Late-presenting right congenital diaphragmatic hernia

Sir,
Congenital diaphragmatic hernia (CDH) is classically left-sided, only 8-15% reported to be right-sided.[1,2] In the present era, a large majority of the CDH are diagnosed prenatally and the rest of these present with distress at birth or within the first few hours of life. Only a small number of cases are diagnosed at an age after than one month—so called “late-presenting CDH”.[3] A one month-old, full-term, male infant presented with respiratory difficulty and failure to gain weight. The patient was born by normal vaginal delivery and no antenatal ultrasonography had been done.

The parents had noticed fast breathing and difficulty in feeding right from the first week of life. The respiratory distress increased especially after feeds. On examination, the child was found to be malnourished and tachypneic with a respiratory rate of 60 breaths per minute with mild intercostal retractions. The abdomen was scaphoid and there was decreased air entry on the right side. Chest radiographs showed intestinal loops lying in the right thorax with a mediastinal shift [Figure 1]. The child was then taken up for laparotomy through a right subcostal incision. Small bowel loops were seen to be herniating into the right chest cavity from behind the liver. Bowel loops were reposited back into the abdomen. The postero-lateral defect was well visualized. A double breasting repair of the diaphragm was done and an intercostal drain was inserted in the right chest.

Postoperatively, the child did not require any ventilatory support. The chest tube was removed on day 2 and a chest X-ray showed normal contours of the right diaphragm and a well-expanded right lung [Figure 2]. The child recovered uneventfully. The postnatal problems in diaphragmatic hernia relate to the effects of the herniated viscera on the developing heart and lungs. The respiratory distress associated with CDH results from a combination of lung hypoplasia and pulmonary hypertension. The most severely affected infants develop respiratory distress at birth. Whereas a large majority demonstrate respiratory symptoms within 24 hours of birth, only 2.6-10% of the cases may present after this period.[3-4] It has been seen that the proportion of right-sided CDH in late-presenting cases is higher.[4]

In right CDH, several clinical diagnostic pitfalls are possible. In a retrospective review of patients with right CDH, Daher et al. reported the mean age at diagnosis to be six months.[5] The delay between the first symptom and diagnosis ranged between 0 and 10.5 months (mean = 4.5 months).[5] A study of late-presenting CDHs was done using a large multicentric database in Tokyo by Kitano et al.[4] Out of 3098 cases, 2.6% (79 cases) met the inclusion criteria of late-presenting CDH. They reported that the presenting symptoms of late-presenting CDH can be respiratory or gastrointestinal but left-sided lesions more commonly presented with gastrointestinal problems and right-
sided defects with respiratory complaints.\textsuperscript{[4]} Kaur et al. have reported a nine-month old infant with a right CDH masquerading as staphylococcal pneumonia.\textsuperscript{[6]} The chest X-ray showed multiple lucent shadows in the right lower zone with pleural effusion.

A late-presenting CDH should always be strongly suspected in such cases so as to avoid inadvertent placement of a chest drain in such patients. Right-sided CDH has been reported to be associated with poorer outcome and higher incidence of complications.\textsuperscript{[5]} The present case was diagnosed to have diaphragmatic hernia at one month of age when he was investigated for respiratory distress which had its onset in the first few days of life. The distress was mild so the parents did not seek any treatment till one month of age when they noticed that the child had not gained any weight. This is unlike the classic presentation of CDH where distress occurs at birth. There were no gastrointestinal symptoms in the present case. Our case illustrates the importance of considering a late-presenting CDH as an important differential diagnosis in such scenarios.

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**REFERENCES**


Gitanjali P. Mansukhani, N. N. Kadam, D. B. Bhusare*, A. K. Singal*
Departments of Pediatrics and *Pediatric Surgery, MGM Medical College, Navi Mumbai, Maharashtra, India. E-mail: arbinders@yahoo.com