CASE REPORT

Diaphragmatic defect or pulmonary sequestration in a five-year-old boy

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ABSTRACT
This case report presents a 5-year-old boy with diaphragmatic defect with a tardy clinical manifestation. The findings of chest x-ray and chest CT scan were seen by pediatricians and radiologists as interstitial pneumonia and suspected pulmonary sequestration, pediatric surgeons saw it as a benign teratoma and during surgery a muscular defect of hemidiaphragm right was found. This case emphasize the rarity of diaphragmatic hernia in children of older age, the importance of “accented” clinical suspicion based on the maintenance of signs of mild respiratory failure, the need for additional radiological tests and surgery with the aim to diagnose this abnormality and “successfulness” of prenatal ultrasound examination.

Key words: diaphragm hernias, lung injury, adult children, ultrasonography, prenatal

INTRODUCTION
Development of the diaphragm occurs early in gestation followed by fusion of pleuropertitoneal membrane and transverse septum, creating two holes in the facing the chest and abdominal cavity, that close during the ninth week of gestation. (1). Multifactorial hereditary disorder or harmful agents can prevent the closing of these channels leading to the development of partial defect of the diaphragm or diaphragmatic agenesis (2,3). In the world literature, so far only about 100 reported cases of asymptomatic diaphragmatic hernia have been found in older children and adults (2,3). We strive to contribute with one more case.

CASE REPORT
A five-year-old boy was hospitalized because of dyspnea, central cyanosis and irritating cough. He was afebrile, and respiratory, auscultation sound was diffused and weakened, over the right base inaudible, low pass pitched wheezing diffuse, prolonged expiratory, respiratory frequency (R) of 40/min, percutaneous oxygen saturation (SaO₂) of 88%, heart frequency (F) 150/min. Other physical findings were normal. There was leukocytosis (le) 17.8x10⁹/L (predominantly granulocytes, 90%), and elevated lactic dehydrogenase 618IU/L. Other biochemical findings were within normal ranges: C-reactive protein (CRP), urea, creatinine, aspartate transaminase or glutamic oxaloacetic transaminase (AST), alanine transaminase or glutamic pyruvic transaminase (ALT)). Antibodies to Mycoplasma pneumoniae and Echinococcus were negative. Chest x-ray showed a triangular, inhomogeneous, circumscribed opacity lung parenchyma with a pair of small oval transparency right paracardially that clears the right cardiac contour shadow (Figure 1). Similarly, it was seen in the right profile in cardiac shadow projection anteriorly and cranially with curved linear shadow. Contoured hemidiaphragm was clearly and normally positioned. The boy was healthy until this hospitalization.

Figure 1. Chest X-ray: both sides bronchopneumonia, dominant at right (Clinical Center Kragujevac, Serbia, 2010)

Treatment was initiated according to the protocol of atypical bronchopneumonia (5 days oxygen therapy, bronchodilators and simultaneous distribution of azithromycin and systemic glucocorticoids for the first 3 days of hospitalization). On
the fifth day of the treatment the boy presented mild hypoxia (SaO2 94%), mild tachypnea (R32/min), tachycardia (F135/min), auscultation respiratory sound was discreetly altered, basal right and spirometric finding showed mild restriction. The boy was afebrile all the time. These findings were an indication to do a computerized tomography (CT) of the chest.

Computerized tomography image of the boy’s chest revealed, in the front-right lower mediastinum, homogeneous and clearly restricted zone of the dimension 78x54x44mm, not separated from the shadows of the mediastinum but clearly limited by the lung parenchyma, which supports extralobar pulmonary sequestration (Figure 2). Sequestration within more consolidated several distinct zones and aerated zones indicated communication with the bronchus. On both sides, parahilar and back-basal, the peribronchial and perivascular infiltrates in terms of interstitial pneumonia were seen. There were no mediastinal lymph nodes and the thymus was normal.

After consulting thoracic surgeon the child was sent on the fifth day of the hospitalization for a surgical treatment with diagnosis of suspected pulmonary sequestration. Slow passage of column curves and water absorption on the X-ray and CT scan of the chest were mimicking, by differential diagnosis, pulmonary sequestration, benign teratoma, right Bochdalek and Morgagni’s hernia. During performed anterolateral thoracotomy at the right side, a defect of the muscular diaphragma (next to and medially) was found, which was presented as partial agenesis of the diaphragm. The patient recovered without complications.

Clinical and radiological presentation of the case seemed as interstitial pneumonia and pulmonary sequestration to pediatricians and radiologist, while for pediatric surgeons it looked like benign teratoma. A muscular defect of the right hemidiaphragm was confirmed during the surgery.

The incidence of diaphragmatic defect in newborns is 1:4000-5000, and it could result in respiratory failure and high risk of mortality without successful surgical intervention (1,2). Rarely, the massive congenital diaphragmatic hernia has a favorable prognosis as in case of appropriate and timely diagnosis and surgical treatment (4), as fortunately happened to our patient. Diaphragmatic defect is five times more common on the left than right side of the thorax and it usually appears in patients with positive family history (1,2). Right-sided diaphragmatic defect, which was found in our patient, is accompanied by lack of symptoms because the liver prevents herniation of other organs through the diaphragmatic defect over time (3). Symptoms depend on a degree of herniation: small hernia goes undetected and can be incidentally detected much later in life (4). Partial agenesis of the diaphragm is often associated with cardiovascular, and rarely, with urogenital, gastrointestinal and chromosomal abnormalities (6). Malformations of these organ systems are not found in our patient.

The diagnosis of congenital diaphragmatic hernia is usually set in advanced stages of pregnancy when some degree of pulmonary hypoplasia, mediastinal shift, polyhydramnios and pseudocyst in the thorax originated from intestinal gyrus can be assessed (5,6). Postnatally, the diagnosis could be confirmed by X-ray or CT or magnetic resonance imaging of the chest (5,6). X-ray and heart auscultation findings of our patient were neither similar nor in line with the findings of other reports (5,6). The heartbeats could not be heard on the opposite side of the chest.

In conclusion, this case indicates the insufficiency of prenatal ultrasound examination although the mother’s pregnancy was controlled by the adopted protocol. This indicates practical and scientific problems. Late surgical correction in children of older ages is always accompanied by a high risk of complications of diaphragmatic hernia, which may be life threatening.