ESOPHAGEAL DUPLICATION CYST IN ADULT WITH SUSPECTED ISOLATED CROHN'S ESOPHAGUS: A RARE CASE REPORT

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ABSTRACT

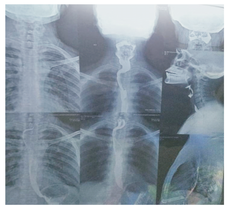
- Esophageal duplication cysts are benign, asymptomatic anomalies of foregut formation[1]. This anomaly is more common in pediatric rather than adult age groups. This is a case of esophageal duplication cyst with acute clinical presentation, treated successfully with surgical resection in which the patient underwent transhiatal esophagectomy with gastric conduit reconstruction with feeding jejunostomy and bilateral intercostal chest tube placement under general anesthesia and a histopathology report of the specimen to investigate the reasons behind symptoms was suggestive of Chronic active esophagitis with deep ulceration and non-caseating granulomas most consistent with Crohn's esophagus.

INTRODUCTION

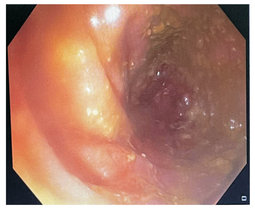
- The esophageal duplication cyst is an embryonic malformation first reported by Blassium in 1711. It is a rare form of congenital gastrointestinal duplication cyst found commonly in the esophagus ileum, jejunum, and colon. Pathogenesis is due to incomplete vacuolization during embryogenesis's fifth to eighth week and migration into the esophageal wall[2,3]. Cysts can be divided into cystic or tubular based on their structural configuration. Cystic duplications are more common (80% of the duplications) and do not communicate with the bowel lumen. Tubular duplications are less common and communicate with the lumen of the bowel.[4] A slight male preponderance has been observed with these cysts. They are mainly detected in childhood, with most duplications symptomatic within the first 2 years of life. However, late presentation in adulthood is also seen.[5] In adults, patients with esophageal duplication cysts usually do not exhibit symptoms and are often diagnosed during chest X-rays, photographs, or computed tomography for other conditions. However, cysts may become symptomatic owing to various complications such as narrowing of the esophagus, respiratory system compression, rupture, infarction (diseased tissue caused by a local lack of oxygen), or malignancy. Malignant degeneration can occur, albeit rarely. Esophageal duplication cysts can be lined by alimentary (squamous epithelium) or tracheobronchial mucosa regardless of where they are located. They have an envelope of smooth muscle and share a common wall with the esophagus[6,7]. Before surgical resection, a comprehensive evaluation of the mass and adjacent structures and other concurrent systemic anomalies is mandatory. Crohn's disease is a chronic inflammatory disease, which involves any segment of the mouth to the anus, but the involvement of the upper tract is less known, especially in the adult population, in which routine upper endoscopy is not indicated, as in the pediatric population.[8,9] The mean age at diagnosis was 31 years, and the mean interval between the diagnosis of CD and involving the esophagus was 1 year. The most frequent histological findings are chronic inflammatory infiltrate with a predominance of lymphocytes in the lamina propria and presence of ulcers[10]. Noncaseous granulomas are rare and observed in fewer than 25% of cases[10,11,12]. The granulomas are focal and are located in the deep submucosal and muscle, which could affect their identification[13].

CASE REPORT

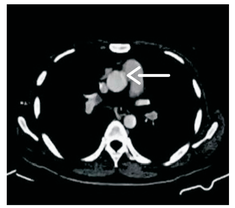
- A 37-year-old male patient presented at our clinical setting of a tertiary care hospital with a chief complaint of dysphagia for 25 days. Passing solid food was more difficult than liquid contents. The patient underwent a Barium Swallow to diagnose the cause behind Mechanical Dysphagia. Barium Swallow results were consistent with mass effect with mild mucosal irregularity noted in the upper thoracic esophagus with proximal dilatation. Approximately 56*27mm sized hypoechoic lesion noted along esophagus on screening sonography. An Upper GI scopy was scheduled which showed a submucosal bulge with overlying ulceration seen at the left upper esophageal wall causing mild luminal narrowing. To get a better view of the extent, a Computed Tomographic scan of the thorax was conducted, which suggested a 43*31*46 mm3-sized well-defined, irregular, non-enhancing, fluid-containing lesion with internal septa involving post cricoid area and upper 1/3rd of the thoracic esophagus on the left side causing enough luminal narrowing to push the esophagus on the right side with extension. The benign etiology was determined as an Esophageal Duplication cyst with possible differentials being a granulomatous esophagus disease. Informed and written consent was taken for resectioning approximately 10cm of the upper esophagus and 13 cm including the fundus and cardiac part of the stomach due to granulomatous changes in these parts.Intraop lesions were found adherent to the carina and left bronchus.The lesion was resected and extracted through cervical incision followed by transhiatal esophagectomy. End-to-side anastomoses of the cervical part of the esophagus and a gastric conduit which was created via a transhiatal open approach at the level of the cricoid. A feeding jejunostomy was placed for the patient's nutrition and bilateral intercostal chest tube placement was for unwanted complications. The resected 23 cm total specimen was sent for histopathology which on microscopic examination of multiple sections from the esophagus shows transmural inflammation, intraepithelial lymphocytes, deep ulceration,aphptoid ulceration, and non-caseating granulomatous inflammation with the presence of fibrosis, dilated lymphatics and hyperplastic squamous epithelium seen. No evidence of lymphocytic vasculitis, viral inclusions, fungal structures, parasitic strictures,dysplasia or malignancy seen. Section from stomach show mild superficial gastritis, no evidence of dysplasia or malignancy. Suggestive of chronic active esophagitis with deep ulceration and non caseating granulomatous inflammation(granulomatous esophagitis) with the possibility of Crohn's disease of the esophagus or tuberculous inflammation of the esophagus.HLA-DR (immunohistochemistry) to confirm Crohn's disease was positive and the patient was diagnosed with isolated Crohn's disease of the esophagus. The patient was followed up under observation as an outpatient and started tablets of azathioprine, methylprednisolone, and Rabeprazole and is doing well.



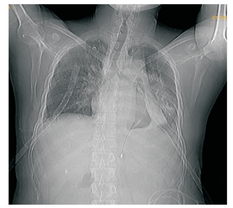
Barium swallow showing esophageal duplicationcyst just Below upper 1/3rd of esophagus



Esophageal duplication cyst on Upper GI scopy



CT scan of esophageal cyst demonstrated by the white line



Post surgery Xray showing nasogastric tube inside the gastric conduit with bilateral intercoastal drainage tubes

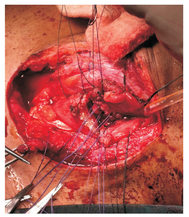
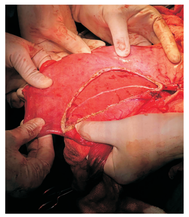
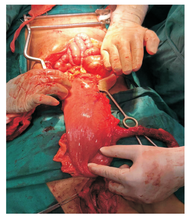


Image showing esophageal and gastric conduit end to side anastomosis



Sleeve gastrectomy for gastric conduit



Externalised esophagus with stomach

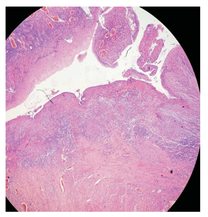


Image-histophathology slide suggestive of crohn's esophagus

DISCUSSION

- Esophageal duplication cysts are primarily diagnosed in early childhood, and in adults, they are primarily asymptomatic and are incidentally discovered during the radiological examination. They are more common in the distal esophagus.The diagnosis and treatment of esophageal cysts are still evolving. Diagnosis is aided by the use of computed tomography (CT) and endoscopic ultrasonography (EUS). Treatment is currently moving from thoracotomy to less invasive procedures, including video assisted thoracoscopic surgery(VATS) and endoscopic techniques. Esophageal involvement is one of the rare complications of CD. In a retrospective review, only 20/9900 patients were diagnosed with CD involving the esophagus (0.2%), with a median age of 31 at diagnosis.[15] The prevalence of endoscopic esophageal CD in the pediatric population is higher. One study showed endoscopic findings of esophageal CD in 7.6% of children with CD, with 17.6% showing histologic evidence of esophageal CD.[16] These numbers may be underestimating the true extent in adults, as there is no standard of performing an upper endoscopy in asymptomatic CD patients, which is commonly performed in the pediatric population. The mid and distal esophagus is more commonly affected.[14,15] The most common histological findings are nonspecific chronic inflammation, and granulomas are rare. A lymphocytic infiltrate in the lamina propria is commonly seen.[17] The findings of noncaseating granulomas are neither sensitive nor specific as they are present in only 7%-9% of esophageal CD patients.[18] The clinical diagnosis of esophageal CD can be difficult and is made through a combination of historical, endoscopic, and pathologic findings. Azathioprine is effective in prolonging remission in CD and in facilitating the discontinuation of steroid therapy.[19]

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