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Case Report

Right Congenital Diaphragmatic Hernia: Report of a Clinical Case in a 10-Day-Old Neonate in the Neonatology Unit of the Mali Hospital

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Abstract: ***Introduction:*** Congenital diaphragmatic hernia (HDV) is a rare malformation. The posterolateral or Bochdalek form is the most frequent (80%) and is located on the right in only 15% of cases. It is a rare cause of respiratory distress in newborns. Our objective was to describe the diagnostic and therapeutic aspect of a case in a 10-day-old newborn in the neonatology unit of the Mali hospital. ***Observation***: He was a 10-day-old male infant admitted for dyspnea from birth. He was born from a pregnancy followed to term without major incidents. Syphilis serology was negative. However, toxoplasmosis, rubella, hepatitis B and HIV serologies have not been performed. Four ultrasounds performed during pregnancy returned to normal. He was born by scheduled Cesarean section uterus and resuscitated at birth. The emergency chest X-ray was not informative. The diagnosis of right congenital diaphragmatic hernia was confirmed by the thoracoabdominal CT scan. A treatment combining oxygen therapy, 10% glucose serum: 100 ml / kg / d in infusion and domperidone: 1 ml / kg / d in 3 administrations per os has been established. After one month and 23 days of hospitalization, after stabilization of respiratory distress, he was operated. The aftermath was simple. ***Conclusion:*** Congenital diaphragmatic hernia is a rare condition. The posterolateral form is more common. Respiratory distress is their main mode of revelation at birth. In the event of any neonatal respiratory distress, the hypothesis of HDC must be evoked and confirmed by an X-ray of the chest.

Keywords: Hernia, diaphragmatic, congenital, hospital in Mali.

INTRODUCTION

Congenital diaphragmatic hernia (HDC) is defined by the presence of an opening in the diaphragm [1].

The etiology of HDC is poorly understood. Abnormal embryogenesis due to defective retinoid signaling could be involved as well as ZFPM2 mutations in some cases [2].

It represents 8% of congenital malformations. There are several types: hiatus hernia, retro-costo-xiphoid hernia and posterolateral hernia or Bochdalek's hernia the most common. Its incidence is estimated at 1/3200 births [3].

In the vast majority of cases (80%) the cupola hernia is on the left [4]. It is accompanied by constant prenatal pulmonary compression responsible for a very high mortality rate of between 30 and 50% of deaths [5].

While the progress made in neonatal resuscitation has markedly improved its prognosis in developed countries, this is not the case in developing countries where the prognosis remains poor [6].

The aim of our work is to report a case of right diaphragmatic hernia with ascension of the colonic angle and of the liver in the right hemithorax in a 10-day-old newborn, diagnosed and operated on in the neonatology unit of the Mali hospital.

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CLINICAL OBSERVATION

It was a 10-day-old male infant admitted with respiratory distress. Her mother with a history of pregnancy-induced hypertension and cesarean section had 6 pregnancies. The 3rd and 4th pregnancy were aborted in the 3rd and 4th month, respectively, spontaneously. There was no notion of inbreeding in their marriage. Our newborn was the 4th of a family of 4 children. Her 3 older sisters were doing well.

He was the result of a pregnancy followed to term without major incidents. The syphilis serology was negative. However, toxoplasmosis, rubella, hepatitis B and HIV serologies were not performed. Four ultrasounds performed during the pregnancy returned to normal. He was born by scheduled cesarean section for a scarred uterus and then resuscitated at birth. The birth weight was 3100 g, the birth size was 50 cm, and the head circumference at birth was 34 cm.

The onset of the disease dated back to birth marked by respiratory distress. In view of the persistence of the respiratory discomfort, a chest x-ray was urgently requested. She showed multiple images of bullous clarities of the right hemithorax delimited by fine septa mistakenly evoking a giant lobar emphysema hence its reference in our service.

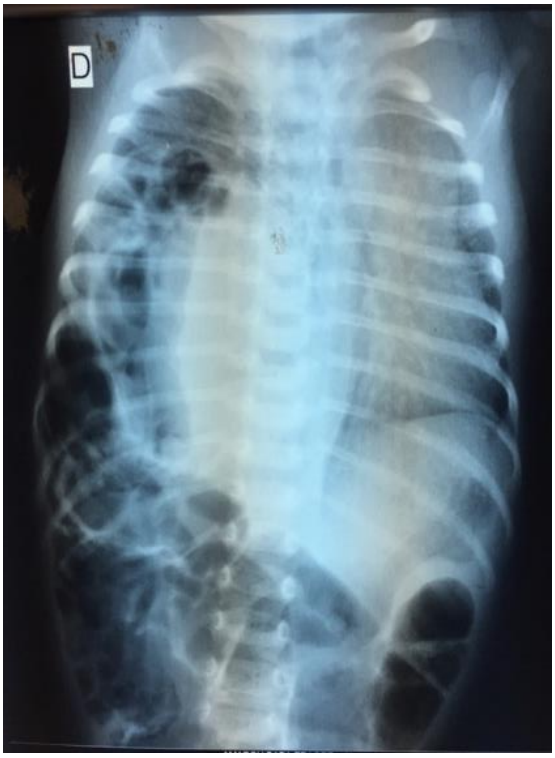

Figure 1: Chest x-ray

On admission the weight was 2980g. The size was 50cm. The head circumference was 34cm. The temperature was 37.1° C. he was in good general condition, with good cutaneous-mucous coloration. The respiratory rate was 65 cycles per minute. Arterial oxygen saturation was 89% in air. There was intercostal indrawing and thoraco-abdominal rocking. On pulmonary auscultation the vesicular murmur was diminished on the right. Cardiac auscultation was normal. The abdomen was hollow with normal transit. The testicles were swollen, painless, translucent. The remainder of the somatic exam was unremarkable.

A chest x-ray performed showed hydro-aeric images in the right hemithorax (Figure 1), the thoraco-abdominal CT scan revealed a right diaphragmatic hernia with presence of the liver, right colonic angle in intrathoracic, right pulmonary collapse and displacement of the mediastinum to the left side (Figures 2, 3).

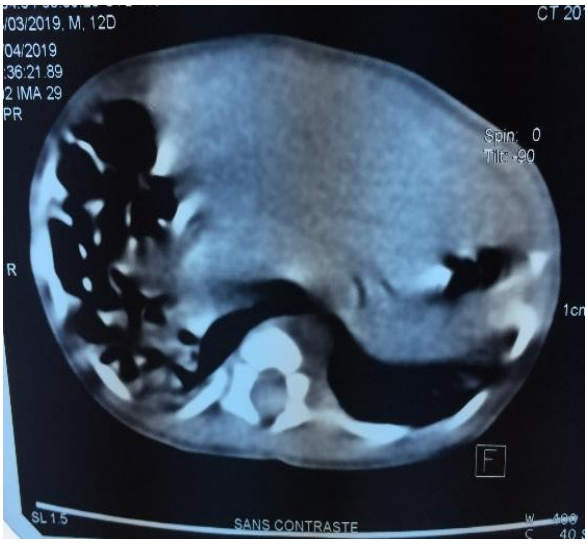

Figure 2: Thoracic CT mediastinal window

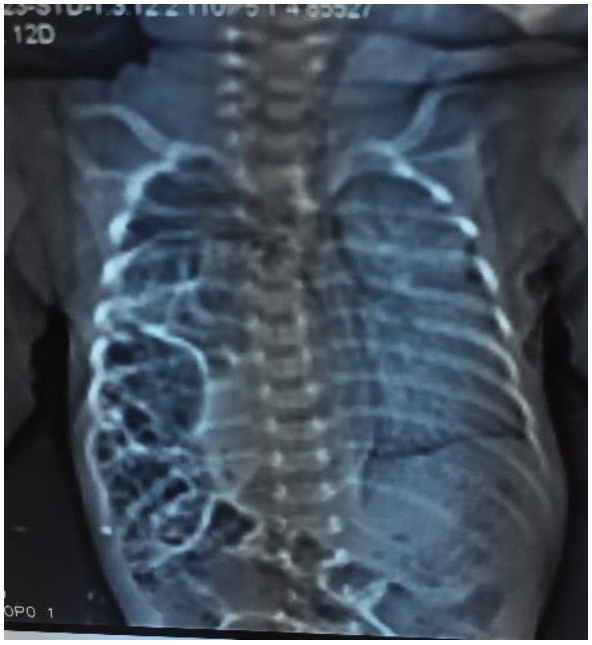

Figure 3: Thoracic CT parenchymal window

The inguino-scrotal ultrasound showed a communicating hydrocele of medium abundance on the

right with septa and low abundance on the left. The requested cardiac ultrasound for an associated heart defect returned to normal.

A treatment combining oxygen therapy, 10% glucose serum: 100 ml / kg / day as an infusion and domperidone: 1 ml / kg / day in 3 oral administrations was initiated. The blood count and C reactive protein were normal.

On D 53 of hospitalization after a multidisciplinary meeting comprising a pediatric surgeon, a thoracic surgeon, a pediatrician and an anesthesiologist, he was operated: under general anesthesia. The surgical technique was as follows: a right subcostal laparotomy was performed of approximately 8cm. On exploration, we found a right posterolateral hernia of approximately 6cm in diameter, the left lobe of the liver and the right colonic angle contained in a bag, were in the right hemithorax compressing the ipsilateral lung (Figure 4 , 5). We repressed the herniated organs. We proceeded to close the hernia with X-stitch separated at 2 suture reinforced by a 2/0 vicryl overlock. A chest tube was placed.

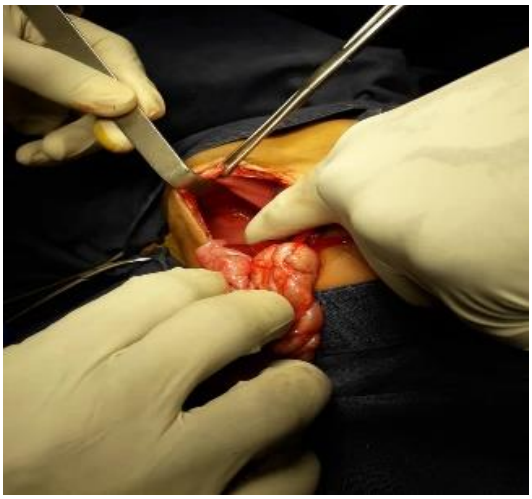

Figure 4: Intraoperative image showing the defect and herniated viscera

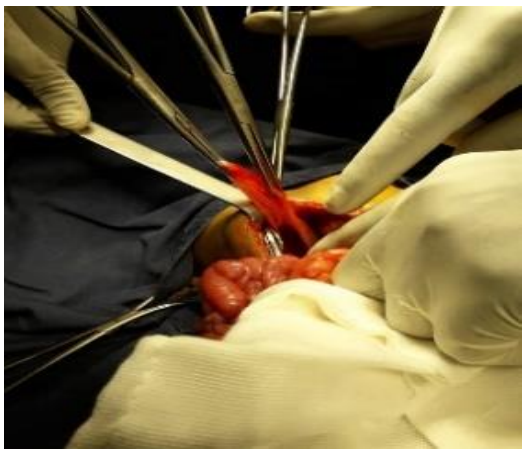

Figure 5: Intraoperative image showing the hernial sac

The immediate postoperative follow-up was straightforward. He was released on the 6th postoperative day with a weight of 3400 g Figures 6 & 7.

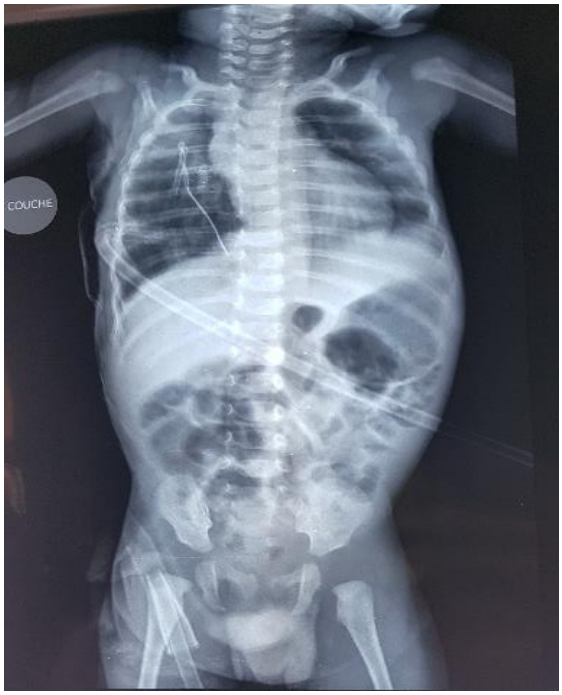

Figure 6: Chest x-ray on day 1 of hospitalization

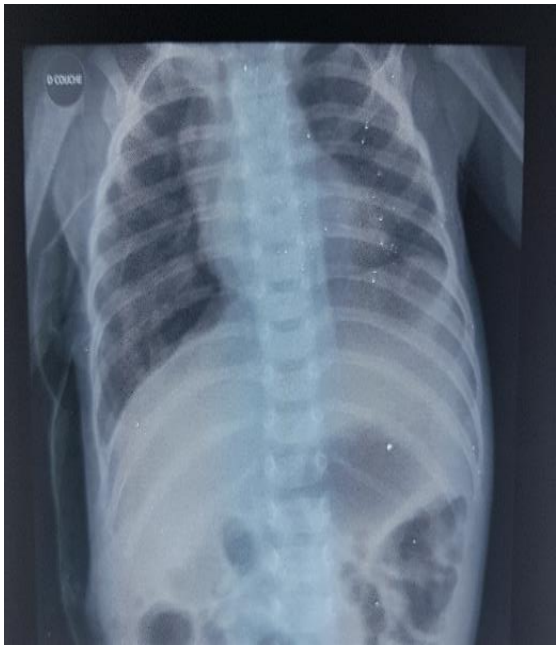

Figure 7: Chest x-ray on day 7 of hospitalization

COMMENTARY

Congenital diaphragmatic hernias are a rare and potentially serious disease entity [7]. Its incidence is estimated at 1 per 2000-5000 births [4, 8]. We have recorded two cases in the service in 8 years. The male sex is predominant [1, 9, 10].

Its etiology is not well known. The diaphragm is formed between the 4th and the 8th week of amenorrhea (SA) separating the pleural and peritoneal cavities by the closure of the foramen of Bochdalek. From 8th to 10th SA the intestinal loop is reintegrated by rotating. The result is a primary failure to close the pleuroperitoneal duct [11].

We can distinguish: the left or right posterolateral form; the anterior retro-xiphoid form of Morgani-Larrey and the hiatus form [12].

About a third of HDC cases are associated with heart defects and lesser proportions have neural, genitourinary and gastrointestinal skeletal abnormalities [1, 2, 13]. In our observation, in addition to the diaphragmatic hernia, our newborn had a congenital hydrocele which spontaneously regressed.

Bochdalek's hernia accounts for 80-90% of all diaphragmatic hernias. About 85% of them occur on the left side, 10% on the right and about 5% are bilateral [14].

Their diagnosis is in 70% of cases is made antenatal. The calling sign on ultrasound is a deviated heart and compressed lung caused by the thoracic ascension of the abdominal viscera [5]. The presence of the liver in intrathoracic form in itself constitutes a criterion of seriousness. A hernial sac may be present. It is associated with a better prognosis [5].

Fetal MRI is increasingly used to confirm the diagnosis of HDC in the second and third trimester as a complementary tool to ultrasound studies to better define the fetal anatomy. Its main advantage is its ability to measure total lung volume and allow an assessment of herniated liver mass in the chest [14].

Bochdalek's hernia can be diagnosed postnatal. Clinically, this malformation is most often manifested by the classic triad: respiratory distress, displacement of heart sounds and flat abdomen [15]. At most, this is a picture of acute asphyxia at birth. Sometimes this is a picture of progressive respiratory distress [15]. The frontal thoraco-abdominal radiograph confirms the diagnosis by showing digestive hydroaeric images in the thorax, a displacement of the mediastinum on the side opposite the lesion and a hemithorax occupied by the cardiac silhouette in case of left congenital posterolateral hernia [15].

The thoraco-abdominal scanner was our diagnostic radiological means in the face of the ambiguity and insufficiency of a previously performed chest X-ray.

The management of Bochdalek's hernia includes: neonatal management and surgical management [5].

Neonatal care is now well codified in developed countries. Its principle is to minimize pulmonary aggression, hypoxia and acidosis in order to limit pulmonary arterial hypertension which is constant and can by itself put a strain on the prognosis [5].

It consists of intubating the newborn and transferring it to a neonatal intensive care unit for preoperative stabilization. Optimally, this transfer takes place within the same hospital where the child can benefit from specific care as early as possible [15].

The most currently accepted treatment strategy includes high frequency oscillation assisted ventilation (HFO), early administration of exogenous surfactant, and inhalation of nitric oxide [15].

Surgical management includes: antenatal surgery and postnatal surgery [5]. The proposed antenatal surgery is tracheal obstruction fetoscopy. It is indicated in cupola hernias with a poor prognosis [5].

Postnatal surgical repair of Bochdalek's hernias begins with reduction of the herniated organs followed by exploration of the diaphragm. If a tension-free suture is possible with a good quality diaphragm, muscle closure is achieved [5]. In the event of a hernia with a sac, it is indifferently preserved or resected [5]. If the approximation of the edges requires too much traction of the diaphragm, a prosthetic plate is placed and most often in polytetrafluothylene (5). Muscle flaps are used by some teams. The repair difficulties are linked to the size of the defect, but this size is also a very good indicator of pulmonary severity and of the prognosis [5].

In newborns, the preferred approach is laparotomy, for left or right hernias, thoracoscopy being reserved for good shapes [5]. Indeed, even in the teams accustomed the rate of recurrences after repair by thoracoscopy remains higher, evaluated around 30% against 10% after open surgery. The reasons for this difference are not well identified [5].

Our newborn was born by cesarean section in a gyneco-obstetrics department without antenatal diagnosis and then transferred to our neonatal unit by public transport. There is no equipment for neonatal resuscitation.

We performed classical surgery which allowed the herniated organs to be pushed back and the diaphragmatic defect to be closed. This classic surgical approach has been the preference of other authors [3]. The laparoscopic approach remains possible but is not yet within our reach. The herniated organs found intraoperatively in our observation were the left lobe of the liver and the right colonic angle. For some authors it was only the colon [3] or the right kidney [10]. The

postoperative follow-up was straightforward with a 6-month follow-up.

CONCLUSION

Congenital diaphragmatic hernia is a rare condition. The posterolateral form is more common. Damage to the right dome is rarer than that of the left dome. Respiratory distress is their main mode of disclosure at birth. In the face of any neonatal respiratory distress, the hypothesis of HDC must be suggested and confirmed by a chest x-ray.

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