Synchronous Gut Duplication Cysts : A Rare Presentation

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Introduction

Duplications of alimentary tract are rare congenital anomalies and multiple duplications occur in a fraction of these cases [1]. Their presentation is diverse and they pose a diagnostic and therapeutic challenge [2]. Majority occur in the small intestine and present in the first year of life [1]. Thoracic duplications usually present with recurrent respiratory tract symptoms or these are detected incidentally on chest radiographs. Pyloric and duodenal duplications present with recurrent vomiting and failure to thrive [1]. The small intestine duplication presents with abdominal mass, intestinal obstruction or gastrointestinal bleeding [1]. The duplication cysts contain gastric mucosa in 50% of cases resulting in ulceration, hemoptysis or hematemesis from perforation [3,4]. The exact site of bleeding is difficult to determine in neonates. We report an unusual presentation of synchronous gastrointestinal duplication.

Case Report

One year old child was detected to have an opacity in the right chest on radiograph taken for anaemia and febrile convulsions. History revealed an episode of lower gastrointestinal bleed two months back, which required blood transfusion without further investigation. The child was pale and underweight. The clinical examination of abdomen and chest were essentially normal. The haemoglobin was 5.4 g% with a reticulocyte count of 2.8%. Peripheral smear showed microcytic hypochromic anaemia. Ultrasound abdomen was normal. Contrast enhanced computed tomography (CECT) revealed a large well circumscribed unilocular cyst in the right paravertebral space (Fig. 1). Right thoracotomy and excision of a large cystic mass was done. The lesion was 10x5x5 cm in size, extending from level of carina to the diaphragm. Histopathology revealed bronchogenic and ectopic gastric mucosa. The patient made uneventful recovery and was discharged.

The patient presented after six weeks with severe hematochezia. After resuscitation and haemodynamic

stabilisation, a technetium scan revealed intense abnormal activity in the right iliac fossa (Fig. 2). Exploratory laparotomy showed a tubular duplication of the ileum, 75 cm long with proximal blind and distal communicating end (Fig. 3). There was an associated malrotation of the midgut. The involved segment was excised and malrotation corrected. The remaining bowel was measured to be of adequate length (160 cm). The patient made an uneventful postoperative recovery.

Discussion

The incidence of intestinal duplication in western literature is one of every 4500 autopsies. No familial incidence has been reported. Males dominate most series. Duplications can occur anywhere from mouth to anus. Sixty seven percent of them present by the first year of life but it has been reported in neonates also [5]. Fifteen percent of the patients have two or more duplication cysts [1]. We report a case of synchronous thoracic oesophageal and ileal duplications.

Fig. 1 : Contrast enhanced CT revealing a large well circumscribed unilocular cyst in the right paravertebral space

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Fig. 2 : Technetium scan revealing intense abnormal activity in the right iliac fossa

Duplications are either cystic or tubular in shape. The thoracic oesophageal duplication in our case was cystic and the small intestinal duplication was tubular. The diagnostic characteristics of duplication cysts include the presence of a well developed smooth muscle coat, the lining epithelium resembling some portion of the alimentary tract and intimate attachment with the adjoining gut [6]. In present case the thoracic duplication had epithelium resembling gastric, oesophageal and bronchogenic mucosa, while the small intestinal duplication mucosa resembled the ileum with islands of gastric mucosa. In literature the mediastinal foregut duplication cysts are reported having neither intraspinal extension nor heterotopic gastric mucosa [7].

The thoracic cyst may be associated with incomplete vertebral fusion and may have an attachment to the dura or spinal cord. It rarely communicates with the oesophageal lumen. The thoracic duplication in our patient did not communicate with either the spinal canal or the oesophagus. Most thoracic cysts remain intrathoracic, but sub-diaphragmatic extensions are known. Holcomb [8], cites 2% of all duplication cysts as being combined thoracoabdominal in nature. Young patients with thoracic cyst present with respiratory symptoms. Malena from the ectopic gastric mucosa is rare and is the usual presentation in older patients. Our patient though young presented with malena. The respiratory symptoms are caused by expansion of the cyst due to collection of secretions. These mediastinal cysts have to be differentiated from neuroblastoma, haemangioma and bronchogenic cysts.

Surgical excision is the only option after diagnosis, even if the cyst is asymptomatic. The complications of compression, bleeding, perforation, infection or malignancy are well known [9]. With the midgut duplication cyst, excision is the best choice. Small cysts can be excised with the normal intestine. If a large length of the normal intestine is involved, excision would lead to malabsorption and short gut syndrome requiring

Fig. 3 : Tubular duplication of the ileum, 75 cm long with proximal blind and distal communicating end.

alternative procedures to retain normal intestine.

Our patient had a foregut duplication cyst in combination with a midgut tubular duplication and presented with recurring malena. This patient had ectopic gastric mucosa in both the locations. We recommend that routine technetium scan should be done in all cases of thoracic duplication cysts in the followup period to detect concomitant midgut duplication.

Conflicts of Interest

None identified

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