Mediastinal Enteric Cyst in a Neonate

Vikram Singhal, Rathika D. Shenoy[1], Nutan Kamath, Sadashiva Rao[2]
Departments of Pediatrics and [2]Pediatric Surgery, Kasturba Medical College, Manipal University, [1]Department of Pediatrics, KS Hegde Medical Academy, Mangalore, Karnataka, India

ABSTRACT

Mediastinal enteric cysts are relatively uncommon, and patients tend to present at a later age compared to those with duplications in other areas of alimentary canal. The tendency of enteric cyst to enlarge and produce airway obstruction is sufficient reason for early surgical removal. We report on a case of mediastinal enteric cyst in a neonate with respiratory distress for its early presentation and management. The embryological basis and anatomical issues relating to duplication cysts of the gastrointestinal tract is discussed.

Key words:
Duplication cyst, respiratory distress, surgical emergency

INTRODUCTION

Surgical emergencies presenting as respiratory distress in the late neonatal period are uncommon. Enteric cysts (or enterogenous cysts, EC) are rare congenital anomalies of the developing foregut and include esophageal duplication cysts and neurenteric cysts. These are often discovered incidentally on X-ray chest in an older child. We report on a case of mediastinal EC in a neonate presenting with respiratory distress for its rarity and issues in diagnosis and management.

CASE REPORT

A 22- day-neonate, first born of a nonconsanguineous parentage, was referred with respiratory distress of 1-day duration. Pregnancy and delivery was uncomplicated; he was born term with a birth weight of 2800 g, length of 54 cms and head circumference of 36 cms. At admission he had poor activity; the respiratory rate was 70 per minute with intercostal retractions; the pulse rate was 160 beats per minute; peripheral pulses were equally palpable and synchronous with normal heart sounds. Chest examination revealed decreased air entry and impaired breath sounds on right side. Abdominal and genital examination was normal and nasogastric tube could easily pass to the stomach. A homogenous opacity in right middle and lower zone with deviation of esophagus to left was seen on X-ray. No vertebral anomalies were noted. Contrast-enhanced computerized tomography confirmed a posterior mediastinal cystic mass [Figure 1]. Hematological profile and serum chemistry results were within normal limits. Our clinical diagnosis included bronchogenic cyst, cysticadenomatoid malformation and neurenteric cyst. The patient underwent a right posterolateral thoracotomy. A large cystic mass (5 × 4 cm) in the posterior mediastinum extending on to the right chest, with a good line of cleavage between the mass and the esophagus was observed. The loculated cyst was aspirated, decompressed and excised. In the postoperative period, neonate required mechanical ventilation. Histopathological examination of the tissue confirmed an EC [Figure 2]. Child is doing well at follow-up.

DISCUSSION

Alimentary tract duplications may be found anywhere from mouth to anus. In a large collective review of the literature of 495 duplications, 50% were midgut, 36% foregut and 12% hindgut. Esophageal duplications formed 19%, and of the midgut duplications, ileal duplications were the most common, forming 35%.[1]

The primitive foregut gives rise to the pharynx and lower respiratory tract as well as upper gastrointestinal tract. The most common foregut cysts, the bronchogenic cyst represents abnormal budding of vertebral primitive foregut whereas the esophageal duplication represents abnormal budding of the dorsal primitive foregut, indicating their common origin from the primitive foregut and close

Address for correspondence:
Dr. Nutan Kamath,
Department of Pediatrics, KMC Hospital, Attavar,
Mangalore – 575001, Karnataka, India.
E-mail: nutankamath@yahoo.com

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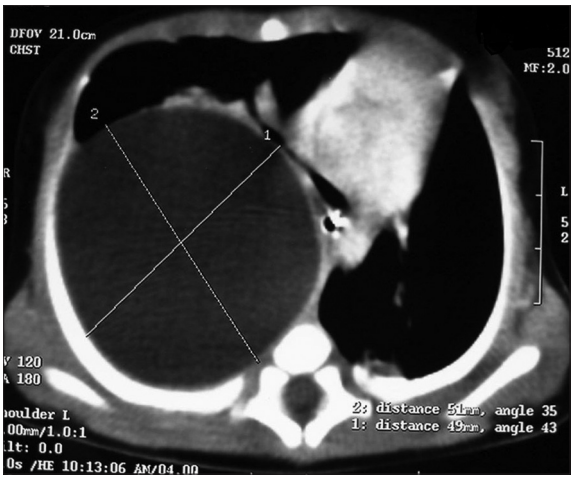

Figure 1: Computerized tomography showed posterior mediastinal cystic mass 5.1×4.9 cm

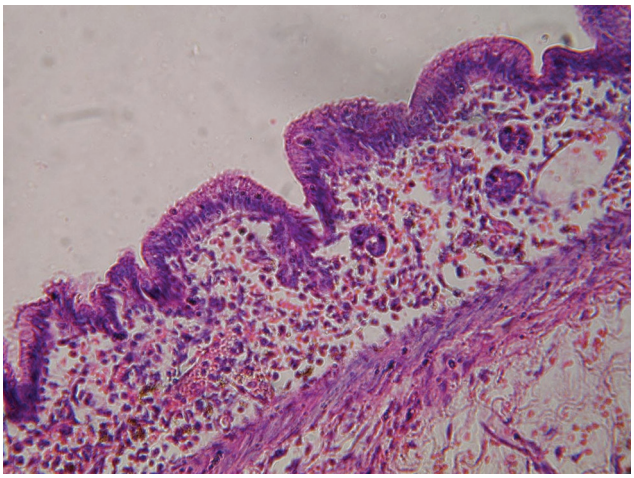

Figure 2: Histopathological examination of the tissue revealed enteric cyst

embryologic relationship.[2] The earliest description of foregut duplication was that of Blassius and Bremer in 1711.[3] The most accepted theory for EC is that of the 'split notochord syndrome' postulated by Bentley and Smith with persistence of a neuroenteric canal.[4] During the third week of intrauterine life, the developing notochord fuses with the embryonic endoderm and at about the fourth week the endoderm separates from the notochord, but if adhesions or a neuroenteric band persists during this separation, traction diverticula and duplication cysts will develop, as well as vertebral defects. This can also explain the frequent association of foregut duplication with malformations of the spinal column and cord. When foregut duplication cysts are associated with vertebral malformations, they are called neuroenteric cysts.[5]

EC are usually separate from the esophagus and lie in the posterior mediastinum. Mediastinal EC are included within esophageal duplication cysts because of its location. They usually develop in the right posterior mediastinum in retrocardiac position and often extend into the right hemithorax.[6] Clinical presentation may include respiratory distress due to mass effect, as in our patient who developed distress in the late neonatal period. Mediastinal EC lumens are partially or completely lined by gastric or intestinal mucosa surrounded by smooth muscle. They arise from the failure of coalescence of vacuoles early in the development of the foregut.[6]

Esophageal duplications are relatively uncommon, and patients tend to present at a later age compared to those with duplications in other areas of alimentary canal. Although duplications of the esophagus are reported throughout its length, most are located in its distal half. These lesions are cystic and located in the posterior mediastinum (commonly on the right side). Duplications can present with wide spectrum of symptoms based on location and type. Dyspnea, stridor or persistent cough are the commonest presentations.[5] Respiratory distress occurs most commonly in small infants because of pressure on bronchi or lung.[6] Hemorrhage is seen in those with heterotopic gastric mucosa, which is present in 29-35% of duplications, which can be identified preoperatively by technetium scintigraphy.[6,7] CT scan is a routine part of diagnostic evaluation of mediastinal tumors, cysts, and other masses. With the liberal use of antenatal scans, these cysts can now be demonstrated prenatally,[8] and since a large number of these patients will have respiratory distress immediately after birth, arrangements can be made for them to be delivered in a tertiary care center where further postnatal evaluation and management with a competent neonatal and pediatric surgery team can be provided. Chest and epigastric pain or recurrent respiratory tract infection are the modes of presentation in older children. The foregut cysts may remain asymptomatic for months or years.[9]

The tendency of EC to enlarge and produce airway obstruction is a sufficient reason for early surgical removal. Enteric variety has variable types of epithelium and well-developed muscular wall, which resembles that of the intestine. Ectopic mucosa if gastric is liable to perforate and bleed. Cartilage is never found in the wall. All the characteristic histological features were present in our patient. Surgical excision of the cyst is the mainstay of treatment along with comprehensive supportive care. To the best of our knowledge there have been only few case reports from India.[10]

CONCLUSIONS

It is important to consider EC as a differential diagnosis in a neonate presenting with respiratory distress. Treatment is surgical removal. Outcome of an EC without associated congenital anomalies is favorable.

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