An unusual presentation of congenital bronchoesophageal fistula

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Summary

We present the case of a 5-week-old neonate with multiple congenital abnormalities including a broncho-oesophageal fistula, which showed radiological features suggestive of congenital diaphragmatic hernia. Emergency limited barium swallow done was initially reported as a case of diaphragmatic hernia. Autopsy revealed pus within the right lung, and a fistulous connection between the oesophagus and an intralobar sequestrated lung. No diaphragmatic hernia or intra-abdominal organ abnormality were seen, and an occipital meningomyelocoele was also confirmed.

Keywords: ***Bronchoesophageal fistula, Hiatus hernia, Meningo-encephalocoele***

Résumé

Nous présentons le cas d'une néonatale agée du cinq semaines atteinte d'une anormalie congénitale multiple y compris une fistule broncho-oesophageal qui avait indiqué des traits radiologique evocateur d'une hernie diaphragmatique congénitale. La méthode de la bouillie de sulfate de barium restreintée d'urgence effectue etait au départ donné comme un cas de l'hernie diaphragmatique. L'autopsie avait indiqué un pus du dedans du poumon du côoté droite et un lien fistule entre l'oesophage et le poumon intralobe séquestré. On n'a pas vu aucune hernie diaphragmatique ou organe intraabdominal anormal, et on a également confirmé un méningomyelocoele occipital.

Introduction

Congenital Broncho-oesophageal fistula (BEF) is a rare abnormal connection between the Gastrointestinal system and Respiratory system. The most frequent site of communication is between the oesophagus and the lower lobe of the right lung especially the superior segment but may occur high up in the neck. Initial diagnosis rests mainly on symptomss and plain radiographs, and other confirmatory examinations include, Barium swallow, Barium oesophagogram, bronchogram and cross sectional imaging (CT and MRI).

Case history

A 5-week-old female infant born at a private hospital at term by emergency caesarean section was noticed to have a soft occipital swelling.

The only significant antenatal history was that the mother received therapy for malaria at about the third week of the pregnancy. The baby was referred to the hospital shortly after birth but did not present till about the fifth week when she was noticed to have difficulty in breathing.

The physical examination revealed an acutely ill-looking infant, small for her age, pale, febrile and in severe respiratory distress. She had resonant percussion notes over the right hemithorax. The cardiovascular system was essentially normal. Ultrasonography of the occipital swelling revealed a meningo-encephalocoele, while that of the abdomen was essentially normal. Chest X-rays (Fig. 1_a and 1_b) showed an area of increased lucency in the lower zone of the right lung field laterally bordered medially by an opacity, which also obliterated the right cardiac margin.There was a subtle shift of the mediastinum to the contra-lateral side. There was

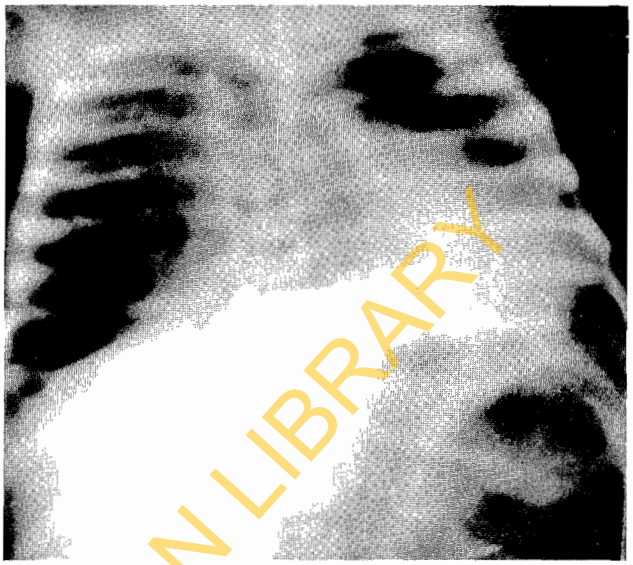

Fig. Ia *Showing the area of lucency to be in the posterior mediastinum and appears to be in continuity with the bowel gas shadow.*

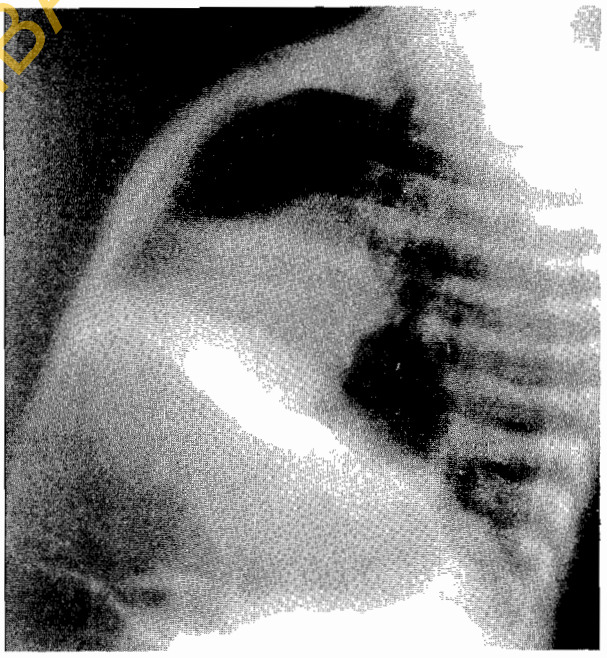

Fig. Ib *Showing the area of lucency to be in the posterior mediastinum and appears to be in continuity with the bowel gas shadow.*

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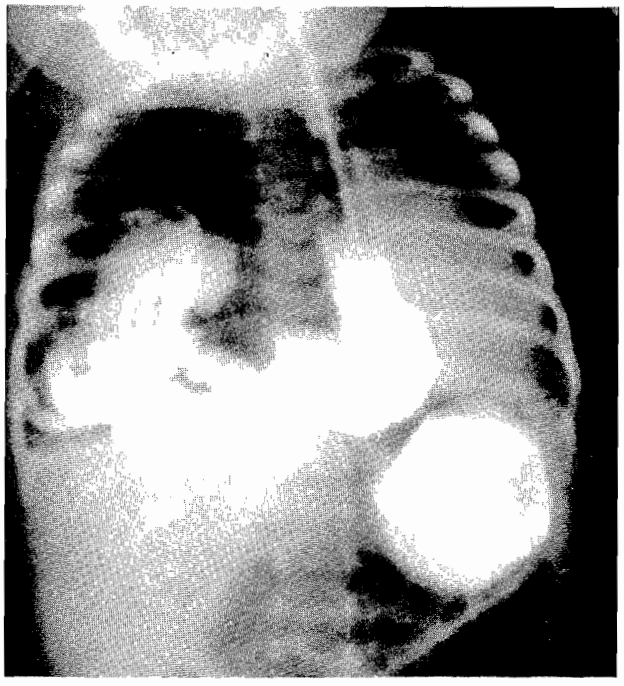

Fig. 2 *Limited barium swallow showing localised contrast with lobulated outline in the right hemithorax*

Fig. 3 *Delayed film showing the contrast to have layered at the base of the lung*

gaseous distension of the visualised bowel loops. The lateral film showed the area of lucency to be in the posterior mediastinum, which appears to be in continuity with the gastric air bubble. Differential diagnoses included congenital hiatus hernia, pneumatocoele, and sequestrated lung. A limited barium swallow (Fig. 2) showed an area of localised contrast in the right hemithorax with lobulated outline and a stalk like connection between the distended lower third of the oesophagus. Barium was also seen in the fundus of the stomach. The delayed film (Fig. 3) showed the contrast to have layered evenly at the base of the right lung. The patient however died before computerised tomogram could be done.

At post mortem the right lung was found to contain thick pus with no evidence of gut herniation or defect in the diaphragm. A fistulous connection between the oesophagus and an intralobar sequestrated lung was documented. The meningo-myelocoele was also confirmed, and no intra-abdominal organ abnormality was seen.

Discussion

Congenital Broncho-oesophageal fistula (BEF) is a rare abnormal connection between the Gastrointestinal system and Respiratory system.[1,2,3,4] It has a female preponderance and more common on the right field. Seventy-five percent of cases are not diagnosed until adult life.[1,2,3] Fistula between the bronchus/trachea and the oesophagus results from failed tracheo-oesophagus separation in the early stage of embryonic development.[3] A fistula may not cause symptoms in childhood until adult life.[3] Bainbridge and Keith have classified BEF into four types.[3] Type I is a fistulous connection to a wide neck diverticulum of the oesophagus, and type II is a short tract running obliquely from oesophagus to the bronchus. Type III is a connection between the oesophagus and a cyst in a lobe, while type IV, is a fistulous connection to a sequestrated segment of the lung. Type II is the simplest and the most common and carries the best prognosis.

Initial diagnosis rests mainly on symptoms and plain radiographs. Also the need and type of further investigation is governed by the most likely diagnosis on assessment of age of the patient, symptoms and initial radiographs.[1,2,3] The case here presented is of type IV, confirmed by the findings at autopsy.

Chest radiographs, will always reveal an abnormality, which may be subtle, but can rarely be normal. There is usually pneumonic changes due to leakage from the oesophagus. Gaseous distension of the bowel in the abdomen is prominent confirming a fistulous connection between the respiratory tract and the gastrointestinal tract.

Contrast swallow should be performed using diluted barium to look for the fistulous tract (which is usually difficult to demonstrate),[7] but this can be enhanced by good oesophageal distention. The most frequent site of communication is between the oesophagus and the lower lobe of the right lung especially the superior segment but may occur high up in the neck.[2,7] There are two main reasons why these lesions may be missed; the tract may be very narrow and have muscular walls, which may close the fistula, or the lesion may occur high in the neck, with examinations failing to include the area of involvement.[7]

Barium oesophagogram is considered the most sensitive and definite tool for diagnosis especially when recorded for detailed study.[3] A bronchoscopy and or bronchogram may be necessary especially in small children.

Cross sectional imaging (CT and MRI) will always show the abnormality and this should be the routine in centres in developing world where these facilities are available.[5,8] It should be noted that sometimes, despite extensive investigations, the dignosis is made only during operation when a

more common disease, such as bronchogenic or oesophageal cyst, is suspected preoperatively.[3] Since BEF is often associated with congenital abnormalities in other systems it is pertinent to do a thorough examination to rule out these other associated abnormalities.

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