MECONIUM THORAX: A CASE OF BOCHDALEK HERNIA AND CECAL PERFORATION WITH CYSTIC PERIVENTRICULAR LEUKOMALAISIA IN A NEONATE

Meral Barlas* ✣ Aydın Yağmurlu**

SUMMARY

A 35 week pre-term male neonate with a prenatal history of polyhydroamnios and intrauterine growth retardation was delivered from a 25 year old mother. Chest radiograph and CT showed pleural effusion, and mediastinal shift to the right. The patient undervent an exploratory laparotomy and a posterolateral left diaphragmatic hernia was identified as well as 3mm perforation of a normally positioned cecum. The left pleural space was cleared of debris, then the diaphragmatic defect was closed. After appendectomy and colonic biopsy cecostomy was performed. His postoperative course was uneventfull. However, the infant presented a spastic posture, opistotonic movement and tonic-clonic seizures. Cystic encephalomalasic areas in both hemispheres, loss of volume in white matter and cortical atrophy were detected by MRI. Performing colostomy closure he was discharged to the pediatric neurology department. At 8 months and 3 years follow up, the child has no problems due to CDH, but he has very severe neurologic deficit and is mentally retarded.

Key Words: *Congenital Diaphragmatic Hernia, Cecal Perforation, Cystic Periventricular Leukomalaisia.*

ÖZET

Mekonyum Toraks; Bochdalek Hernisi Olan Bir Yenidoğanda Çekal Perforasyon ve Kistik Periventriküler lökomalazi

Gelişme geriliği ve polihidroamniyos nedeniyle 25 yaşındaki anneden normal vajinal yolla doğan 2500 gr ağırlığındaki 35 haftalık prematür erkek yenidoğan, sol Bochdalek herni öntanısıyla yatırıldı.Radyolojik görüntüleme yöntemlerini takiben laparatomi yapıldı. Sol hemitoraksta ve karın içindeki mekonyum bulaşısının, normal yerleşimli çekumdaki 3 mm perforasyondan kaynaklandığı diğer barsakların normal çap ve görünümde olduğu saptandı. Karın ve toraks yıkanıp temizlendi, diyafrağma defekti dikilerek onarıldı. Apendektomi, Kolon biyopsisi ve çekostomi yapıldı, cerrahi girişim sonrası 7. gün şifa ile taburcu edildi. Hasta 4 aylık iken spastik kasılmaları nedeniyle MRI yapıldı. Tüm beyinde kistik oluşumlar, beyaz cevherde hacim kaybı ve kortikal atrofi saptandı. Çekostomi kapatılarak, çocuk nörolojisi kliniğine yollandı. Gelişme geriliği olmayan 3 yaşındaki hastada KDH nedeniyle ilgili bir komplikasyon yoktur, ancak ileri derecede nörolojik defisit ve mental retardasyonla yaşamını özel eğitimle sürdürmektedir.

Anahtar Kelimeler: *Bochdalek Hernisi, Çekal Perforasyon, Kistik Ensefalomalazi.*

The incidence of associated malformations in infants with congenital diaphragmatic hernia (CDH) is approximately 30-40 percent. The predominance during these associated anomalies is in neural tube defects including anencephaly, myelomeningocele, hydrocephalus and encephaloceles. Cardiac defects are the second most common group (1-3). Periventricular leukoencephalopathy is a pathologic process that has attracted little attention in neurodiagnosis. However its association with extremely low birth weight infants, hypocarbia and mechanical ventilation is well demonstrated (4). We present herein as a case of CDH and cecal perforation and cystic periventricular leukomalasia.

* *Associate Professor, Department of Pediatric Surgery, Ankara University School of Medicine, 06100 Ankara, TURKEY*

** *Clinical Fellow / Instructor, Department of Pediatric Surgery, Ankara University School of Medicine, 06100 Ankara, TURKEY*

Received: *May 20, 2002* Accepted: *Sept 30, 2002*

Case Report

A 35-week-preterm male neonate with a prenatal history of polyhydroamnios and intrauterine growth retardation was delivered from a 25 year old mother via spontaneous vaginal route. The baby weighed 2500 g. APGAR scores were 5 and 8 at 5 and 10 minutes respectively.

Cyanosis and respiratory distress required as rush to the neonatal intensive care unit. He received oxygen in the hood. On physical examination the respiratory sounds were diminished in the left side and the heart sounds were displaced to the right. With the suspect of CDH a radiocontrast enema was performed and the colonic segments under the diaphragma was observed. A normal sized and placed cecum, contrast leaked into an ill-defined collection in the right lower quadrant. Also the upper gastrointestinal series with radiocontrast material revealed no abnormality. Chest radiograph and CT showed pleural effusion, and mediastinal shift to the right. The patient undervent an exploratory laparotomy on day 2 of life. A posterolateral left diaphragmatic hernia was identified as well as 3mm diameter perforation of a normally positioned cecum (Figure 1). A significant degree of meconium staining was seen throughout the peritoneal cavity and thorax. The left pleural space was cleared of debris then the diaphragmatic defect was closed, after which appendectomy and colonic biopsy cecostomy was performed. Pathologic examination showed a congenital deficiency of the muscularis propria in the perforated area and normal ganglion cells. The postoperative period was uneventful. No mechanical ventilation was required, and he was discharged from the hospital in the 10th postoperative day. Cecostomy closure was planned. The child presented four months later with a spastic posture, opistotonic movement and tonic-clonic seizures. Magnetic resonance imaging revealed cysticencephalomalasic areas in both hemispheres, loss of volume in white matter, and cortical atrophy (Figure 2). The patient's screening for inborn errors of metabolism did not reveal any abnormality.

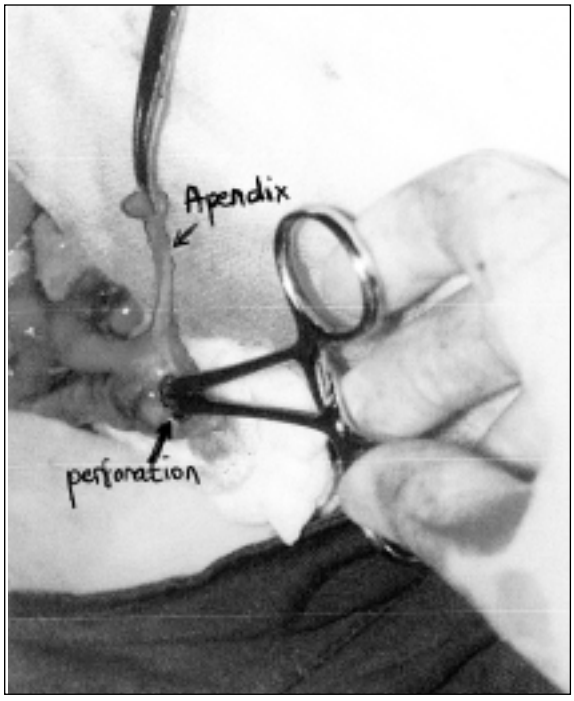

Figure 1: *Tip of Mosquito clamp directs to the cecal perforation. Please note the rest of the bowel in normal appearance.Operative photograph on day 2.*

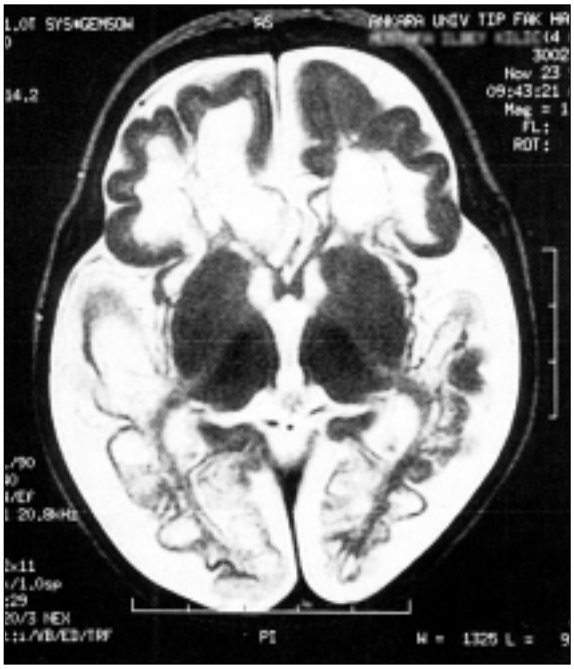

Figure 2: *Cystic periventricular leukomalasia in both hemispheres ,loss of volume in white matter, and cortical atrophy on T2 images in MRI at 4 months postoperatively*

Performing colostomy closure he was discharged to the Pediatric Neurology Department for this rare neurological entity. At 8 months and 3 years follow up, the child has no problems due to CDH, but he has a very severe neurologic deficit and is mentally retarded.

Discussion

Although neonatal care has improved over the past 20 years, congenital diaphragmatic hernia remains as an anomaly with a high mortality rate (3). This is not only due to the defect itself but also a combination of associated anomalies. Cardiovascular malformations and neural tube defects including anencephaly, myelomeningocele, hydrocephalus, and encephaloceles have a predominance among these anomalies (1,2,5). The rate of gastrointestinal disorders remains low. Patole et al, have presented a case in which extension of meconium peritonitis through muscular defect in the diaphragm lead to intrathoracic calcifications diagnosed ultrasonographically at 23 weeks of gestation (6). Butterworth et al. reported third meconium torax with Job's syndrome (3). The case we present here is the second one in the English literature with CDH and gastrointestinal perforation presented as fecaloid material in the thorax. The reason of cecal perforation has not been clearly defined. Except the perforation site, rest of the intestines were macroscopically normal both vascularisation and diameter. The histopatologic examination excluded Hirschsprung's Disease.

The association of neurological defects seems to be high with CDH but this is the first case of congenital leukoencephalopathy. It was reported by Wisewell et al. that mechanically ventilated premature infants are at increased risk for cystic periventricular leukomalasia, particularly if hypocapnia occurs (4). The presented case had no need for mechanical ventilation because of the small diaphragmatic defect and lung hypoplasia was not severe. Though it is hard to differentiate the congenital form from the acquired one, we believe that this pathologic entitiy is a congenital form which was previously demonstrated in three Turkish children by Oliver et al. (7). The neurological findings were noted in the first months of life and include spasticity and impairment of motor and mental retardation, just like the present case with spastic posture and opistotonic movements and tonic-clonic seizures. Magnetic resonance imaging of the brain showed extensive cysts within the cerebral hemispheres, ventricular enlargement and white matter disease similar to the previously reported three cases.

To conclude; meconium Thorax in CDH with cecal perforation is a rare case especially when accompanied with cystic periventricular leukomalacia.

REFERENCES

1. Kaiser JR, Rosenfeld CR: A population based study of congenital diaphragmatic hernia: Impact of associated anomalies and preopertive blood gases on survival. J Pediatr Surg 1999;4: 1196-1202
2. Langer CL, Harrison MR: Congenital diaphragmatic hernia and evantration of the diaphragm. In: Puri P (ed) Newborn Surgery, Butterworth, Heinemann, Oxford, 1995;pp209-216
3. Butterworth SA,Weber EM Meconium torax: A case of Bochdalek Hernia and cecal perforation in a neonate with Job's syndrom, J Pediatr Surg 2002;37:673-674
4. Wisewell TE, Graziani LJ, Kornhauser MS et al: Effecs of hypocarbia on the development of cystic periventricular leukomalaisia in premature infants treated with high frequency jet ventilation. Pediatrics 1996;98: 918-924
5. Ahmad A, Gangitano E, Odell RM et al: Survival, intracranial lesions, and neurodevelopmental outcome in infants with congenital diaphragmatic hernia treated with ECMO. J Perinatol 1999;19:436-40
6. Patole S, Whitehall J, Almonte R et al.: Meconium thorax: case report and review of the literature. Am J Perinatol 1998; 15:53-56
7. Olivier M, Lenard HG, Aksu F et al :A new leukoencephalopathy with bilateral anterior temporal lobe cyts. Neuropediatrics 1998; 29:225-228