Diaphragmatic eventration in children

Eventration diaphragmatique chez l'enfant

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RÉSUMÉ

Prérequis : L'éventration diaphragmatique (ED) est une surélévation permanente du diaphragme sans solution de continuité. Les manifestations cliniques sont diverses, allant des formes asymptomatique à une détresse respiratoire mortelle.

Le but de cette étude était d'évaluer notre expérience dans la prise en charge de 8 enfants hospitalisés pour éventration diaphragmatique au cours des 15 dernières années.

Méthodes : Etude rétrospective colligeant huit cas d'ED au département de chirurgie pédiatrique de l'hôpital d'enfant de Tunis.

Résultats : Cinq patients (2 garçons et 3 filles) dont l'âge a varié de 5 mois à 7 ans, ont bénéficié d'une plicature diaphragmatique (3 du côté droit et 2 du côté gauche) pour ED symptomatique avec une amélioration spectaculaire de leur état respiratoire. Un autre patient (garçon de 3 mois) a été admis pour une détresse respiratoire ayant nécessité une ventilation mécanique. Il est décédé avant l'opération dans un tableau de septicémie. Deux autres enfants, suivis pour ED de découverte fortuite, étaient asymptomatiques pour une durée moyenne de 5 ans. La radiographie thoracique a suggéré l'éventration dans tous les cas. L'échographie a permis le diagnostic dans 6 cas. Aucun cas de malformation associée n'a été noté.

Conclusion : L'échographie est un examen radiologique important pour le diagnostic. La plicature diaphragmatique semble être sûre et efficace. La chirurgie n'est pas recommandée pour les enfants asymptomatiques.

Mots-clés

Eventration diaphragmatique, plicature, enfant

SUMMARY

Background: Diaphragmatic eventration (ED) is a rare anomaly defined by a permanent elevation of a hemidiaphragm without defects. Clinical manifestations are diverse, ranging from asymptomatic to life-threatening respiratory distress. The aim of this study is to report our experience of management of eight children with ED over the past 15 years.

Methods: A retrospective study was conducted involving 8 infants and children with ED managed at the department of pediatric surgery of Tunis Children's Hospital.

Results: Five patients (2 males, 3 females) whose ages ranged from 5 month to 7 years (mean, 13 months) were operated on using diaphragmatic plication (3 right-sided and 2 left-sided plications) for symptomatic ED with a dramatic improvement in their respiratory status. Another infant (a 3-month-old boy) was admitted for respiratory distress that required mechanical ventilatory support. He died before operation because of sepsis. Two other asymptomatic patients with incidentally diagnosed ED were followed up for 5 years on average. Chest x-ray suggested eventration in all cases. Ultrasound was found to be a useful modality for diagnosis in 6 cases. There were no cases of associated malformation.

Conclusion: Diaphragmatic eventration in children is usually congenital but may be acquired. Chest ultrasound is an important imaging modality for diagnosis. Diaphragmatic plication appears to be safe and effective. Surgical intervention is not recommended in asymptomatic patients.

Key-words

Diaphragmatic eventration, plication, children

Diaphragmatic eventration (ED) is an abnormal elevation of part or the whole of the hemidiaphragm as a result of paralysis, aplasia or atrophy of varying degrees of muscle fibres. It is usually congenital but may be acquired. The incidence is uncertain, although it is known as a rare condition. Clinical manifestations in infants and children are diverse, ranging from asymptomatic to life-threatening respiratory distress. Although ED may be amenable to conservative treatment, symptomatic ED in children may require surgical treatment. The aim of this study is to report our experience of management of eight children with ED over the past15 years.

METHODS

A retrospective study was conducted involving 8 infants and children with ED managed at the department of pediatric surgery of Tunis Children's Hospital, over a 15-year period, going from 1998 through 2012. Information considered included clinical presentation, radiological findings, and results of treatment. The report discusses the question of birth trauma to brachial plexus. Surgical treatment was required for five patients consisting of plication of the diaphragm through an intercostal approach: Duval grasper was used to grasp and invaginate the apex of the eventration downward into the abdomen, thus creating a transverse fold from the periphery to the cardiophrenic angle behind the phrenic nerve. This fold is closed first using a series of interrupted nonabsorbable sutures. A second series was carried out similarly, thus burying the first series of suture lines. Stitches were inserted through a more peripheral portion of diaphragm to obtain the desired tension of the diaphragmatic dome. One infant died before operation and two others were followed up for asymptomatic ED.

RESULTS

Five patients (2 males, 3 females) aged between 5 months and 7 years (mean, 13 months) were treated using diaphragmatic plication. All of these patients experienced symptoms of recurrent pneumonia (4 cases), dyspnea (2 cases), and vomiting (1 case). A lesion of the brachial plexus was found in one infant. Chest radiography was revealed total elevation of the right dome in 3 cases and of the left dome in 2 cases above the fourth intercostal space (figure1) and a mediastinal shift in 2 cases.

Figure 1 : Preoperative chest radiography shows elevation of the right herni diaphragm

On ultrasound, hemidiaphragm was found to be thinned out but intact with reduced movement in 3 cases. Complete immobility or paradoxical movements were not seen. CT scan indicated in 2 cases where the diagnosis was still in doubt. Fluoroscopy was not realized in any case. The postoperative course was uneventful in all cases. There was no recurrent or residual eventration that necessitated reoperation in any patient. Follow-up time ranged from 6 months to 12 years with good clinical and radiological course (figure 2,3). Another infant (a 3-month-old boy) was admitted for respiratory distress that required ventilatory support. Chest X-ray revealed elevation of left hemidiaphragm. Ultrasound and thoracic computed tomography scan confirmed the integrity and reduced mobility of the diaphragm. The patient died before operation because of sepsis. Two other asymptomatic patients with left ED diagnosed incidentally were followed up. Chest ultrasound confirmed the eventration in both cases. They didn't experience any symptoms after a mean follow-up time of 5 years. There were no cases of associated malformation.

Figure 2 : Preoperative lateral chest film shows a diaphragm elevated more than 2 intercostal spaces over the normal side

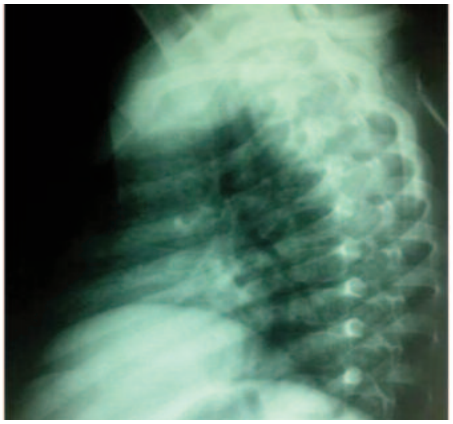
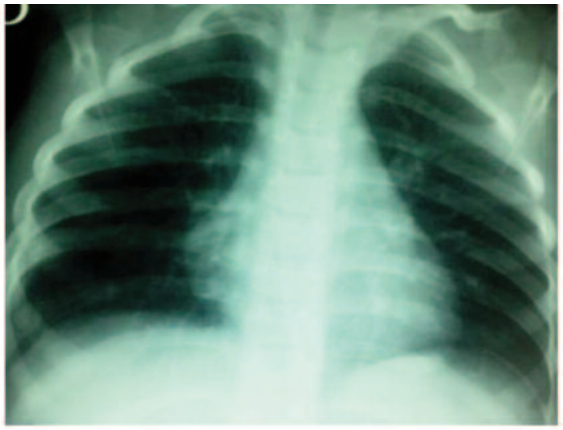


Figure 3 : Postoperative chest radiograph shows dramatic improvement



DISCUSSION

Eventration of diaphragm can be congenital or acquired. Congenital eventration results from inadequate development of the muscle or absence of the phrenic nerves. The most common cause of acquired

eventration is injury to the phrenic nerve, resulting from either birth trauma or thoracic surgery for congenital heart disease (1,2). A lesion of brachial plexus (seen in one case in our series) indicated acquired eventration. The terms eventration and paralysis are often confused: paralysis may be the instigator of eventration because of degenerative muscular changes after denervation, whereas eventration is not always associated with paralysis. Nevertheless, both eventration and paralysis have the same consequences (3). ED is more commonly left sided (4), like in our series (62.5%). In infants and children who have diaphragmatic eventration, clinical manifestations are diverse, ranging from mild gastrointestinal symptoms to life-threatening respiratory distress requiring mechanical ventilatory support (5). While usually asymptomatic, diaphragmatic eventration can cause progressive dyspnea on exertion or respiratory infection in small children, especially in those who are less than 2 years of age (5). In our series, recurrent ipsilateral pneumonias were noted in four cases (50%). Eventration must be differentiated from diaphragmatic hernia. In eventration, the diaphragm retains its continuity and attachments to the costal margin. With diaphragmatic hernia, in contrast, this continuity between the diaphragm and the costal margin is disrupted. Diaphragm is uniform in eventration but with sac in diaphragmatic hernia. There is no pulmonary hypoplasia in eventration compared to diaphragmatic hernia, therefore undetected at birth and is picked up as a coincidental finding (2,4). Conventional chest radiography suggested ED demonstrating an elevated diaphragm. Fluoroscopy is considered the most reliable way to document eventration showing reduced, paradoxal or absent movement of diaphragm. Ultrasonography is an important imaging modality for diagnosis. It was effective in 75% of our patients. The diaphragm can be seen as a continuous thin layer above the elevated abdominal viscera with reduced mobility. CT scan and magnetic resonance imaging may be performed as adjuvant techniques in cases where the diagnosis still remains in doubt (1,4,6). Associated findings include Horner's syndrome, contralateral ptosis, and chest wall deformities with and without missing ribs, gastric volvulus, coarctation of the aorta, cleft palate, hemivertebrae, congenital heart disease, renal ectopia, hypoplastic aorta, situs inversus, club foot…(4).

Diaphragmatic plication was indicated in symptomatic patients. It increases hemithorax volume, stiffens the diaphragm, and reduces paradoxical motion, improving pulmonary functions. The operative procedure is safe, simple and suitable. It improves symptoms dramatically (1,5), like in our series. Recently, there have been reports that thoracoscopy is effective in diagnosis and treatment of patients with diaphragmatic disorders. Thoracoscopic or minimally invasive operation may be an acceptable alternative to the conventional operative technique.

The benefits of endoscopic surgery are evident in the literature and include smaller incision, quicker recovery, and decreased pain (7,8,9). If the patient experiences no or a few symptoms, they should still be strictly followed-up to promptly indicate the necessity for operation in the event of an even slight deterioration in respiratory function (5,6).

CONCLUSION

Diaphragmatic eventration in children is usually congenital but may be acquired. Chest Ultrasound is an important imaging modality for diagnosis. Diaphragmatic plication appears to be safe and effective in the treatment of patient with recurrent ipsilateral pneumonia. Laparoscopic diaphragmatic plication circumvents the limitations of open transthoracic and appears to be as effective. Surgical intervention in asymptomatic patients is not recommended according to the literature to date.

References

1. Tiryaki T, Livanelioglu Z, Atayurt H. Eventration of the diaphragm. Asian J Surg.2006; 29:8-10.
2. Yazici M, Karaca I, Arikan A, et al. Congenital eventration of the diaphragm in children: 25 years' experience in three pediatric surgery centers. Eur J Pediatr Surg.2003; 13:298-301.
3. Verhey PT, Gosselin MV, Primack SL, et al. Differentiating diaphragmatic paralysis and eventration. Acad Radiol.2007; 14:420-5.
4. Kulkarni ML, Sneharoopa B, Vani HN et al. Eventration of the diaphragmatic and associations. Indian J Pediatr 2007; 74: 202-205.
5. Tsugawa C, Kimura K, Nishijima E, et al. Diaphragmatic eventration in infants and children: is conservative treatment justified?. J Pediatr Surg.1997; 32:1643-4.
6. Kansal AP, Chopra V, Chahal AS, et al. Right-sided diaphragmatic eventration: A rare entity. Lung India.2009; 26:48-50.
7. Borruto FA, Ferreira CG, Kaselas C, et al. Thoracoscopic Treatment of Congenital Diaphragmatic Eventration in Children: Lessons Learned After 15 Years of Experience. Eur J Pediatr Surg. 2013 Jun 19.
8. Groth SS, Rueth NM, Kast T et al. Laparoscopic diaphragmatic plication for diaphragmatic paralysis and eventration: an objective evaluation of short-term and midterm results. J Thorac Cardiovasc Surg.2010; 139:1452-6.
9. Becmeur F, Talon I, Schaarschmidt K, et al. Thoracoscopic diaphragmatic eventration repair in children: about 10 cases. J Pediatr Surg.2005; 40:1712-5.