ORIGINAL RESEARCH PAPER

Neonatology

KEY WORDS: congenital diaphragmatic eventration, congenital diaphragmatic hernia, congenital diaphragmatic defect.

CONGENITAL DIAPHRAGMATIC EVENTRATION- AN INCIDENTAL FINDING

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ABSTRACT

The diaphragmatic eventration is a rare disorder, due to mal-development of muscular portion of hemidiaphragm. The condition is commonly asymptomatic, but can manifest as respiratory distress and recurrent respiratory infections. It can be congenital or acquired. Congenital being more common, acquired variety is due to phrenic nerve injury. It can be differentiated from diaphragmatic hernia due its unbroken continuity. We report a case of a full-term female child born to a primigravida mother with uneventful antenatal and perinatal history. On routine examination child was stable, asymptomatic except presence of heart sounds on right side. In suspicion of dextrocardia, chest X-ray done, suggestive of elevation of left hemidiaphragm and shifting of abdominal viscera in upward direction. 2-D-Echo was normal. CT chest ruled out congenital diaphragmatic hernia and confirmed eventration. Child was managed conservatively and discharged later.

Introduction

Eventration of diaphragm is a rare disorder which occurs due to maldevelopment of muscular portion of diaphragm. It can be congenital or acquired type [1]. Incidence is 1 in 10,000 live births with a male sex and left hemi diaphragmatic preponderance [2]. In eventration, the diaphragm retains its continuity and attachments to the costal margin. Symptomatic cases present with respiratory compromise due to weakness and abnormal elevation of dome of diaphragm causing thoracic displacement. We are reporting a case of full-term neonate with incidental detection of eventration of left dome of diaphragm. Baby was asymptomatic throughout the course of hospitalization and managed conservatively.

Case Report-

A full term female child was born to 25-year-old primigravida mother with uneventful antenatal course Birth weight was 2.9 kg. Baby cried immediately after birth. At birth clinical examination was normal child was active and was feeding well. On routine assessment heart sounds were found on right side hence dextrocardia was suspected, rest systemic examination was normal and liver was on right side. Chest Xray was done which showed elevation of left hemi-diaphragm and upward shifting of abdominal viscera and 2-D -Echo was normal. However diaphragmatic continuity was maintained and its outline could be seen on Chest Xray. Computed tomography was done to rule out other associated diaphragmatic defects. It showed eventration of left hemidiaphragm and no other anomaly was detected. Child was asymptomatic throughout her hospital course and was managed conservatively and discharged with advice for close follow up.

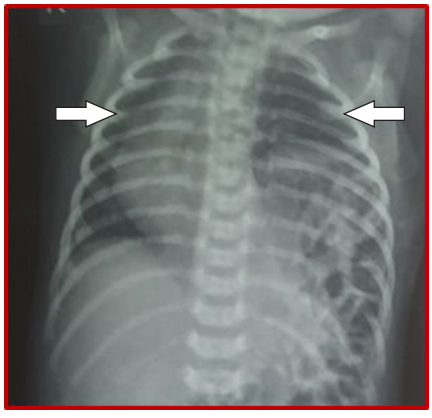

Fig1 -CXR showing elevation of left dome of diaphragm with shifting of heart on right side

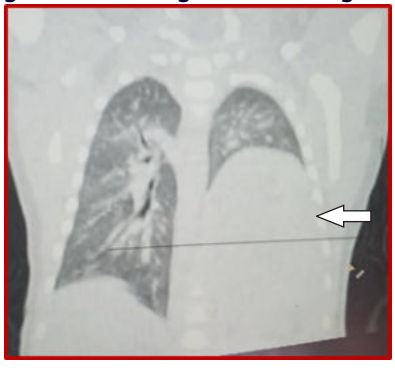

Fig 2- CT chest showing elevation of left dome of diaphragm.

DISCUSSION

Entity of eventration of diaphragm was first recognized by Jean Louis Petit in 1774. Beclard in 1829, coined the word eventration. In 1923 Morrison performed the first successful repair of an eventration of diaphragm [1]. CDE results from absence of the phrenic nerves or inadequate development of the diaphragmatic muscle due to abnormal migration of myeloblasts from upper cervical somites into septum transversum and pleuroperitoneal membrane [2]. Acquired eventration of diaphragm is due to injury to the phrenic nerve as a result of traumatic birth or cardio thoracic surgery [3]. The thin and hypoplastic musculature of diaphragm is unable to restrain the abdominal viscera leading to its cephalad elevation and upward displacement of abdominal organs. The diaphragmatic muscle maintains its normal costal attachments but it has limited motility due to its elevation. Etiology of diaphragmatic is unknown, however association with fetal rubella or cytomegalovirus is found in some cases [4]. Close differential diagnosis of diaphragmatic eventration is congenital diaphragmatic hernia (CDH). In eventration continuity and attachments to the costal margin is maintained, however there is breach in continuity between the diaphragm and the costal margin in CDH [2,3]. Other differential diagnoses are pneumothorax, pleural effusion, congenital pulmonary lesions (Congenital Pulmonary Airway Malformation (CPAM), bronchogenic cyst); Fryns syndrome CDE could be total or focal, bilateral or unilateral. Most common type unilateral and left side [8]. It is usually asymptomatic and may be found as incidental radiographic finding, as in our case [5].

Symptomatic cases may present with respiratory distress which could be life threatening, pulmonary consolidation, repeated respiratory infections. In adults, symptoms like dyspepsia, epigastric discomfort, or burning sensation, abdominal pain, vomiting and flatulence may be seen. [5,6]. Bilateral CDE reduces lung functions even more, especially in supine position [8]. CDE can be associated with other congenital malformations like pulmonary hypoplasia or aplasia, cardiac defects, hydrocephalus, pectus excavatum, cleft palate, hypospadias, cryptorchidism and congenital torticollis [7,8]. In CDE thinned weakened musculature is unable to hold abdominal viscera hence diaphragm elevates and abdominal organs gets displaced in thorax. Depending on sites of herniation organs can be small intestine, stomach, pancreas, kidney, liver [5]. Posterior diaphragmatic eventration is usually difficult to distinguish from Bochdalek hernia [5]. CDE can be diagnosed by chest radiograph in which elevated diaphragm is seen. Computed Tomography scan will show details of herniated abdominal viscera. Fluoroscopy is considered the most reliable diagnostic technique for CDE to document eventration showing reduced, paradoxical or absent movement of diaphragm [11]. Ultrasonography and magnetic resonance imaging can also be used for diagnosis [9, 10].

Treatment of symptomatic CDE is surgical plication of diaphragm. Asymptomatic cases can be strictly followed up regularly for assessment of respiratory function and need of repair [11]. Surgical approach can be either open thoracotomy or minimally invasive video-assisted thoracic surgery [8,10,11]. Surgical repair of CDE increase thoracic volume, increases vital capacity thereby reduces work of breathing [10,11]. In our case patient was conservatively managed as she was asymptomatic and was advised close follow-up.

CONCLUSION-

Asymptomatic CDE is diagnosed usually as an incidental finding. Timely diagnosis and treatment of symptomatic cases of CDE can avoid development of recurrent respiratory infections. Plication of diaphragm is usually the treatment of choice. Close follow up is needed in asymptomatic cases.

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