VOLUME - 12, ISSUE - 02, FEBRUARY - 2023 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Pathology

A RARE CASE OF OESOPHAGEAL DUPLICATION CYST IN ADULT

Shivangi Shukla	Post Graduate Resident Pathology, Era's Lucknow Medical College and Hospital, Era University, Lucknow, Uttar Pradesh- 226003, India
Noorin Zaidi	Associate Professor- Pathology, Era's Lucknow Medical College and Hospital, Era University, Lucknow, Uttar Pradesh- 226003, India
Ankita Yadav	Post graduate Resident- Pathology, Era's Lucknow Medical College and Hospital, Era University, Lucknow, Uttar Pradesh- 226003, India
Nirupma Lal	Professor and head of the Department- Pathology, Era's Lucknow Medical College and Hospital, Era University, Lucknow, Uttar Pradesh - 226003 India
Sumaiya Irfan*	Associate Professor- Pathology, Era's Lucknow Medical College and Hospital, Era University, Lucknow, Uttar Pradesh- 226003, India *Corresponding Author

ABSTRACT A Duplication cyst of the oesophagus is the 2nd most common congenital anomaly of the Gastrointestinal tract. It has an estimated prevalence of 0.012% with a higher predominance in males, representing the 10-15% of all gastrointestinal duplication defects. Although it is a common finding in children, diagnosis of an oesophageal duplication in adults is rare. They are often asymptomatic but they may experience symptoms such as stridor or dysphagia corresponding to the lesion within the neck or mediastinum or be complicated by intracystic haemorrhage, rupture & infections. Here we present a case of an adult patient presenting with an oesophageal duplication cyst.

KEYWORDS : Duplication cyst; Dysphagia,

INTRODUCTION-

Gastrointestinal (GI) duplication cysts are rare inherited disorders commonly occurring in the jejunum, ileum, colon and oesophagus out of which oesophageal duplication cyst constitutes 10-15% of all congenital duplication cysts involving the GI tract [1,2]. Congenital oesophageal malformations like oesophageal duplication cysts are uncommon, especially in adults. Oesophageal duplication cysts in infants or children are usually detected during an examination for respiratory distress or feeding problems. The natural progression of the illness is known to be complicated by bleeding, infection, and mass effects.

Oesophageal duplication cysts in adults are usually asymptomatic, and the diagnosis is frequently based on an unexpected finding on a chest radiograph. For symptomatic oesophageal duplication cysts, complete surgical excision is the standard treatment. Oesophageal duplication cysts can be accurately diagnosed before surgery with endoscopic ultrasonography.[2]

Here we present a case of a young adult with complaints of gradual right-sided pleuritic chest pain and fever. Limited data is present in the literature regarding this rare congenital malformation in symptomatic adults.

Case Report:

Clinical history:

A 25-year-old male presented with complaints of gradual right-sided pleuritic chest pain and fever. He had on-and-off abdominal pain and recurrent vomiting associated with pain in the chest and a cough with shortness of breath after taking meals for 20-22 days which was relieved by taking medications for a short period. Patient gave no history of any significant weight loss, dyspnoea, hematemesis, melena, or peripheral edema.

On examination, his vitals were stable with blood pressure of 120/70 mmHg, pulse rate of 90/min and respiratory rate of 24/min. The rest of the abdominal and respiratory examinations were within normal limits. Routine haematological and biochemical parameters were also tested and they were within normal limits, on further evaluation.

CECT thorax revealed a "non-enhancing fluid accumulation cystic lesion measuring ~3.5 x 3.8 x 5.8 cm (AP x TR x CC) closely abutting the mid and lower esophagus, extending from the lower border of T9 to mid of T12 vertebrae on the right side.

The lesion was causing a mass effect on the esophagus leading to luminal narrowing. The anterior lesion was abutting the posterior wall of the left atrium and inferior vena cava. Complete surgical excision of the cyst with exteriorization of the base was performed and it was sent for histopathological evaluation.

Grossly:

The specimen was received at our histopathology lab as two gray-white to gray-brown soft tissue pieces measuring 3.6 x 1.5 x 1.0 cm. Routine tissue processing and H&E staining as per the protocol standardized in our histopathology lab were executed.

Microscopically:

H&E (Haematoxylin and Eosin) stained section showed cyst wall lined by stratified columnar epithelium underlying submucosa and thickened smooth muscle-muscular layer present. Showed features consistent with oesophageal duplication cyst. No neoplastic pathology was identified.

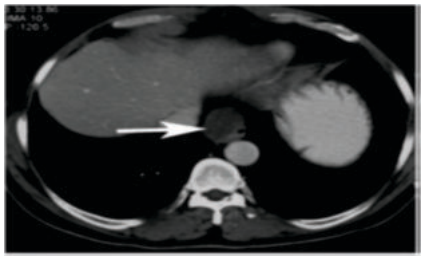

Fig 1: CT Scan shows a 3.5 × 3.8 × 5.8 cm Showing a well-defined homogenous cystic lesion (white arrow)

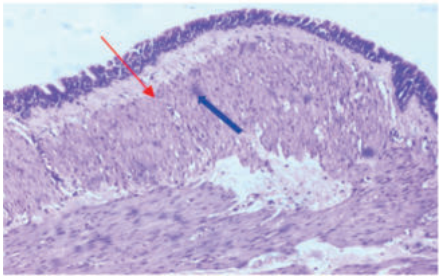

Fig 2: 4x view of duplication cyst: Stratified columnar epithelium (Red arrow) Underlying Submucosa (Blue arrow)

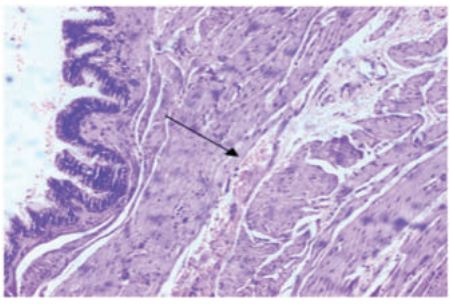

Fig 3: 10x view of duplication cyst: Thick smooth muscle-muscularis layer (Black arrow)

DISCUSSION

The estimated prevalence of oesophageal duplication cysts, an uncommon congenital embryonal abnormality, is 0.0122% [1]. These cysts occur as a result of the primitive oesophagus failing to vacuolate during embryologic development between the fourth and eighth gestational week [3] Nikhil Sonthalia et al. reported a case that was similar to ours in a 30-year-old, male patient. [13].

The majority of them are found in the mediastinum, seen as distinct masses that run alongside or in parallel with the oesophagus. Adult intramural oesophageal cysts are thought to be quite rare [4,5]. Squamous or enteric epithelium lines duplication cysts, which also have a second layer of oesophageal smooth muscle around them. They are most commonly seen in male with male-to-female ratio of around 2:1, and they are most frequently found in the lower region of the oesophagus. There have also been a few reports of intraabdominal oesophageal duplication cysts [6]. Duplication cysts can be either cystic or tubular, however, cystic types are more common. They might additionally appear as a diverticulum in children and may be asymptomatic, while the oesophageal duplication cysts when diagnosed in adulthood are more symptomatic [7]. Dysphagia, chest pain, epigastric discomfort, vomiting, stridor, cough, haemorrhage, and hematemesis may all be present in the patient. Additionally, these symptoms might be found by chance. The majority of symptomatic duplication cyst reports involve children [8]. On upper GI endoscopy, the differential diagnosis must be taken into consideration for lesions like lipomas, leiomyomas, and GI stromal tumours (6,9). In these cases, an endoscopy-guided mucosal biopsy is not very helpful. Other differential diagnoses that can be taken into account in these situations include mediastinal cysts such as hydatid cysts, pericardial cysts, bronchogenic cysts, or mediastinal lymphadenopathy.

On CT scans of the thorax, a homogenous low attenuating cystic lesion with well-defined boundaries along the oesophageal wall producing varying lumen deficit is shown. EUS is the preferred imaging technique for identifying mediastinal cysts and lymph nodes. While the histopathological evaluation offers to be the best diagnostic tool (14), certain pathological criteria like the cyst being attached to the oesophageal wall, the cyst communicating with the epithelia, its origin from the gastrointestinal tract and it is underlined by two layers of muscularis propria and the muscularis propria of the oesophagus continuing with the muscular layer of the cyst are extremely helpful. The role of surgery is not entirely clear because, in asymptomatic patients, surgical intervention can result in long-term consequences such as heartburn and reflux esophagitis and have a mortality rate of about 1% [12].

CONCLUSIONS

Esophageal duplication cysts in adults are rarely diagnosed. These are more prevalent in males and are usually asymptomatic. However, depending on their position and size, the esophageal duplication cysts may be linked to the common symptoms of dysphagia, chest tightness, and weight loss. The cysts may be discovered incidentally. Esophageal duplication cysts are treated with complete surgical resection avoiding severe complications like acute rupture, rapid enlargement, and even malignant transformation of esophageal duplication cyst.

REFERENCES

1. Olajide AR, Yisau AA, Abdulraseed NA, Kashim IO, Olaniyi AJ, Morohunfade AO. Gastrointestinal duplications: Experience in seven children and a review of the literature. Saudi J Gastroenterol. 2010;16(2):105-109.
2. Javan NG, Debnath J, Kumar A, Das CJ. Oesophageal duplication cyst: an unusual cause of retrosternal pain and dysphagia in an adult. Singapore Med J. 2008 Sep 1;49(9):e243.
3. Pisello F, Geraci G, Arnone E, Sciutto A, Modica G and Sciume C. Acute onset of esophageal duplication cyst in adult. Case report. G Chir 2009; 30: 17–20. (2nd line in discussion)
4. Oteiza F. Congenital esophageal cysts - two cases in adult patients. Hepatogastroenterology. 1999;46:2405-2408.
5. Cioffi U, Bonavina L, De Simone M, Santambrogio L, Pavoni G, Testori A, Peracchia A. Presentation and surgical management of bronchogenic and esophageal duplication cysts in adults. Chest. 1998;113(6):1492-1496.
6. Martin ND, Kim JC, Verma SK, Rubin R, Mitchell DG, Bergin D, Yeo CJ. Intra-abdominal esophageal duplication cysts: a review. J Gastrointest Surg. 2007;11(6):773- 777
7. Karnak I, Ocal T, Senocak ME, Tanyel FC, Buyukpamukcu N. Alimentary tract duplications in children: report of 26 years' experience. Turk J Pediatr. 2000;42(2):118-125
8. Wiechowska-Kozlowska A, Wunsch E, Majewski M, Milkiewicz P. Esophageal duplication cysts: endosonographic findings in asymptomatic patients. World J Gastroenterol. 2012;18(11):1270-1272.
9. Kuhlman JE, Fishman EK, Wang KP, Siegelman SS. Esophageal duplication cyst: CT and transesophageal needle aspiration. AJR Am J Roentgenol. 1985;145(3):531- 532.
10. Diehl DL, Cheruvattath R, Facktor MA, Go BD. Infection after endoscopic ultrasound-guided aspiration of mediastinal cysts. Interact Cardiovasc Thorac Surg. 2010;10(2):338-340.
11. Wildi SM, Hoda RS, Fickling W, Schmulewitz N, Varadarajulu S, Roberts SS, Ferguson B, et al. Diagnosis of benign cysts of the mediastinum: the role and risks of EUS and FNA. Gastrointest Endosc. 2003;58(3):362- 368.
12. Sonthalia N, Jain SS, Surude RG, Mohite AR, Rathi PM. Congenital esophageal duplication cyst: a rare cause of dysphagia in an adult. Gastroenterology Research. 2016 Oct;9(4-5):79.
13. Moulton MS, Moir C, Matsumoto J, Thompson DM: Esophageal duplication cyst: a rare cause of biphasic stridor and feeding difficulty. Int J PediatrOtorhinolaryngol 2005, 69:1129-1133.
14. Simons-Linares CR, Wander P, Vargo J, Chahal P. Endoscopic ultrasonography: An inside view. Cleve Clin J Med [Internet]. 2020;87(3): 175–83. Available from: http://dx.doi.org/10.3949/ccjm.87a.19003
15. Sangüesa Nebot C, Llorens Salvador R, Carazo Palacios E, Picó Aliaga S, Ibañez Pradas V. Enteric duplication cysts in children: varied presentations, varied imaging findings. Insights Imaging [Internet]. 2018;9(6):1097–106. Available from: http://dx.doi.org/10.1007/s13244-018-0660-z
16. Liu R, Adler DG. Duplication cysts: Diagnosis, management, and the role of endoscopic ultrasound. Endosc Ultrasound [Internet]. 2014;3(3):152–60. Available from: http://dx.doi.org/10.4103/2303-9027.138783
17. Papparella A, Nino F, Noviello C, Marte A, Parmeggiani P, Martino A, Cobellis G. Laparoscopic approach to Meckel's diverticulum. *World J Gastroenterol* 2014; 20(25): 8173-8178 [PMID: 25009390 DOI: 10.3748/wjg.v20.i25.8173]
18. Gandhi D, Garg T, Shah J, Sawhney H, Crowder BJ, Nagar A. Gastrointestinal duplication cysts: what a radiologist needs to know. Abdom Radiol (NY) [Internet]. 2022;47(1):13–27. Available from: http://dx.doi.org/10.1007/s00261-021-03239-w
19. Tripathy TP, Patel R, Mohanty SK, Mohanty SP. Ileocolic Intussusception in an Infant with Ileal Duplication Cyst as a Lead Point- A Case Report. J Clin Diagn Res [Internet]. 2021; Available from: http://dx.doi.org/10.7860/ jcdr/2021/49890.15114
20. Gonzalez-Urquijo M, Hinojosa-Gonzalez DE, Padilla-Armendariz DP, Saldaña-Rodriguez JA, Leyva-Alvizo A, Rodarte-Shade M, et al. Esophageal duplication cysts in 97 adult patients: A systematic review. World J Surg [Internet]. 2022;46(1):154–62. Available from: http://dx.doi.org/10.1007/s00268-021-06325-8