Delayed presentation of congenital diaphragmatic hernia in association with group B streptococcus infection in a preterm Omani neonate

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ABSTRACT

We present an interesting case of a preterm Omani newborn that had delayed onset of congenital diaphragmatic hernia in association with group B streptococcus infection. The association and the pathogenesis are supported by literature review. The message to follow is that any neonate with prolonged course of streptococcal pneumonia, with unusual course, should be investigated for presence of congenital diaphragmatic hernia.

Keywords: Congenital diaphragmatic hernia, group B streptococcal infection, neonate, delayed.

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The reported incidence of congenital diaphragmatic hernia is about 1 in 2000 to 1 in 5000 live births.[1] Mostly, it occurs on the left side.[2] In early neonatal life, Group B streptococcus is one of the most common organism causing pneumonia and sepsis. The association between the Group B streptococcus (GBS) pneumonia and the delayed presentation of congenital diaphragmatic hernia has been reported from different parts of the world.[3-8] We are reporting a case of delayed congenital diaphragmatic hernia in an Omani neonate, as no such case has been reported earlier from this region of the world.

Case Report. A 12 hour-old, preterm male infant was admitted to the neonatal intensive care unit with history of lethargy. He was born by spontaneous vaginal delivery to a 26 year old lady with history of pregnancy induced hypertension. There was no history of prolonged or premature rupture of membrane or maternal fever. Apgar score was 8 at one minute and 9 at five minutes. His birth weight was 1620 grams, with length of 42 cms and head circumference of 30 cms. His admission vitals were normal. On examination he was noted to be lethargic. No deformity was noted. A septic work up was carried out and he was started on ampicillin and gentamicin. His CBC revealed a WBC of 1.6 x 10^9/L, hemoglobin of 14 g/dl and platelet of 162 x 10^9/L. His first chest x-ray showed slight bilateral haziness with no signs of hernia (Figure 1). The blood culture grew group B streptococcus. He remained stable until 12 days of life, when his chest x-ray showed suspicion of right-sided diaphragmatic hernia (Figure 2). He was intubated and ventilated and was kept nil per oral. He remained on very low

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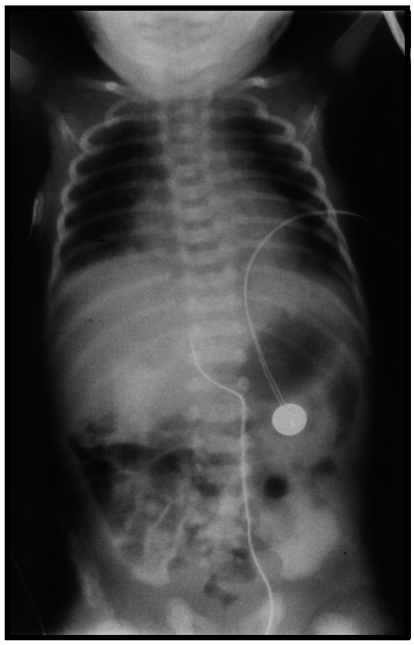

Figure 1 - Chest x-ray taken at birth (no signs of hernia.

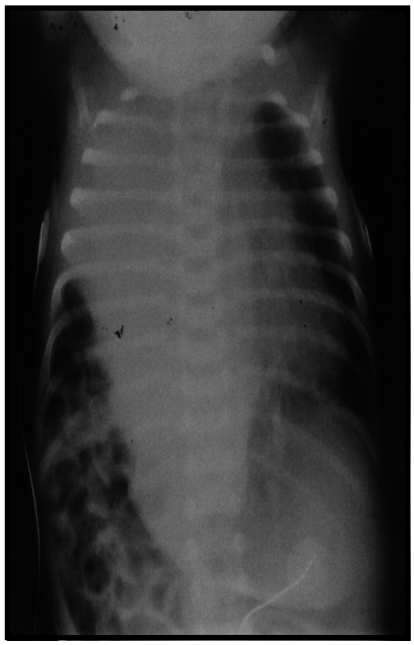

Figure 2 - Chest x-ray taken at day 12 (suspected right-sided hernia).

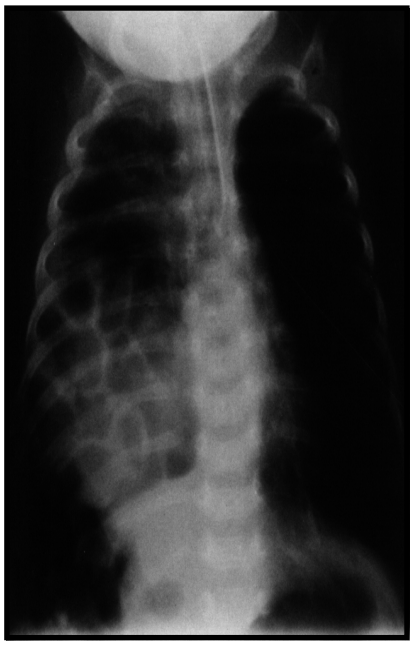

Figure 3 - Chest x-ray taken at day 27 (full-blown) hernia.

ventilatory settings. During the course an ultrasound and a series of follow up chest x-rays were carried out for confirmation of our suspicion, but nothing was confirmatory. The infant was extubated on day 22 of life and then started on feeds. On day 27 he developed respiratory distress. A chest x-ray revealed full-blown diaphragmatic hernia (Figure 3). He was again intubated and referred for surgery. Laparotomy revealed a large central defect in the right hemi-diaphragm with smooth edges. The content included the right lobe of the liver with gall bladder and the hepatic flexure of colon with no sac. The contents were reduced and the edges were approximated. The post-operative course was uneventful. Infant was extubated in 48 hours. He remained stable on feeds and was discharged home. On the last follow visit in the clinic, he was noted to be thriving well with good weight gain.

Discussion. Congenital diaphragmatic hernia is an uncommon cause of respiratory distress in neonates, occurring in 1 in 2000 to 5000 live births.[1] Approximately 85-90% occurs on the left side as compared to 15-20% on the right side.[2] The delayed presentation of congenital diaphragmatic hernia, associated with Group B streptococcus (GBS) pneumonia, has been reported earlier from east and west,[3-8] but no such case is reported earlier from middle eastern region of the world.

The pathogenesis of the delayed presentation of congenital diaphragmatic hernia with GBS infection is interestingly reported and several hypothesis have been put forward to explain the mechanism involved in this association. The situation is like the "chicken-and-the egg". Is it infection destroying diaphragm and causing hernia or hernia causing infection?

In the support of the theory that GBS infection causes necrosis and destruction of diaphragm resulting in herniation, Suresh et al[3] reported an interesting case. They found jagged and friable margins of the diaphragm in an infant with GBS infection and congenital diaphragmatic hernia. In their report the pathological findings also favored the acquired defect due to extension of GBS infection.

One the other hand, Ashcraft et al[4] have suggested that the GBS infection was secondary to the congenital diaphragmatic hernia. They hypothesize that movements of the defective diaphragm are dysfunctional which predispose to the development of pneumonia. The infective process causes atelectasis and tachypnea. These changes in the pulmonary mechanics lead to herniation through the defect. They further suggested that the tamponade by

the liver prevent the herniation through the defect earlier.

The other explanation for the delayed herniation was given by Bangale and Watters.[5] They suggested that inflammatory changes in the lung, especially right lower lobe, together with abnormal lung compliance could act as a splint for the defect, thereby delaying visceral herniation.

Looking at our case, it is difficult to establish a cause and effect relation between GBS infection and congenital diaphragmatic hernia. As noted in our case that the defect in the diaphragm was very smooth, this negated the theory of Suresh et al,[3] that infection causing necrosis and perforation of diaphragm leading to herniation. At the same time the smooth edges favored the hypothesis of Ashcraft et al[4] and Bangale and Watters[5] that GBS infection was secondary to the congenital diaphragmatic hernia.

In conclusion, any neonate with group B streptococcal pneumonia, who has been treated adequately and is doing fine and then suddenly deteriorates, they should be investigated for presence of congenital diaphragmatic hernia.

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