



## Right Sided Eventration of Diaphragm with Corpulmonale-A Case Report

### KEYWORDS

diaphragmatic eventration, respiratory symptoms, corpulmonale, old age, incidental

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### ABSTRACT

*Eventration of the diaphragm is generally regarded as a condition in which the left or the right leaf of the diaphragm has ascended abnormally high into the chest. It is a condition in which all or part of the diaphragm is largely composed of fibrous tissue with only a few or no interspersed muscle fibers. It can be congenital or acquired. Congenital presents at an early age of life. In adults it is mostly asymptomatic and found incidentally on radiographs. If symptomatic, repeated upper respiratory tract infections are the most common findings associated with eventration of the diaphragm in older age groups. Here we present a case of 62 year old female with right sided eventration of diaphragm presented with corpulmonale which is a rare entity.*

### INTRODUCTION:

Eventration of the diaphragm refers to an "abnormal elevation of one leaf of an intact diaphragm as a result of paralysis, aplasia or atrophy of varying degrees of muscle fibres". The muscular insertions are normal, normal orifices sealed and there is no interruption of pleural or peritoneal layers.<sup>1</sup> It is most often an isolated entity and detected incidentally, mostly on left side and is asymptomatic. Eventration may be congenital or acquired. An eventrated diaphragm may result from congenital etiologies; muscular hypoplasia or aplasia. Acquired causes include phrenic nerve injury during breech delivery, operative trauma (crush, transection and hypothermia), fibrosis, inflammation and neoplasia Stauffer and Rickman (1972)<sup>2</sup>. Symptomatic adults have dyspeptic, respiratory and cardiac features. Although it is conceivable that secondary hypoplasia of the homolateral lung, usually presenting with severe cardiorespiratory symptoms leads to corpulmonale, there is no evidence substantiating this assumption. Here we present a rare case of right sided eventration of diaphragm with corpulmonale.

### CASE REPORT:

A 62 year old female presented with 1 year history of breathlessness and discomfort in the right chest for which she has not taken any medical advice. The breathlessness of the patient was gradually progressive from class III to class IV NYHA (New York Heart Association) and was more severe for the past 10 days with orthopnea and paroxysmal nocturnal dyspnea. This complaint was accompanied by swelling of feet and discomfort in right side of chest and upper abdomen. There is no history of trauma, palpitations, syncope, cough, abdominal distension, vomiting, pain abdomen. Patient denied any history of fever, any similar complaints since childhood or any surgeries. On physical examination she had pedal edema. Chest examination revealed decreased movements on right side in inframammary, infra-axillary, infra scapular areas. Tactile vocal fremitus was decreased and note was impaired to dull with decreased breath sounds on the same areas. A provisional diagnosis of right pleural effusion with heart failure was considered.

Laboratory investigations were within normal limits. Diag-

nostic pleural tap was dry without any fluid. Chest X ray PA view showed homogenous opacity in right lower and middle zones with sharp upper margin of opacity and contour of diaphragm preserved. Electrocardiograph showed right axis deviation, poor R wave progression. 2D echo revealed mild tricuspid regurgitation with pulmonary arterial hypertension with grade 1 diastolic dysfunction with a PR jet of 36 mm. X ray lateral view showed elevated right dome with preserved contour (figure:1). Ultrasonography showed congested liver with dilated hepatic veins and decreased movements of right hemidiaphragm and no paradoxical movements. Computed tomography of chest revealed elevated right dome of diaphragm and displacement of liver, right kidney and right colon with intact contour. (figure:2). There is no mediastinal lymphadenopathy. MRI abdomen was done and findings confirmed (figure:3). With all the above findings, eventration of right diaphragm with compromised right lung, lead to corpulmonale.

### DISCUSSION:

Eventration of diaphragm (ED) consists of thinned diaphragmatic muscle producing elevation of entire or part of the hemidiaphragm. Jean Louis Petit first recognized this disorder in 1774<sup>3</sup> and the term 'eventration' was coined by Becard in 1916<sup>4</sup>. Its incidence is about 1 in 10000<sup>5</sup> more common in males. It may originate from a congenital defect or may be acquired. Congenital diaphragmatic eventration is a consequence of incomplete or absent muscularization of the pleuroperitoneal membrane which may be due to early return of the midgut to the abdomen and is frequently associated with prematurity, chromosomal and other developmental abnormalities. Acquired diaphragmatic eventration is due to phrenic nerve pathology which may be caused by trauma, especially following instrumentation during delivery, neoplastic infiltration or compression, or following surgery.

ED can be partial, localized to a part of the hemidiaphragm (anterior, posterolateral, medial) or complete affecting the whole hemidiaphragm<sup>6</sup>. Complete eventration invariably occurs on left side but partial eventration occurs virtually on right side<sup>7</sup>. In this case, complete eventration was seen on right side which is a rarity. Congenital diaphragmatic eventra-

tion is often asymptomatic and may be an incidental finding on a chest radiograph, at thoracotomy or at post mortem. It may present in infancy with respiratory insufficiency which may require artificial ventilation or with recurrent chest infections. Macroscopically, the (affected portion of the) diaphragm is attenuated, abundant, membranous, without muscular appearance. Microscopically there is paucity or absence of muscular fibers and diffuse fibroelastic changes. Atrophy, thinned wall and progressive distension of diaphragm represent the main characteristics of diaphragmatic eventration.<sup>8</sup>

Congenital eventration can be associated with syndromes like Kabuki make up syndrome, Beckwith-Weidemann syndrome, Poland syndrome, Wandering spleen syndrome, Jarcho Levin syndrome, infections like fetal rubella, cytomegalovirus infection, trisomies, chromosomal abnormalities and congenital anomalies like pulmonary hypoplasia, congenital heart disease, tracheomalacia, cerebral agenesis, renal ectopia, malrotation, deformities of pinna, Meckel's diverticulum, Werdnig Hoff man disease (a neurological cause of congenital eventration).<sup>9</sup>

Clinically it is typically asymptomatic, but can cause dyspeptic, respiratory or cardiac symptoms. The respiratory symptoms of dyspnoea, pain in the chest, cough, and cyanosis are due to the reduced lung space and atelectasis due to compression of the basal part of the lung.<sup>10</sup> Depending on the extent and the degree of muscle layer deprivation there is absence or less effective caudal movement of the diaphragm during inspiration. This leads to reduction of lung volumes and impaired ventilation. The dyspeptic symptoms are pain in the abdomen, nausea, vomiting, belching, etc., and are due to the displacement of the abdominal contents. The cardiac symptoms of palpitations, tachycardia and extrasystole are probably caused by the displacement of the mediastinum. In our case eventration is on right side, respiratory symptoms predominated and as it is long standing it lead to cor pulmonale.

The diagnosis of diaphragmatic eventration can usually be made on standard PA and lateral chest films. In the distinct cases they show an unbroken, curved line representing the elevated diaphragm. The unbroken continuity differentiates it from diaphragmatic hernia

Thomas (1970). On the lateral film the difference of level between the normal and the elevated leaf is more pronounced than it appears from the PA film. Eventration of diaphragm may cause pleural effusion appearance in PA and lateral decubitus chest x-ray (Tehrani sign).<sup>3</sup>

Previously, pneumoperitoneography served as the 'gold standard' but has been replaced by fluoroscopy or high resolution ultrasound, Kaplan et al (1994). Ultrasound may present valuable information about diaphragm integrity, with eventration content or the other diaphragmatic pathologies. Although the other imaging modalities such as fluoroscopy, computed tomography, and MR imaging may be performed as adjunct techniques in cases of the diagnosis still in doubt, they are frequently unnecessary after ultrasonography.<sup>11,12</sup> The diaphragm can be seen as a continuous thin layer above the elevated abdominal viscera and on real-time ultrasound the abnormal region can be seen to move downward with the normal portion although it may show a slight lag in its inspiratory excursion. Fluoroscopy may demonstrate smooth elevation of the hemidiaphragm with little or only slightly paradoxical movement of the affected portion of the hemidiaphragm during forced inspiration, although a paralyzed hemidiaphragm is likely to have paradoxical motion.

In our case the diaphragmatic eventration on right side is thought to be idiopathic after ruling out all potential causes like iatrogenic, traumatic, neuromuscular disorders, malignancy. Rare possibility of congenital eventration presenting late in the life is considered as she never had a hospital re-

cord in her life. Associated Cor pulmonale in patient with eventration is a theoretical possibility but is very rare, which is present in this case. Congenital diaphragmatic eventration in asymptomatic individuals does not require any treatment. The routine surgical procedure generally performed in symptomatic individuals is plication of the diaphragm.<sup>13</sup> In view of old age, unwillingness for surgery, presence of pulmonary arterial hypertension, surgery was deferred and patient was managed conservatively. She was now doing well and is under follow up.



Fig-1: CHEST X-RAY PA & Lateral views showing eventration of right diaphragm (yellow arrow)



Fig-2: CT ABDOMEN showing eventration of right diaphragm with liver (red arrow), kidney (white arrow) & colon (yellow arrow) displacement.

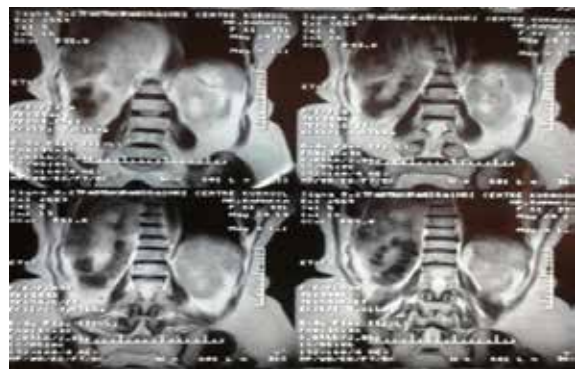


Fig-3: MRI Reconstruction films also showing eventration and displacement of liver, right kidney and colon

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