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Case Report

Atypical presentation of congenital diaphragmatic hernia



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Introduction

Congenital diaphragmatic hernia (CDH) usually presents as respiratory distress in the newborn period but atypical presentation can mimic a vast array of clinical symptoms frequently masquerading other more common paediatric entities. Prompt and accurate diagnosis is essential in the management of CDH.¹

Case report

Seven days old female neonate was brought by the mother with history of refusal of feeds and having not passed urine for

over 24 h. She was born full term normal delivery in a hospital with birth weight of 2.5 kg and discharged home on day 3 of life following normal perinatal transition. On examination the baby was lethargic with doughy feel of the skin and had petechial rash over both feet. She had lost 700 gm of weight since birth (28%). Anterior fontanel was tense with normal neonatal reflexes. There was no breathing difficulty or any sign of respiratory distress at presentation. Abdomen was soft with no significant organomegaly. Bowel sounds were heard and the baby had passed meconium normally. She was noted to have subtle seizures during examination and was managed as a case of Late Onset Sepsis with hypernatraemic dehydration.

Investigation revealed total leukocyte count of $14 \times 10^3/L$ with polymorphs of 70%, platelets $97 \times 10^3/L$ and shift to left with 25% of band forms on peripheral blood smear. Renal functions were deranged (urea 376 mg/dL, creatinine 6.1 mg/dL) with dyselectrolytaemia (serum sodium 186 mEq/L and potassium 8.8 mEq/L). CSF analysis was suggestive of meningitis (WBC $730 \times 10^3/L$ with predominant neutrophils, protein 123 mg/dL, sugar 187 mg/dL, globulin increased and Gram and Zn strain—Negative). Venous blood gas revealed pH 7.25, Na 164 mEq/L and HCO₃ 14.3 mEq/L. Blood sugar was 480 mg/dL. USG abdomen revealed evidence of medullary hyperechogenicity in both kidneys however transcranium USG was normal. Chest skiagram showed homogenous opacity in left lower zone (Fig. 1).

The dehydration and dyselectrolytaemia were corrected slowly over 3 days and euglycaemia achieved with insulin infusion (Table 1). Renal functions showed an improving

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Fig. 1 — Chest radiograph AP view showing homogeneous opacity left lower zone with no clear bowel shadows in the chest making diagnosis of CDH difficult.

trend and repeat USG abdomen showed no evidence of calcinosis.

Breast feeds reinitiated on day 3 of admission but baby had deterioration with respiratory distress. On auscultation there was decreased air entry on left side with crackles not varying with respiration. Baby desaturated further with fall in saturation to 60%, was resuscitated and put on ventilator.

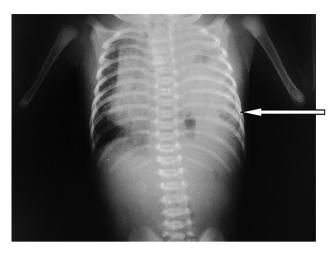


Fig. 2 – Chest radiograph showing shift of mediastinum to right and homogenous opacity left hemithorax with gas shadow in lower zone (arrow).

Repeat chest skiagram revealed mediastinal shift to right and homogenous opacity left hemithorax with gas shadows in lower zone suggestive of diaphragmatic hernia (Fig. 2). However, presence of a nasogastric tube and the stomach gas shadow in the abdominal cavity posed a diagnostic dilemma (Fig. 3). Antibiotics were upgraded in view of consolidation. Urgent computed tomography revealed intestinal loops in left hemithorax (Fig. 4). After stabilisation the baby was taken up for surgery.

Intraoperatively there was a defect of 4×6 cm in the posterior part of the diaphragm with herniation of small intestine, ascending colon, part of transverse colon and spleen. The stomach was in situ. There was associated malrotation of gut which was corrected. Baby was weaned off from the ventilator after 2 days and made an uneventful recovery.

Discussion

CDH occurs in about 1 in 3000 live birth. The most common defect is the posterolateral (Bochdalek) type and 80% of it is left sided. Over 90% of the patients will be diagnosed either antenatally or will present with respiratory distress in the first few hours of life requiring mechanical ventilation. However, about 5%–30% of diaphragmatic hernias present beyond the

Table 1 $-$ Serial biochemical parameters in the patient with CDH.							
	Day-1	After 6 h	Day-2	Day-3	Day-4	Day-5	Day-10
Serum Na ⁺ mEq/L	186	176	164	160	148	145	137
Serum K ⁺	8.6	6.6	5.5	4.5	5.3	4.4	4.8
Serum urea mg/dl	376	_	348	240	119	18	10
Serum creatinine mg/dL	6.1	_	4.9	3.8	1.1	0.5	0.5
Fluid for correction	NS bolus 20 ml/kg	N/2	N/2	N/2	N/3	N/5	N/5

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