Case Report

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Congenital Esophageal Duplication Cyst-A Rare Case Report and Review of Literature

Pore Anant[1], Sarwal Ankush[1*], Barsiwala Nargish[2] and Gheewala Hussain[3]

[1]*Tata Medical Centre, Kolkata, India*

[2]*Children`s Hospital, Mumbai, India*

[3]*Saifee Hospital, Mumbai, India*

[*]Corresponding author
Dr. Ankush Sarwal, Fellow, Department of Surgical Oncology, Tata Medical Centre, Kolkata, India, E-mail: drankushsarwal@yahoo.in

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Abstract

Alimentary tract duplication is a relatively rare congenital anomaly. Esophageal duplication cyst is an uncommon developmental anomaly among them. Respiratory symptoms are the most common symptoms in foregut duplication, most case present with respiratory distress which may be present from birth, or symptoms may insidious with cough, wheeze, or recurrent respiratory infection. We present a case of newly born baby with respiratory distress, which on radiological investigation showed left mediastinal mass that was removed by excisional biopsy and final histopathology revealed an esophageal cyst.

Keywords: esophageal duplication cyst, foregut duplication cyst, congenital malformation

Introduction

Gastrointestinal tract duplication cysts are rare congenital malformation. Foregut duplication cyst are categorised on the basis of their origin into esophageal, bronchogenic, neuroenteric cyst [1]. Bronchogenic and esophageal duplication cyst are thought to arise from abnormal budding of the embryonic foregut at 5-8th week of gestation, although the exact embryonic origin of different types of duplication cyst remains mystery [2]. Among the foregut duplication cyst, 50-70% is nitrogenous, while 7-15% of them are bronchogenic [3]. Foregut duplication cyst constitutes 6-15% of primary mediastina masses [4]. They may be contained within the gastrointestinal tract wall or can be extrinsic to it. The reported prevalence of esophageal duplication cyst is 0.0122 %. Around 80% of the lesions are diagnosed in childhood, with majority of them being symptomatic [3]. Most of the esophageal duplication cysts are located in right posterior inferior mediastinum. Two-thirds of this lesion is found in the lower third of the esophagus and one-third of the upper esophagus [5]. Esophageal duplication cyst have a double layer of surrounding smooth muscle, are lined by alimentary (squamous or enteric) epithelium, and are either attached to esophagus in para esophageal or intraluminal fashion [3,4,5]. Patients of esophageal duplication cyst are asymptomatic but can develop symptoms such as dysphagia or chest pain due to compression of surrounding structure. Middle and lower esophageal cyst can cause dysphagia, epigastria discomfort, chest pain, vomiting. Upper esophageal cyst can cause stridor and non productive cough. Rare symptoms such as cardiac arrhythmia, retrosternal and thoracic back pain, cyst ulceration, bleeding or cyst rupture with secondary mediastinitis have also been reported. . Surgical removal is not difficult and is needed to provide strong clinical evidence for diagnosis. Complete surgical excision is typically curative because recurrence is rare [6-8].

Case Presentation

A full term male child was delivered by Lower segment Caesarean section (LSCS) in a female with bad obstetrics history. The mother was 34 year old Gravida 5 and para 3 female who previously had 2 miscarriages in Gravida 1 and 3 with 2 live births. In current pregnancy patient was on Insulin for her Gestational diabetes. Antenatal USG diagnosed cystic adenoid or congenital diaphragmatic hernia. The birth weight was 3.32 kg; baby cried normally at birth, no resuscitation was required. The anthropometric parameter of the baby were length 49 cms, head circumference 33 cms, chest circumference 32 cms respectively The infant started having respiratory distress immediately after crying. Post LSCS baby was shifted to Neonatal Intensive Care Unit (NICU) for further management. In NICU, chest x ray was done which showed left hemi thorax opacity suggesting a mediastinal mass as shown in Figure 1.

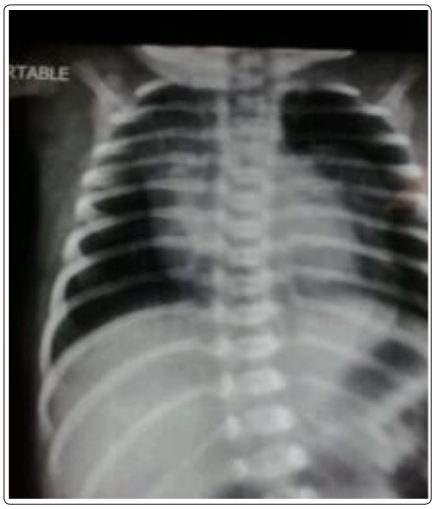

Figure 1: Chest X ray: Showing Left mediastinal mass

Computed tomography scan done subsequently showed a large well defined cystic lesion in the para-mediastinal aspect located posteriorly in the left hemi thorax with minimal peripheral enhancement suggesting it to be an esophageal duplication cyst. Blood parameters were normal. Ultrasonography of brain was normal. Infant was on full oral feed during this period. Thoracotomy and excisional biopsy was planned in order to prevent the complications of cyst and confirm the diagnosis. Intraoperative, a large cystic lesion of size around 3.5x 2 cms was seen in posterior mediastinum, parallel to esophagus. There was no visible communication seen to the esophagus and aberrant vessels. Around 30 ml of mucinous clear fluid was aspirated from the cyst. The cyst was excised and sent for Histopathological examination as shown in figure 2. Histopathology of the specimen confirmed it to a benign gastroenteric cyst with gastrointestinal-type mucosa with well-developed muscular is propria with no evidence of dysplasia. The infant had an uneventful recovery postoperatively and was discharged on 7th post operative day. The weight at time of discharge was 3.4 kg and tolerating full oral feeds.

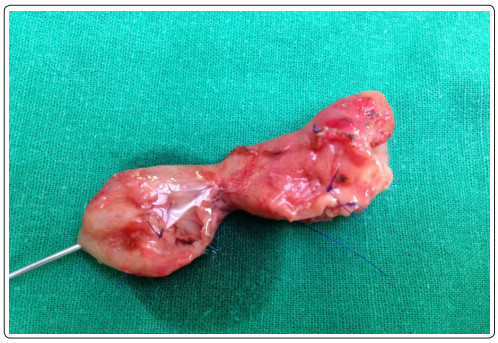

Figure 2: esophageal duplication cyst

Discussion

Embryo logically, the foregut at the cranial end of the primitive gut develops into the pharynx, respiratory tract, esophagus, stomach, and the first part and proximal half of the second part of the duodenum [1]. W. E. Ladd first introduced the term duplication in 1934.Esophageal duplication cysts are exceedingly rare congenital embryonal malformations with estimated prevalence of 0.0122 % in literature [3]. Majority of them are found in the mediastinum where they present either as separate mass along or in continuity with native esophagus. Even though duplications cysts are mostly benign, but presence of ectopic gastric mucosa and the potential for malignant degeneration remains a concern. The symptoms are related to size, location, type of duplication, and presence of heterotopic mucosa. In our case the infant was symptomatic for respiratory distress and doubt was raised since the time antenatal Ultrasound was done. Esophageal duplication cyst may remain undetected on plain chest radiograph. Noncontract and contrast-enhanced CT scan can demonstrate relation of the duplication cyst with the esophagus and tracheobronchial tree as was done in the present case. The mainstay of treatment is surgical excision, which can be accomplished by means of a thoracotomy or thoracoscopy [7]. We recommend complete excision to confirm the diagnosis, relieve symptoms, and prevent complications.

Conclusion

Esophageal duplication cysts are relatively rare and may have no specific symptoms. Surgical removal or enucleation is the treatment of choice in most symptomatic cases. In asymptomatic cases, surgery should be considered as the cyst could develop ulceration or perforation and there is risk for malignant transformation too. Curative surgery is not difficult and is needed to provide strong clinical evidence for diagnosis. Complete surgical excision is typically curative because recurrence is rare.

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