Congenital diaphragmatic eventration complicated with neonatal gastric perforation.

Eventration congénitale du diaphragme compliquée d'une perforation gastrique

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RÉSUMÉ

L'éventration congénitale du diaphragme est un défaut de développement rare et habituellement asymptomatique. La perforation gastrique néonatale est également une pathologie rare, mais pouvant engager le pronostic vital chez le nouveau-né. L'association de ces deux pathologies a été rarement rapportée. Nous rapportons le cas d'un nouveau-né de sexe masculin à terme issu d'une grossesse et d'un accouchement sans incident et qui avait un examen clinique normal à a naissance. Le nouveau-né a été admis le 4ème jour de la vie dans un tableau de choc hémodynamique, ballonnement et contracture abdominale et de refus de téter suggérant une péritonite. La radiographie du thorax et l'échographie ont conclu à une hernie diaphragmatique. Une laparotomie a été réalisée après une brève réanimation et a mis en évidence une éventration diaphragmatique avec perforation gastrique. Une suture de la perforation gastrique avec une plicature diaphragmatique ont été réalisée avec l'évolution favorable.

Mots-clés

La perforation gastrique , éventration diaphragmatique , congénitale , nouveau-né

SUMMARY

Congenital diaphragm eventration is a rare and usually asymptomatic developmental defect. Neonatal gastric perforation is also a rare but life-threatening condition. In our knowledge, the association of these two pathologies has been, exceptionally reported. We report a case who illustrates clinical and radiological features of this possible co-morbidity. A full-term male neonate was born from uneventful pregnancy and delivery. The antenatal scan was reported as normal. At birth, clinical exam was normal, no special resuscitation was necessary. The newborn was examined and admitted the 4th day of life for fever, tachypnea, cyanosis, hemodynamic shock and refusing feeds. Clinical examination suggested peritonitis. Chest radiography and ultrasonography suggested congenital hernia. A laparotomy was performed after a brief resuscitation and confirmed the presence of diaphragm eventration with gastric perforation. Suturing of gastric perforation with a diaphragmatic plication was performed with favorable evolution.

Key-words

Gastric perforation , Congenital diaphragmatic , eventration , neonate

Congenital Diaphragm eventration (CDE) is a rare developmental defect with an incidence of 1 in 10.000 live births [1]. CDE is characterized by aplasia of a part or the whole of the diaphragm muscle which is replaced by fibroblastic tissue leading to the displacement of hemi diaphragm into the thorax [2]. While usually asymptomatic and accidentally diagnosed [3], CDE can be associated with life-threatening situations especially in new-born and infant.

Neonatal gastric perforation (NGP) is also a rare but life-threatening condition [4]. In our knowledge, the association of CDE with NGP has been, exceptionally reported in the literature [5].

We aimed to illustrate clinical and radiological features of this possible co-morbidity.

CASE

A 3655 g term male neonate was born to 23 years old gravid 2 lady by normal vaginal delivery. The antenatal period was uneventful. The antenatal scan was reported as normal. At birth, Apgar scores were 8/10 at 1 min and 9/10 at 5 min, clinical exam was normal, no special resuscitation was necessary. At 3rd day of life, he presented fever, bilious vomiting, refusing feeds and jaundice. The newborn was examined and admitted 24hours later in the Resuscitation and Intensive Care Unit of Neonatology of Military Hospital of Tunis. The vital parameters reveled fever at 39.8°C, heart rate of 175/min, capillary refill time of 3 sec, blood pressure of 60/49 mmHg, tachypnea of 70/ min with Silverman score of 4/10, oxygen saturation of 90 % in air. There was bowel sound heard on left lung base. The heart sound was shifted to right. Jaundice and peritonitis clinics signs were noted: distended abdomen with generalized contracture, bilious gastric residuals, and absence of stools. Chest and abdominal X-ray (figure 1) showed a left intra thoracic digestive clarity surmounted by left upper lobe opacity with respect to a lung collapse.

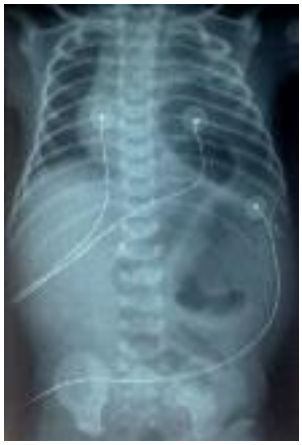

Figure 1 : Preoperative chest and abdominal radiography shows left intra thoracic digestive clarity, diffuse ground-glass opacity with paucity of bowel gas

The ultrasonography showed the diaphragmatic discontinuity associated with an intra peritoneal effusion of average abundance.

After a brief resuscitation, a laparotomy was performed and noted a left diaphragmatic eventration and a longitudinal perforation at the back surface of the greater gastric curvature.

A surgical suturing of gastric perforation with a diaphragm plication was performed with favorable evolution. The infant was well at the age of 15 months.

DISCUSSION

CDE is a rare and usually asymptomatic developmental defect, which is generally, diagnosed accidentally [3]. Neonatal gastric perforation (NGP) is also a rare but life-threatening condition. Early diagnosis of NGP is difficult because most patients usually present with nonspecific signs such as abdominal distension and lethargy [4]. It often occurs without any apparent precipitating event with rapid deterioration [5]. In our knowledge, the association of these two pathologies has been, exceptionally reported [5]. It was admitted that CDE results from inadequate development of the diaphragm muscle or absence of the phrenic nerves [1]. However, the exact etiology of CDE remains unknown, although it may be associated with fetal rubella or cytomegalovirus infection [6].

Otherwise, NGP may occur in patients with mechanical obstruction distal to the stomach, such as duodenal atresia or malrotation [7], or with iatrogenic perforation (e.g., feeding tube placement) [4]. NGP is likely to be more frequent in premature and low birth weight neonates [5]. All these conditions were lacking in our patient. Although many theories regarding the pathogenesis of NGP have been proposed, the etiology remains varied and may be combined [7].

NGP was originally thought to result from congenital anatomic defects of the gastric musculature among neonates, especially in prematurity [5]. Nevertheless, Shaw et al [8] suggested that NGP was caused by a mechanical rupture of the stomach secondary to increased intragastric pressure, rather than a congenital agenesis of the gastric muscle. In the studies of Lin CM et al [5], Yan CY et al [4], and in our report, the associations of NGP with digestive malrotation, hiatal hernia, and CDE in the affected neonates further support Shaw's theory. The physiopathology of NGP may include an intragastric hyper pressure leading to a rupture of gaps normally present in the circular muscle layer of the newborn stomach. Theses gaps are most prominently in the fundus, near the greater curvature [5] and may have little clinical significance under normal circumstances. However, they are potential weak points in the stomach wall that might be susceptible to rupture if intragastric pressure increases [5]. CDE may favorite NGP by

accidentally increase of intragastric pressure.
CDE was reported in a male newborn infant as commonly reported [9]. While partial defects mostly affect the right hemidiaphragm, diffuse defects tend to be unilateral and more commonly affect the left side [9]. Early clinical manifestations of CDE are nonspecific and not evident. Thus, they could often lead to misdiagnosis [9]. CDE must be, differenced from DH. Diaphragm is uniform in eventration but with sac in DH. There is no pulmonary hypoplasia in CDE compared to DH, therefore undetected at birth and is picked up as a coincidental finding [6].
While usually asymptomatic and incidentally diagnosed, CDE may have diverse features. Clinical manifestations range from mild gastrointestinal symptoms, progressive dyspnea or respiratory infection to life-threatening respiratory distress requiring mechanical ventilation support [2, 6].
The main symptoms in CDE are related to the elevated intra-abdominal organs resulting in compression of the lower lobe of the lung [9]. Newborns may present with dyspnea, cyanosis, acute respiratory distress, vascular dysfunction, and cardiac symptoms. Anorexia, nausea, and vomiting secondary to gastric volvulus may also be associated [1, 9].
The diagnosis of isolated CDE can be established by routine chest radiographs and fluoroscopy. Conventional chest radiography usually suggests ED demonstrating an elevated diaphragm [10]. Fluoroscopy is considered the most reliable way to document eventration showing reduced, paradoxal or absent movement of diaphragm [10]. Upper gastrointestinal contrast, chest computed tomography scan, magnetic resonance imaging scan, ultrasound scan, or isotope scan could also help to identify diaphragmatic paralysis and DH [9]. Ultrasonography may be an effective imaging for diagnosis. The diaphragm can been seen as a continuous thin layer above the elevated abdominal viscera with reduced mobility [10].
In our case, CDE was confounded with DH. The association of radiologic signs of gastric perforation with those of CDE contributed to the misdiagnosis of CDE. The diagnosis was performed while laparotomy.
CDE can be isolated or associated with other developmental defects. The main associated findings include hypoplastic lung, congenital heart disease, Horner's syndrome, contralateral ptosis, chest wall deformities, hypoplastic ribs, cleft palate, hemi vertebrae, kidney ectopia, gastric volvulus, and clubfoot [1, 9].
Diaphragmatic plication is the treatment of choice in case of CDE [2, 6]. It stiffens the diaphragm, reduces paradoxical motion, and increases hemithorax volume, restoring pulmonary functions. This operative procedure is simple, safe, and effective [2, 10].
Thoracoscopic surgery may be a suitable alternative to the conventional operative technique avoiding the problems of open surgery [3].

CONCLUSION

While usually asymptomatic, CDE may be associated with diverse life-threatening situations requiring resuscitation and/or emergency surgery especially, in newborn.
NGP is a rare complication in case of CDE, probably due to increase of intragastric pressure. Chest Ultrasound is generally an important imaging modality for diagnosis. Diaphragmatic plication especially using endoscopic surgery seems to be safe and effective in the management of symptomatic patient. In the present case, even the diagnosis of CDE was difficult in presence of clinical and radiological findings of NGP and peritonitis, the patient was managed successfully with emergency resuscitation, gastric suturing and diaphragm plication.

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