

LATE PRESENTING CONGENITAL DIAPHRAGMATIC HERNIA IN A 4-YEAR-OLD CHILD

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Abstract

The majority of congenital diaphragmatic hernia (CDH) are diagnosed antenatally, immediately after birth or in the neonatal period. We report case of 4-year-old child who presented with recurrent wheezing and was diagnosed to have late presenting CDH.

A 4-year-old girl was admitted to the hospital with a 5-days history of increasing shortness of breath and cough. The child had normal birth history and has enjoyed relatively good health till 3 years of age. Her medical history over the last year revealed that she had 4 episodes of wheezing variously treated with inhaled bronchodilators, steroids and antibiotics.

On physical examination she was in mild respiratory distress with bilateral wheezing.

The x-ray showed an unusual appearance of the chest with air-filled bowel loops. A barium X-ray examination of the gastrointestinal tract confirmed the diagnosis of CDH. The hernial contents were reduced back into the abdominal cavity and surgical repair of the diaphragmatic defect was done.

Late presenting CDH can be misdiagnosed and lead to delayed treatment. It should be suspected in children presenting with recurrent respiratory symptoms.

When the diagnosis is confirmed, the treatment of choice is surgical intervention. Clinical awareness, early diagnosis and operative treatment can lead to good outcomes.

Key words: Congenital diaphragmatic hernia, child, wheezing.

Introduction

Congenital diaphragmatic hernia (CDH) is one of the most common major congenital malformations in newborns. Most cases are detected prenatally or present with clinical symptoms in the newborn period. Survival depends on timely diagnosis and treatment.

On the other hand there is a subgroup of patients with CDH who appear outside the first 4 weeks of a child's life.

Case report

A 4-year-old girl was brought to the hospital in order to get a medical evaluation because of 5-days history of increasing shortness of breath and cough.

A review of previous medical history revealed that the child had normal birth history and was in very good clinical condition and did not complain of cough or any other respiratory clinical manifestation till 3 years of age. She had recurrent episodes of wheezing variously treated with inhaled bronchodilators, steroids and antibiotics over the last year.

On admission during physical examination she was in mild respiratory distress with cough, prolonged expiratory phase and bilateral wheezes.

A complete blood count, immunoglobulins, sweat test, tuberculin skin test and skin prick test were found to be normal.

Anteroposterior (AP) view of the chest showed several bowel loops in the projection of the heart. The lateral x-ray also demonstrated this finding behind the sternum (Figure 1a & 1b).

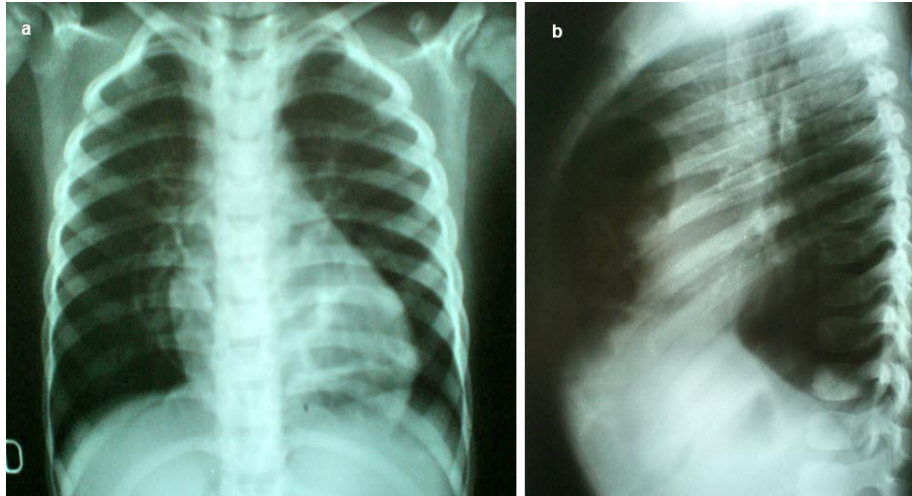


Figure 1a. Chest radiograph (AP) shows herniated gut loops in thorax. **Figure 1b.** Chest radiograph (lateral view) shows herniated gut loops in thorax retrosternally.

A barium x-ray examination demonstrated contrast within herniated part of small intestine and part of the transverse colon in thorax (Figure 2a & 2b).

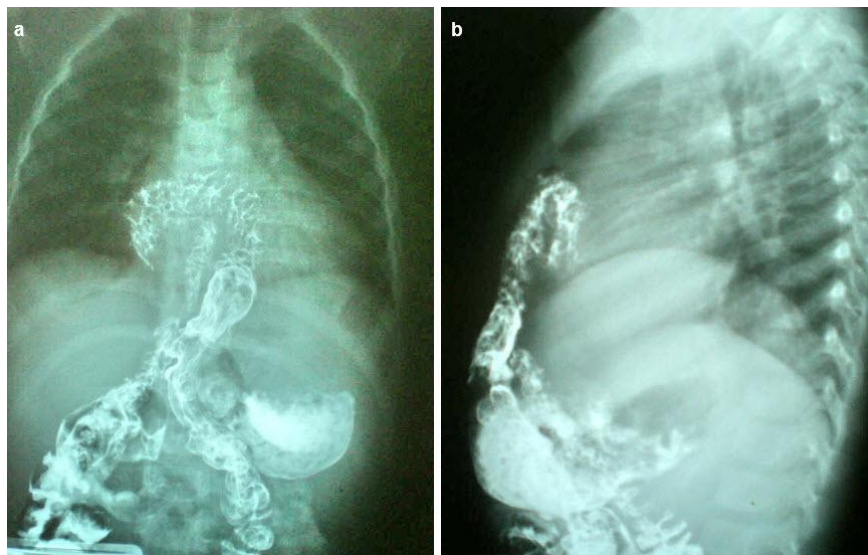


Figure 2a Barium swallow study in the chest cavity. **Figure 2b** shows loops of intestine

The child was referred to the Clinic of pediatric surgery for further diagnostic investigations and appropriate therapeutic intervention.

Congenital Morgagni hernia was confirmed at surgery. Herniated contents were replaced in the abdominal cavity and primary repair of diaphragm was done (Figure 3a & 3b).

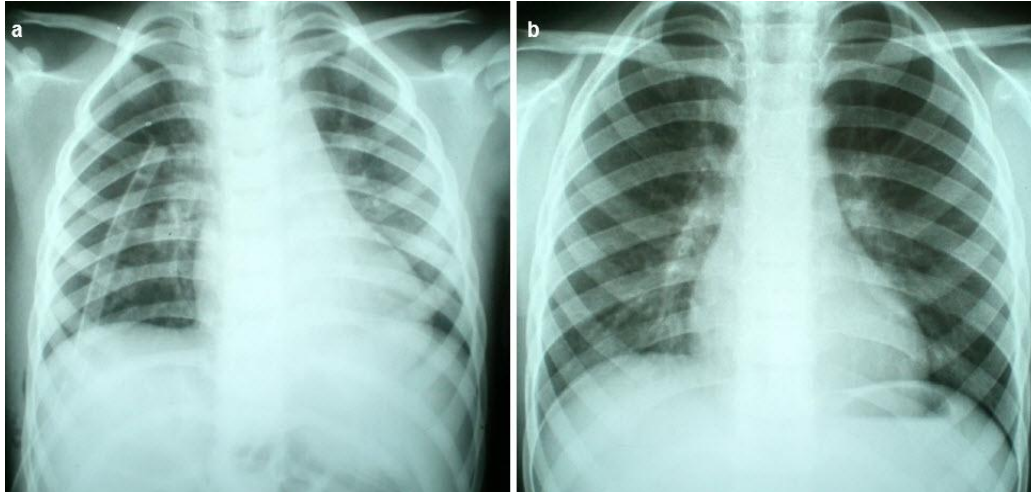


Figure 3a & 3b. Chest x-ray after primary repair of diaphragmal hernia.

Discussion

Congenital diaphragmatic hernia (CDH) is a defect of the diaphragm characterized by penetration of abdominal organs into chest cavity. Most cases are detected prenatally or present with clinical symptoms in the newborn period.

Clinical features depend on location and size of the diaphragm defect and the presence of abdominal organs in thorax. If it is a major defect, the symptoms appear immediately after birth accompanied by severe respiratory distress and cardiac involvement. In some patients, symptoms related to CDH at birth are minimal and the diagnosis is made after the neonatal period due to examination of other disturbances [1,2].

The incidence of late presenting CDH is up to 25% of all cases of this malformation [3,4]. Children with late presenting CDH have less morbidity compared to patients whom the diagnosis was made antenatally or in the neonatal period [5].

The symptoms of delayed presentation of CDH are most often respiratory difficulties, gastrointestinal disorders or combined [6].

In children with unexplained acute or recurrent respiratory symptoms, the diagnosis is usually established by chest X-ray when gas-filled intestinal loops are seen in the chest cavity [7].

An abdominal radiograph may be obtained in patients presenting with gastrointestinal symptoms. A computerized tomography scan, magnetic resonance imaging findings or barium swallow examination can be used to differentiate CDH from other chest masses.

Patients with a delayed clinical occurrence of CDH, unlike children with early-diagnosed CDH in the neonatal period have better outcome due to less severe or absent lung hypoplasia [8].

Late-onset CDH is generally considered as curable condition with a good recovery [9,10].

It must be treated surgically immediately to prevent complications.

Conclusion

Early-presenting CDH usually presents acute symptoms and has a higher mortality rate. In contrast, late presenting CDH has a less acute course and therefore may be undiagnosed at birth or in the neonatal period.

Late presentations of CDH should be considered in young children with recurrent wheezing. Once the diagnosis of CDH is established, treatment of choice is operative with great benefit to the patient.

References

1. Anaya-Ayala JE, Naik-Mathuria B, Olutoye O. Delayed presentation of congenital diaphragmatic hernia manifesting as combined-type acute gastric volvulus: a case report and review of the literature. *J Pediatr Surg* 2008; 43(3): 35-9.
2. Radovic S. Late presentation of congenital diaphragmatic hernia-Case report. *Srp Arh Celok Lek* 2015;143(9-10): 604-8.
3. Berman L, Stringer D, Ein S, Shandling B. The late presenting pediatric Bochdalek hernia: a 20-year review. *J Pediatr Surg* 1988; 23:735-8.
4. Weber T, Tarcy T, Bailey P, Lewis E. Congenital diaphragmatic hernia beyond infancy. *Am J Surg* 1991;162: 643-6.
5. Chang SW, Lee HC, Yeung CY, Chan WT, Hsu CH, Kao HA, Hung HY, Chang JH, JC Sheu, Wang NL. A twenty-year review of early and late-presenting congenital Bochdalek diaphragmatic hernia: are they different clinical spectra? *Pediatr Neonatol* 2010;51(1): 26-30.
6. Bağlaj M. Late-presenting congenital diaphragmatic hernia in children: a clinical spectrum. *Pediatr Surg Int* 2004;20: 658-69.
7. Kumbhar S, Siddiqui S, Thakrar P. Avoiding misdiagnosis in postnatal presentation of congenital diaphragmatic hernia: A report of two cases and review of radiologic features. *Radiol Case Rep* 2019;14(10): 1288-92.
8. Yuan M, Li F, Xu C, Fan X, Yang G, Xiang B, Huang L. Emergency management of acute late-presenting congenital diaphragmatic hernia in infants and children. *Pediatr Emerg Care* 2021;37(7): 357-9.
9. Elhalaby E, Abo Sikeena M. Delayed presentation of congenital diaphragmatic hernia. *Pediatr Surg Int* 2002;18: 480-5.
10. Mei-Zahav M, Solomon M, Trachsel D, Langer J. Bochdalek diaphragmatic hernia: not only a neonatal disease. *Arch Dis Child* 2003;88: 532-5.