

normal in the patient as well as in the parents and sibling Fourthly, there was no family history suggestive of methemoglobinemia and methemoglobin levels of both the parents and elder sibling were normal Fifthly, There is transient physiologic deficiency of cytochrome b5 reductive activity in neonates approximately 60% of the normal adult value, and even greater reduction in premature neonates is known making them more susceptible to oxidant stress leading to methemoglobinemia(3,4) Sixthly, if we consider the possibility of neonate having homozygous cytochrome b5 reductase deficiency and both parents heterozygotes having normal levels of methemoglobin, the mother, if she was heterozygote could have manifested with methemoglobinemia to strong oxidant stress of Dapsone therapy(3) Finally though we were unable to estimate cytochrome b5 reductase enzymatic activity, the prompt response to single dose of methylene blue and non recurrence of methemoglobinemia in five months of follow up in the patient subsequently, probably excludes enzyme deficiency as a cause of methemoglobinemia(3)

Esophageal Atresia with Right Pulmonary Agenesis

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The association of pulmonary agenesis with esophageal atresia with or without tracheo-esophageal fistula is extremely rare. This combination is often fatal as there is progressive respiratory embarrassment

We recommend that all the neonates born to mothers who receive potential oxidant drugs like Dapsone just before the time of delivery should be carefully monitored for methemoglobinemia

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which complicates anesthesia. Only seven such cases have been reported so far(1-4).

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We are reporting a similar case because of its rarity and as there was complete esophageal atresia without tracheo-esophageal fistula along with right pulmonary agenesis.

Case Report

A 25-year-old unbooked second gravida was admitted in labor with massive hydramnios. An artificial rupture of membrane was done and five litres of liquor was drained. She delivered a preterm female baby weighing 1.5 kg with mild respiratory distress.

Physical examination showed moderate chest retraction, absent breath sounds on the right side and apex beat on the right side. Nasogastric tube was difficult to negotiate in the stomach. X-ray of the chest and abdomen revealed blind proximal esophageal pouch, opacification of the right hemithorax with shift of the mediastinum to right (Fig. 1). There was no gas in the abdominal cavity.

The severity of the condition was explained to the parents, and as they were not ready for operation, the baby was treated conservatively. The baby expired on the fifth day.

Discussion

Esophageal atresia occurs in 1:3000-4500 live births and about one third are born prematurely. However, its association with pulmonary atresia especially of severe forms is exceedingly rare. Fifty per cent of babies with atresia have associated malformations, which include cardiovascular, skeletal, renal, anorectal and urogenital malformations, collectively known as Vater syndrome(5-7). There is no predilection for sex or the side of agenesis/aplasia of lung. Bilateral pulmonary agenesis has been reported in association only twice(2,3).

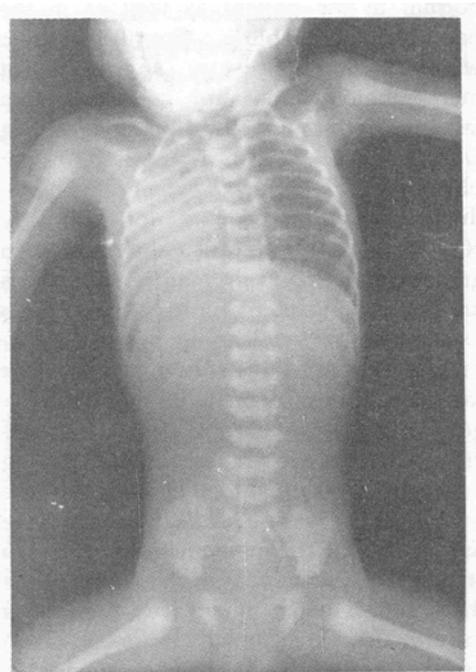


Fig. 1. Chest radiograph of the neonate showing opacification of right hemithorax and no gas shadow in the abdomen.

This combination is often fatal because of the progressing respiratory embarrassment with already marked pulmonary compromise of pulmonary reserve. All such babies require prolonged course of intubation and ventilation followed by corrective surgery which is possible only in tertiary care hospitals.

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Paravertebral Extraosseous Ewing's Sarcoma

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Extra Osseous Ewing's Sarcoma (EES) has been considered a distinct clinicopathological entity despite its striking ultrastructural similarity to Ewing's Sarcoma of Bone (ESB) and same translocation involving band q12 of chromosome 22(1,2) Today more than 150 cases of EES in different locations have been described, for example, larynx, scalp, nasal fossa, neck, chest wall, lung, perineum, finger, arm, lip and toe(3) Paravertebral location of EES

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have been extremely rare To the best of our knowledge EES located in paravertebral area has not been reported in Indian literature We report one such case

Case Report

A 12-year-old male child was admitted with the complaints of progressively increasing dull aching pain in the upper back and right shoulder, radiating to the right hand since last two months and progressive weakness of his right upper limb since last one month Around the same time he noticed a painless swelling in the right side of neck which was slowly increasing in size There was history of loss of appetite and weight, however, he did not suffer from headache, vomiting, visual disturbance, dysphagia, dyspnea or disturbance of gait He was a thin built pale child Examination of central nervous system revealed weakness (power 3/5) and hypotonia of right upper limb The biceps, the triceps, and supinator reflexes in right upper limb were elicitable with difficulty The thenar and hypothenar eminences were significantly wasted and there was no sensory loss Rest of the systemic examination did not reveal any abnormality There was