DELAYED PRESENTATION OF RIGHT-SIDED DIAPHRAGMATIC HERNIA AFTER MECHANICAL VENTILATION IN A NEWBORN WITH HYALINE MEMBRANE DISEASE: A CASE REPORT

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SUMMARY: Delayed presentation of right-sided diaphragmatic hernia occurred after the improvement of hyaline membrane disease in a male newborn. He presented with respiratory distress during the first few hours of life and had hyaline membrane disease confirmed by chest roentgenogram. After initial improvement with antibiotic therapy and ventilatory support for 2 weeks, sudden deterioration of respiration occurred and subsequent chest roentgenogram revealed herniated viscera in the right hemi-thorax. Two days ago his chest roentgenogram was normal. He did not have any other causes for diaphragmatic hernia such as group B streptococcal infection or necrotizing enterocolitis. To our knowledge, this association has not been previously reported in newborns.

Key Words: Diaphragmatic hernia, hyaline membrane disease, neonate.

INTRODUCTION

The majority of neonates with diaphragmatic hernia present as respiratory emergencies within the first 72 hours of life (1) and most of them (91%) has evidences of hyaline membrane disease (2). In a proportion (estimated between 5% to 30%) gastrointestinal or pulmonary symptoms occur beyond the neonatal period. However, this diagnosis is often not considered by physicians after the immediate neonatal period (3). Because the symptoms, signs and radiologic findings of patients with diaphragmatic hernia, presenting after this period, may be extremely varied and difficult to interpret. Delay in diagnosis may lead to misguided therapy, and to a potentially fatal outcome (4).

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Therefore, late-presentation of congenital diaphragmatic hernia (CDH) should be included in the differential diagnosis of any child with persistent gastrointestinal or respiratory problems associated with an abnormal chest x-ray film (5).

We report a case of an 18-day-old male newborn who presented with diaphragmatic hernia after the improvement of hyaline membrane disease (HMD).

This unusual presentation of diaphragmatic hernia following HMD was found to be unique following a search of the English literature.

REPORT OF CASE

A male neonate weighing 2750 grs was born to a 20-year-old woman (gravid 1, para 1, abortus O) at 37 weeks, gestation. The patient was born full-term after a

Figure 1: Anteroposterior chest x-ray of newborn at age of one day.

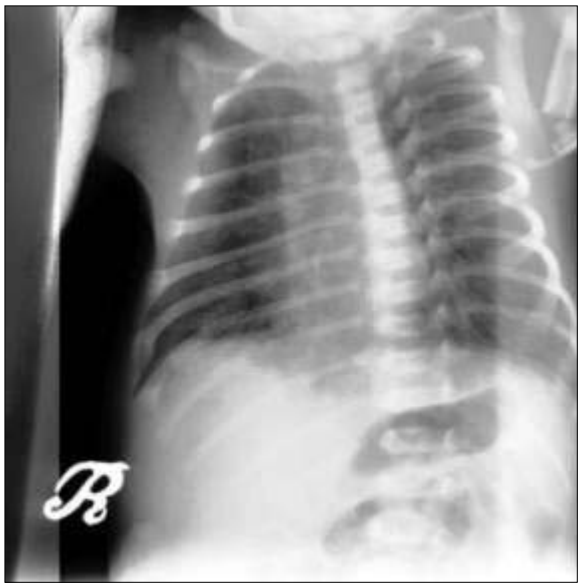
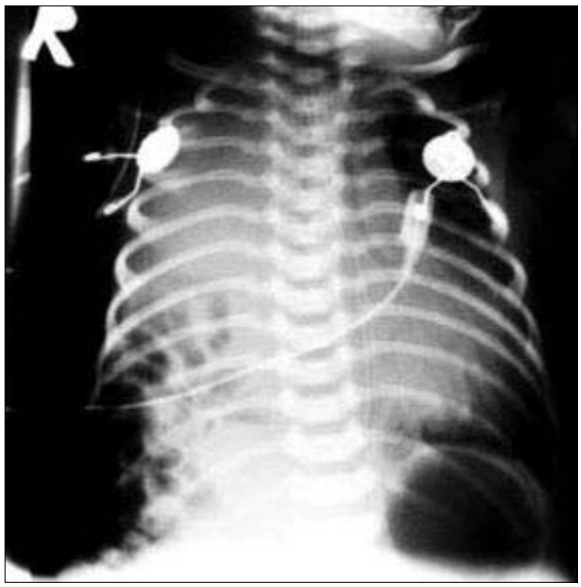


Figure 2: Anteroposterior chest x-ray film showing multiple loops of intestine in right hemi-thorax.



normal pregnancy and an uncomplicated vaginal delivery. Apgar scores were 6 and 8 at first and fifth minutes, respectively. The newborn was noted to have tachypnea and grunting respiration, and was placed in an oxygen hood. Culture of blood, urine, and cerebrospinal fluid were obtained, and the neonate was placed on a regimen of ampicillin and gentamicin. A chest roentgenogram was obtained and interpreted as alveolar haziness due to hyaline membrane disease (Figure 1). Assisted ventilation was required on the third day of life because of hypoxia and acidosis. Blood and CSF cultures were negative. The other blood tests revealed hemoglobin level of 16.8 gr/dl, hematocrit 50.8% and white blood cell count 8000/mm^3 with a differential of 25% polymorphonuclear cells and 73% of lymphocytes. C-reactive protein and ESR were normal. The oxygen requirement gradually decreased and neonate was extubated on the 15th day of life following a normal chest film. After 2 days sudden deterioration of respiratory status occurred and he necessitated mechanical ventilation for hypercapnia.

The patient's vital signs showed an axillary temperature of 36.6°C, a pulse rate of 156 beats per minute, and a respiratory rate of 35 respirations per minute. The lung fields were considered clear to percussion and auscultation. The liver was palpable below the edge of the right costal margin; the spleen tip was also palpable. Bowel sounds were noted in the abdomen, which was flat but not scaphoid. The remaining results of physical examination were normal. Chest x-ray films showed multiple loops of bowel in the right hemi-thorax (Figure 2). Repeated laboratory studies included cultures of blood, CSF, urinalysis, tracheal tube aspiration, and complete blood count, blood sugar, bilirubin, electrolytes, urea, creatinine, c-reactive protein, and ESR. All were found normal. Emergency consultation with pediatric surgeon was asked, but cardiorespiratory arrest possibly due to mediastinal compression by the herniated viscera occurred before surgical intervention.

DISCUSSION

Most of patients with CDH had evidence of diffuse alveolar damage and hyaline membrane formation even at full-term (6), which was more evident in the ipsilateral lung. But delayed presentation of right-sided diaphragmatic hernia after recovery from hyaline membrane disease in the neonate is an unusual clinical relationship.

Patients with CDH usually present soon after birth with respiratory distress. Occasionally presentation is delayed (7).

Radiologic findings of late-onset diaphragmatic hernia vary greatly from one case to another, and even in the

same patient at different times, because of differences in herniated organs size and intermittent spontaneous reduction (8). The spectrum of roentgenographic manifestations included: paravertebral mass, foreign material aspiration (8), lower lobe pneumonia (9), tension pneumothorax (4, 9, 10), congenital lung cyst (4), massive pleural effusion associated with reflex bowel ileus (11), massive pleuropneumonia (12), opacification of right hemi-thorax (13), left side haziness (14), and bowels in the hemi-thorax (13). The spectrum of clinical manifestations of delayed presentation of congenital posterolateral diaphragmatic hernia was described in 1959 by Kirkland in 34 adult patients. In 1976, Osebold and Soper described an additional 27 patients, many of whom were children and as in previous study, most of their patients had gastrointestinal or pulmonary complaints (1).

The etiology of late-onset diaphragmatic hernia has been debated. The role of invasiveness of the virulent group B streptococcus has been suggested as a cause by Harris *et al.* (15). All cultures of our neonate were negative. Giacocia and Jegathesen proposed that the abnormal compliance of the lung coupled with the use of positive pressure ventilation delayed the herniation of viscera or liver through the defect (16). Our neonate required positive pressure ventilation in 3rd day of life and two chest x-rays without CDH appearance. Banagale and Waters postulated that a decrease in intra-thoracic pressure, produced by weaning from mechanical ventilation, contributed to the delayed herniation (17). Our experience also support this theory, as the condition of our neonate deteriorated after apparent improvement in their pulmonary status and after discontinuing ventilatory support. Although the previous chest roentgenograms were normal, follow up roentgenogram demonstrated herniation of bowel loops into the right-side of the thorax.

It is likely that there are two groups of patients with diaphragmatic hernias; in one group it is suggested that the defect is long-standing, but the viscera are confined by a hernia sac or obturated by a solid organ. Presentation occurs when the sac ruptures or the intra-abdominal pressure is raised, or intra-thoracic pressure is decreased, causing the viscera to herniate. In this group previous chest radiographs may be normal. Other group has long-standing herniation, but who only present when a complication of the herniated contents such as volvulus or strangulation occurs (4).

Thus, the possibility of diaphragmatic hernia should be kept in mind to avoid a wrong diagnosis, undue delay in diagnosis, inappropriate treatment, and potentially fatal outcome. A normal previous chest radiograph does not exclude the diagnosis.

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