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Late-presenting right congenital diaphragmatic hernia with severe hypotrophy of the right lobe of the liver

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ABSTRACT

INTRODUCTION: Congenital diaphragmatic hernia (CDH) presenting after 30 days of life is unusual and has a variant pattern of presentation.
PRESENTATION OF CASE: We present a death case occurred to a 34-days-old infant. The infant arrived to our emergency department in cardiac arrest after having suffered from intermittent acute abdominal pain. Autopsy confirmed the presence of a right CDH, with herniation of the right lobe of the liver into the thorax.
DISCUSSION: Most of the cases of CDH are diagnosed prenatally or in the neonatal period. However, some patients do not develop symptoms until after the neonatal period. The relevance of our case is the co-existence of right CDH and important hypotrophy of the right lobe of the liver.
CONCLUSIONS: Evidence of this phenomenon represents an absolute novelty in the extant scientific literature. Even if rare, we suggest to suspect the presence of CDH in fetus with disparity in right and left liver lobe at prenatal ultrasound.

1. Introduction

Congenital diaphragmatic hernia (CDH) is a relatively common malformation approximately occurring in 1 out of 2500 newborns [1]. CDH usually raises respiratory distress in the neonatal period and has a high mortality rate. Some patients do not show symptoms until later stages of life, and asymptomatic period usually ends with an acute attack of disease. With a correct diagnosis of the disease, the prognosis can be excellent. Nevertheless, clinical conditions of the patient are sometimes so serious that they rapidly worsen insomuch as making surgical intervention not feasible.

2. Presentation of case

A 34-days-old Caucasian male arrived to our emergency department in cardiac arrest after referred intermittent acute abdominal colic. Gestational history was unremarkable; he was born at 37 weeks of gestational age, after spontaneous vaginal delivery (neonatal weight 2650 g). The postnatal period was uneventful. At the admission to our Pediatric Emergency Unit, cardiopulmonary resuscitation was performed for 20 min before spontaneous circulation restarted. Medical staff then practiced intubation and therapeutic hypothermia. The chest X-ray (Fig. 1) showed a well-defined circled aerial content in the right hemi thorax, which was presumed to be an intestinal ansa. On computed tomography (Fig. 2), part of the right-lobe of the liver was seen in the right hemi thorax with intestinal ansa herniated in a posterolateral defect. The liver presented modified anatomy with hypoplasia

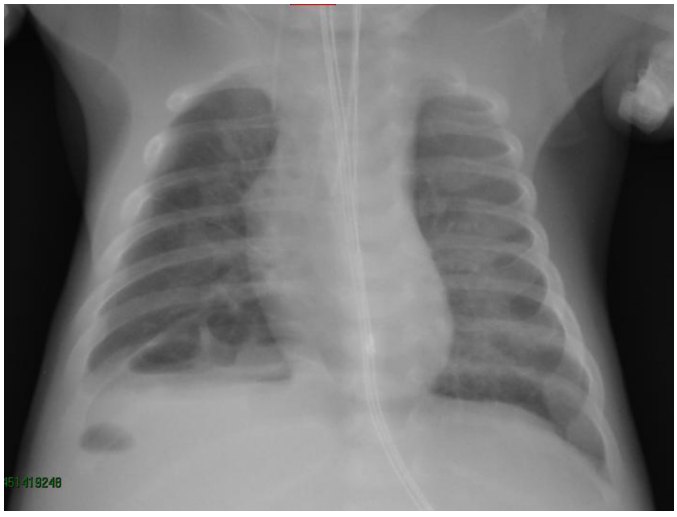

Fig. 1. Chest X ray showing a circled aerial content in the right hemi thorax (black and white print).

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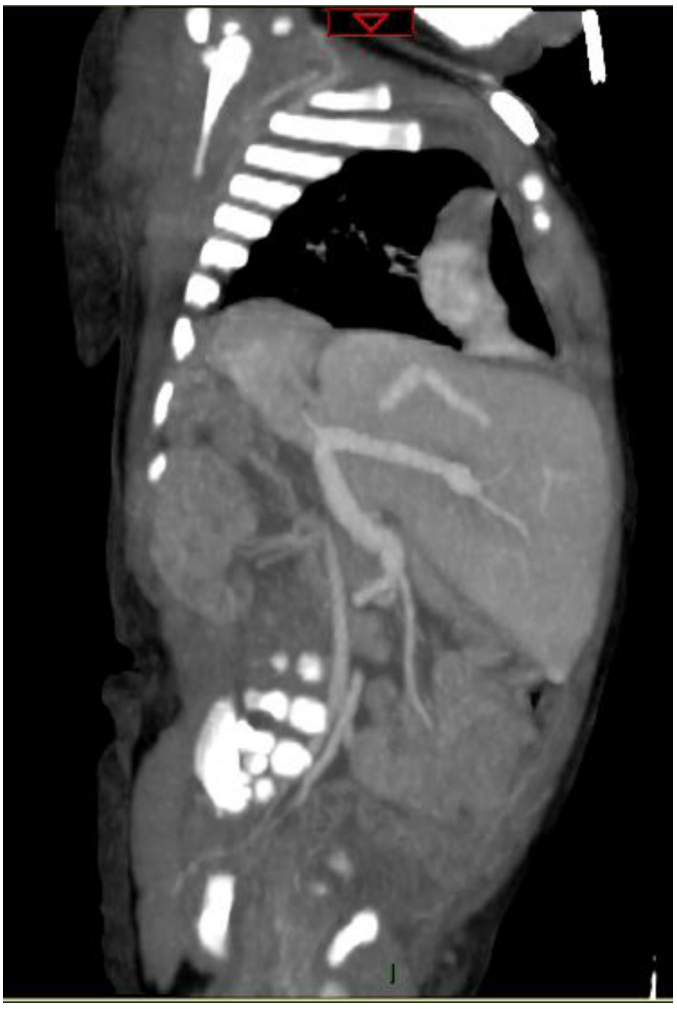

Fig. 2. CT scan demonstrating liver with modified anatomy: right lobe hypoplasia and compensatory hypertrophy of the left lobe (black and white print).

of the right lobe and compensatory hypertrophy of the left lobe. Right portal branch presented threadlike. No other malformations were detected. Aggressive therapeutic hypothermia, therapy with inotropes vasopressors and bicarbonate were nonproductive and the patient was subsequently pronounced dead after six days of coma. The autopsy (Fig. 3) found a 4 × 3 cm right posterior hernia, with part of the right lobe of the liver in the thorax. The diaphragmatic surface of the right liver presented signs of parenchymal atrophy and fibrosis.

3. Discussion

Congenital diaphragmatic hernia (CDH) approximately occurs in 1 out of 2500 newborn infants [1]. Most of the cases are diagnosed prenatally or in the neonatal period. However, some patients do not develop symptoms until after the neonatal period. Late-presenting CDH cases are about 5–45.5% of all cases of CHD [2,3]. Late-presenting CDH is characterized by a variety of clinical manifestations. When the late-presenting CDH is left-sided, it typically produces acute, obstructive, gastrointestinal symptoms, while the right-sided CDH is usually associated with respiratory issues [4]. Late presenting CDH in characterized by a wide clinical spectrum; partial liver displacement, that occurs in most right-sided CDH children, may block the further herniation of hollow viscera. That is the reason why in the right CDH group most children have respiratory symptoms only. Associated anomalies in late-presenting CDH patients have been reported with variable frequencies, ranging from 8.6 to 80% [3–6]. The most commonly reported radiological finding of right-sided CDH is the opaqueness of the right hemi thorax usually associated with mediastinal shift to the contralateral side. Computed tomography can be considered the ideal non-invasive technique for diagnosis, offering the unique opportunity to evaluate the presence of diaphragmatic defect, size, exact location and contents of the various types of diaphragmatic hernia [7]. Once diagnosed, surgical intervention is necessary for the prevention of complications; late-presenting CDH is usually considered as a benign condition with a good prognosis. Nevertheless, because of its wide spectrum of clinical manifestation, a high index of suspicion is important for diagnosis because it can rapidly become a life-threatening condition [4,8].

4. Conclusions

Late-presenting CDH have a variety of clinical manifestations that can be either acute or chronic, and can include respiratory and gastrointestinal symptoms, or both. Acute presentation may be due to rapid visceral displacement into the chest or, in most of the cases, to rapid distension of previously herniated hollow viscera. Rapid displacement of intra-abdominal organs may lead to sudden cardiorespiratory arrest. We suppose that sudden displacement of right lobe of the liver and bowel in the thorax caused severe kinking of hepatic veins with decrease of the blood flow and venous return to heart. Hypotrophy of the right lobe of the liver in right-sided CDH is uncommon. Such hypotrophy is not on a developmental basis but

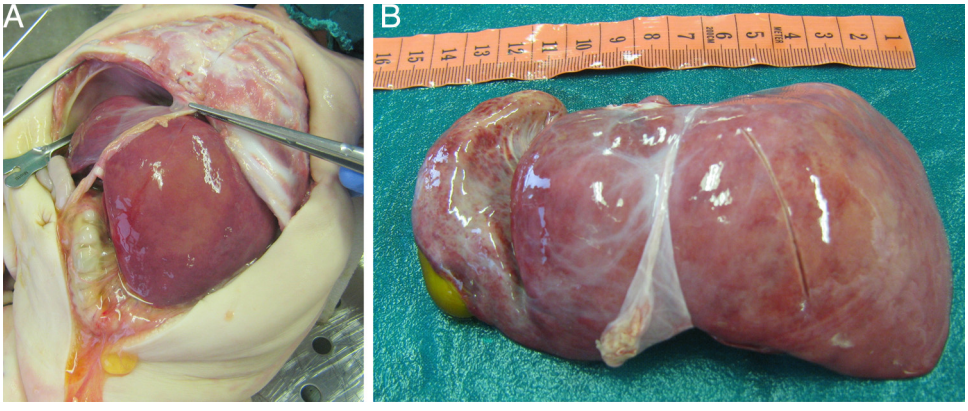

Fig. 3. Autopsy photographs showing the posterolateral diaphragmatic defect (a) and the important hypotrophy of the right lobe of the liver (b) with the diaphragmatic surface presenting signs of parenchimal atrophy and fibrosis (color print).

occurs secondary to nutritional, vascular or toxic influences and always produces a compensatory hypertrophy of the left lobe of the liver.

Even if rare, we suggest to suspect the presence of CDH in fetus with disparity in right and left liver lobe at prenatal ultrasound.

Conflicts of interest

None.

Funding

None.

Ethical approval

Present report complies with the guidelines for human studies and animal welfare regulations. Ethics Committee of our Hospital has not been consulted because no human and animal experiments were conducted.

Author contribution

E.A. De Marco: data collection and paper writing. L. Merli: data collection, paper writing and interpretation. A.Taddei: data collection and paper revision. S.M. Pulitanò: data interpretation. C. Manzoni: data collection. L. Nanni: revision.

Consent

Although no identifying details are reported, parents of the patient have given their informed consent.

Guarantor

Lorenzo Nanni.

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