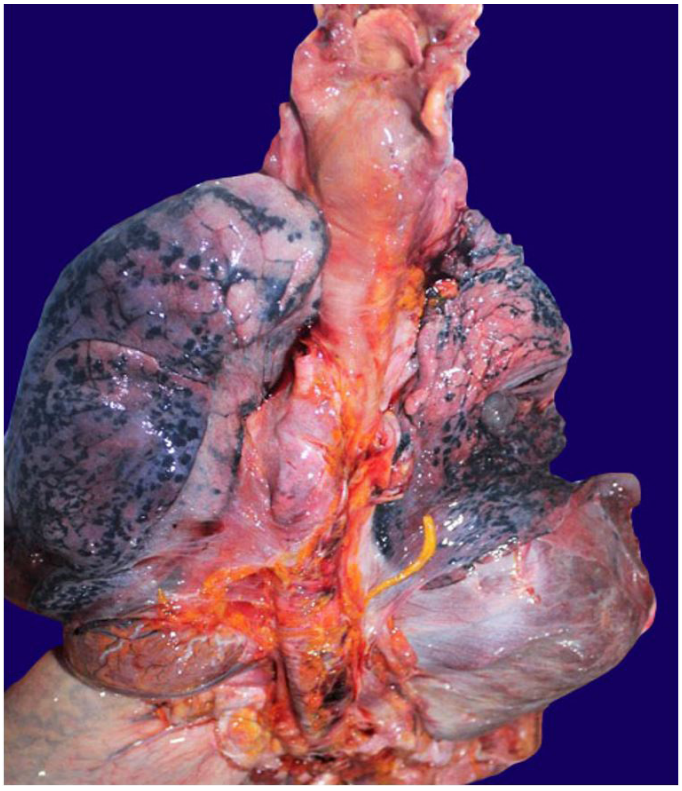

Figure 3. Ventral view of the thoracic organs showing the complete collapsed right lung with marked anthracosis.

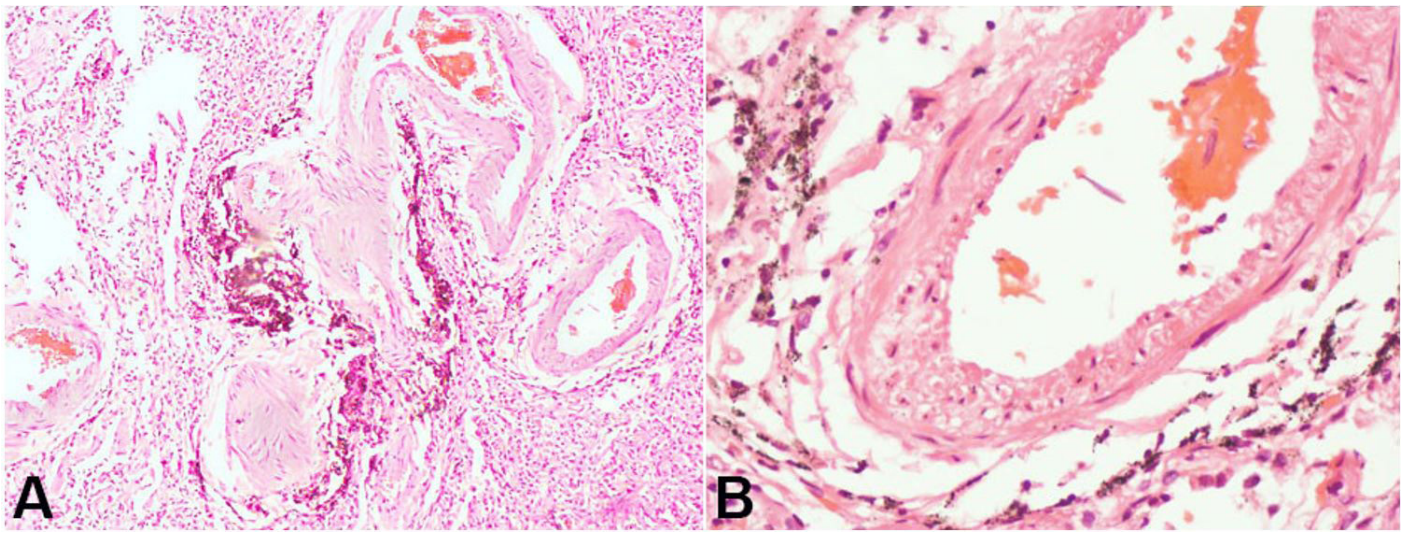

Figure 4. Photomicrographs of the lung showing in **A –** Thickening of vascular walls with the collapsed airways. (H&E x10 objective); **B –** Pulmonary artery showing atherosclerosis (H&E x40 objective).

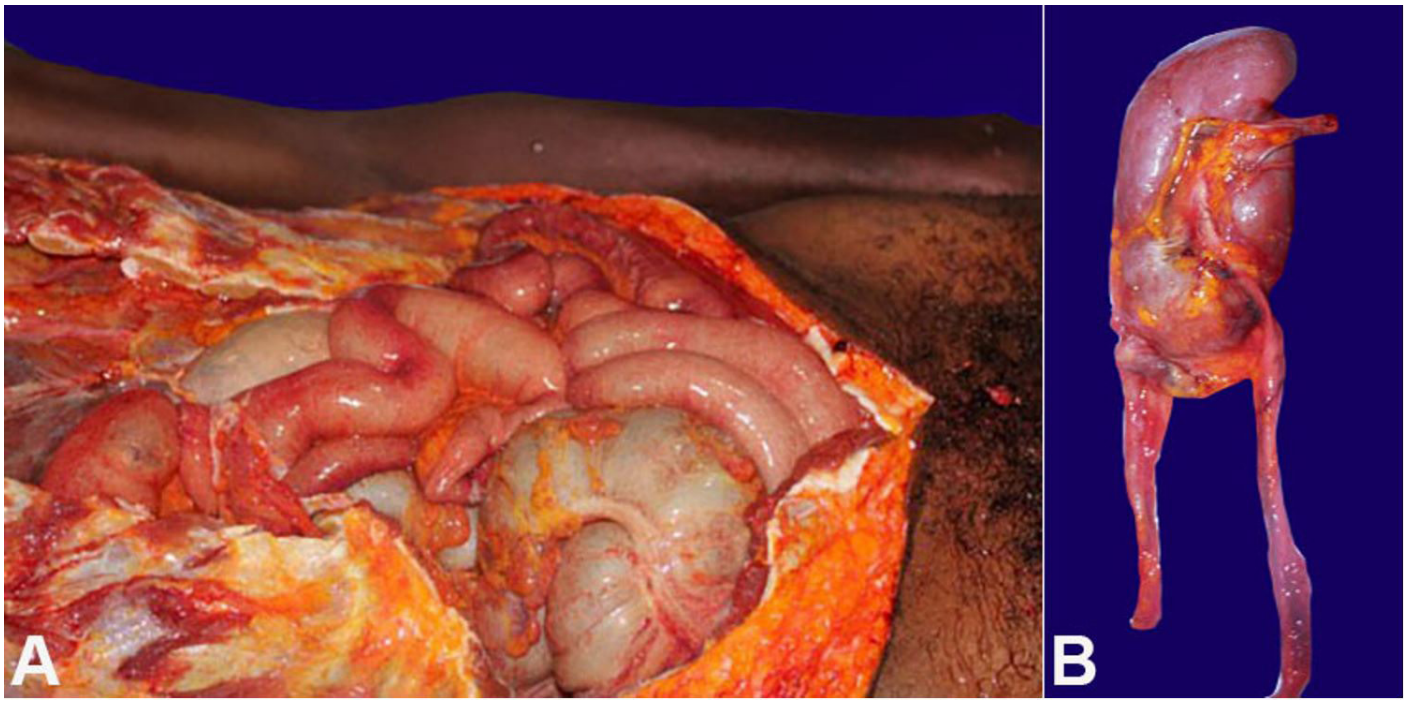

Figure 5. A – Gross view of the malrotation of the gut. The small intestine is majorly within the right hemidiaphragm; **B –** Gross view of the anteriorly located right kidney with prominent lobulations.

DISCUSSION

Complete lung collapse with mediastinal shift has been described as one of the complications of DE.[9] This was found in this index case; the total collapse of the right lung with the left mediastinal shift. The right hemithorax was filled with the liver, part of the gut, and the compressed remnants of the right lung. Pulmonary hypertension, noted in this case, could be secondary to the anatomical shift and the resultant effect on the pulmonary vessels and bronchus.[11] The compression of the lung by abdominal viscera can result in hypoplasia of a developing lung or lung collapse. In pulmonary hypoplasia, there is impaired homeostasis of the immature respiratory epithelium which leads to vascular remodeling (endothelial dysfunction, increased media thickness and increased adventitia thickness), and vasoconstriction, while in mediastinal diseases there is compression of pulmonary vessels caused by the abdominal content pressure against the atelectatic lung in this index case.[12-14] Both of these situations cause increased resistance to blood

flow resulting in pulmonary hypertension in a patient with DE. Pulmonary hypertension would worsen an already impaired heart function due to the existing dilated cardiomyopathy resulting in subsequent heart failure.[15] Also, the autopsy findings ruled out major coronaries' obstruction, valvopathy, and signs of small vasculopathy in the systemic circulation consistent with hypertension.

Eventration is complete or near-complete replacement of the diaphragm with a thin fibrous tissue with strands of muscles.[16] It is much more common on the right side, as was noted in this case report. DE can either be acquired or congenital, more often it is a congenital abnormality. His available premorbid and long-term medical history was sparse, presenting in the University College Hospital - Ibadan for specialized care based on verbal referral and because of his worsening clinical state. He died within 36 hours after admission. Hence, it is difficult to conclusively say if this was a congenital eventration, although the presence of other congenital abnormalities supports this suspicion.

DE is associated with some other abnormalities, such as intestinal malrotation, renal agenesis, and dextrocardia.[2,5,8] Dextrocardia that is commonly found in the very rare left-sided hemidiaphragm eventration was not found in this patient. This patient had a solitary kidney, which was anteriorly rotated and located in the lumbar area. Malrotation of the kidney is a congenital abnormality in which the developing kidney fails to rotate medially as it ascends to the lumbar area from the pelvis, mal-rotated kidney usually retains normal functions.[17,18]

CONCLUSION

DE of the hemidiaphragm can be complicated by thoracoabdominal compartment syndrome if left uncorrected. Thoracoabdominal compartment syndrome can lead to death by causing pulmonary hypertension complicated with chronic cor-pulmonale. This should be considered when a patient with eventration presents with features of heart failure as the symptomatology of both are similar.

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