

DELAYED PRESENTATION OF CONGENITAL DIAPHRAGMATIC HERNIA WITH SPLENIC AND CAECAL HERNIATION

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Received: August 22, 2011
Accepted: October 22, 2011

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The vast majority of posterolateral congenital diaphragmatic hernia (CDH) diagnosed on prenatal ultrasound screening after 24 weeks of gestation present with respiratory distress at birth. It has been observed that late presenting CDH has better outcome and is not associated with pulmonary hypoplasia. We report a case of posterolateral CDH with herniated spleen presented first in a four year old male child with chest infection. This case report highlights the fact that posterolateral CDH which presents late is not associated with pulmonary hypoplasia, but may cause respiratory symptoms.

Key words: Delayed presentation ▪ Congenital diaphragmatic hernia

Introduction

Congenital diaphragmatic hernia (CDH) was first described by Riverius (1). CDH occurs in about 1 in 3000 births. The most common defect is the posterolateral (Bochdalek) type. Over 90% of the patients will be diagnosed either in the antenatal period or will present with respiratory distress in the first few hours of life. There is significant mortality associated with this group (2).

However, about 5% to 30% of diaphragmatic hernias present beyond the neonatal period (3). Although the mortality in this group is low, the morbidity may be significant. The late presenting CDH poses considerable diagnostic challenges because of its varied presentation, often resulting in diagnostic delay, inappropriate treatment, and potential fatal outcome.

We report a case of posterolateral CDH with herniated spleen presented first in a four year old male child with chest infection.

Case report

A four year old male child, born to a non-consanguineous marriage, was moved from a private nursing home complaining of fever, cough and dyspnoea for one week. His postnatal history was uneventful and his past history was suggestive of recurrent chest infections.

On examination, the patient was conscious, febrile and had respiratory distress with respiratory rate of 34 per minute with 100% oxygen saturation, without any sign of congestive cardiac failure. His chest was symmetrical with decreased chest movements on the left side, with minimal shift of the mediastinum to the opposite side. On auscultation air entry was decreased on the left side with peristaltic sounds heard in the left hemithorax. The abdomen was not scaphoid and there was no organomegaly. Other systems were normal. Chest roentgenogram showed air filled bowel loops in the chest



Figure 1 Plain chest radiograph showing air filled bowel loops in the chest cavity



Figure 2 Barium meal follow through study showing loops of intestine in the left hemithorax

cavity, with minimal shift of the mediastinum to the right. The left hemidiaphragm was not clearly visualized (Fig. 1). Barium meal follow through was done, which revealed bowel loops in the left hemithorax, suggestive of left-sided diaphragmatic hernia (Fig. 2).

The patient underwent surgery for this. On exploration, a 4x3 cm muscular defect was found in the posterolateral region of the left dome of the diaphragm (suggestive of Bochdalek hernia); the jejunum, ileum, caecum with appendix and spleen were found herniating through the defect. The organs were repositioned and primary repair was done by approximating the margins of the defect. The postoperative period was uneventful and the patient was discharged 18 days after surgery.

Discussion

Presentation of Bochdalek type CDH may vary. The vast majority of cases are picked up on prenatal ultrasound screening; however, due to the dynamic component, the diagnosis can be missed (4). Most cases of late presenting CDH occur in the first few years of life (5, 6), but cases are reported with some frequency well into adolescence. It is not known whether the herniation of the viscera is continuously present for such a long time, or only happens with the presentation of symptoms. It is easy to hypothesize that a cause, like a simple chest infection with coughing, may provoke herniation due to increased abdominal pressure.

Chronic symptoms, such as recurrent chest infections may result from inadequate ventilation caused by longstanding compression of the ipsilateral lung by the herniated viscera. In our case, the history of recurrent chest infections may suggest long term herniation. However acute deterioration of the respiratory symptoms may indicate propagation of more viscera into the chest.

Late presenting CDHs may manifest with gastrointestinal syndromes, gastric volvulus,

gastric perforation, bowel strangulation necrosis, splenic torsion, predominantly due to a narrow diaphragmatic defect (7, 8, 9). In our patient, the size of the defect on the diaphragm cannot be considered large, however no gastrointestinal syndromes were present. The Barium meal did not indicate dilated bowel loops or obstruction at the level of the diaphragm.

The outcome of late-presenting CDH is usually favourable, which is related to the absence of accompanying pulmonary hypoplasia and low incidence of other congenital malformations (8). In our case no lung hypoplasia was noted at the reconstruction site and no postoperative respiratory complications have been experienced as expected in the vast majority of perinatally detected CDH cases.

On the basis of the nitrofen induced rat model, it is nowadays believed that lung hypoplasia associated with CDH does not only originate from mechanical compression of the herniated viscera (10). The lack of pulmonary hypoplasia and the better outcome in the late presenting group of CDH pati-

ent suggest that this subpopulation may have other origins of the ailment.

In conclusion, delayed presentation of posterolateral CDH is uncommon. Late presentation is not associated with severe pulmonary hypoplasia, but may result in respiratory and gastrointestinal complications. The prognosis is favourable with correct diagnosis and surgical repair.

Acknowledgement

The authors are thankful to Dr. Kuldeep Shah for technical support in formatting the article.

Authors' contributions: Conception and design: VYK, MA; Acquisition, analysis and interpretation of data: MA, CA, SN; Drafting the Manuscript: VYK, MA CA SN ; Revising it critically for important intellectual content: VYK, MA.

Conflict of interest: The authors declare that they have no conflict of interest. This article was not sponsored by any external organisation.

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Citation: Kshirsagar VY, Amithkumar Ch, Nagarsenkar S, Ahmed M. Delayed presentation of congenital diaphragmatic hernia with splenic and caecal herniation. *Paediatrics Today.* 2012;8(1):75-77.