CASE REPORT

Complicated oesophageal duplication cyst in a 55-year-old man: a rare case from Pakistan

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Abstract

Duplication cysts are congenital anomalies that arise during early embryonic development. They occur in the small intestine, oesophagus, stomach, and colon. Oesophageal duplication cyst (ODC) is rare among newborns, but even rarer in adults.

We report the case of a 55-year-old male with the presenting complaints of haematemesis, dysphagia, and significant weight loss for the past eight months. Clinical examination was unremarkable, except that the patient had pallor. Computed tomography scan confirmed the presence of ODC. Subtotal oesophagectomy with gastric pull-up and sub-aortic oesophagogastrostomy was completed. Histopathology was consistent with foregut duplication cyst. Post-operative period was complicated by left basal atelectasis, and the patient was discharged without long-term sequelae. Adult ODC can be regarded as a diagnosis of exclusion after ruling out oesophageal malignancy, high-grade oesophageal varices, mediastinal masses, and submucosal lesions. Complete surgical excision with restoration of gut continuity is optimal management in complicated, large (>5cm) ODC.

Keywords: Oesophageal cyst, Oesophagectomy, Dysphagia, Jejunostomy, Pakistan.

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Introduction

Duplication cyst is a rather rare embryological entity. It was first reported by Blassium in 1711 and is mostly seen in the oesophagus, ileum, jejunum, and colon.[1] Alimentary tract duplications can potentially occur anywhere from tongue to anus.[2] They are usually single, variable in size, and morphology can include spherical, tubular, or cystic types. Sometimes these cysts have heterotrophic gastric mucosa, or pancreatic tissue or are even associated with spinal abnormalities.[3]

Oesophageal duplication cyst (ODC) accounts for 20% of all gastrointestinal duplication cysts and commonly involves the distal oesophagus.[4] ODC is usually an early childhood diagnosis. However, it can be revealed incidentally or present symptomatically in early or late adulthood.[1,2] The aetiology of ODC is not clear. They are formed due to faulty intrauterine recanalization of the oesophagus during the fourth to eighth week of foetal development.[4] The symptoms of ODC can vary from dysphagia and chest pain to massive haematemesis leading to dire surgical emergency.[1] ODC can manifest as intestinal obstruction, gut perforation, and respiratory distress in children.[5]

The existing English scientific literature contains only a handful of case reports illustrating management by surgical or non-surgical techniques. There are no specific guidelines for the management of asymptomatic or symptomatic ODC in a resource-limited setting. After a comprehensive literature search using PubMed Central and Google Scholar, we document the first case of a successfully managed ODC in an adult from Pakistan.

Case Report

A 55-year-old man, with no known comorbidities, presented to the surgical outdoor clinic of Shaikh Zayed Medical Complex, Lahore, in January 2023 with the presenting complaints of sudden haematemesis, dysphagia, and a significant weight loss of more than 10 kg in the past eight months. He had five-to-six episodes of haematemesis, mostly post-prandial. He got symptomatic treatment from local clinics and ignored his presenting complaint in the long run. Haematemesis was accompanied by progressive dysphagia which worsened the intake to liquids only until presentation. General physical examination and systemic examination was not remarkable, except that the patient was clinically pale.

The patient was subjected to relevant haematological and biochemical laboratory workup, followed by radiological investigations. Laboratory results showed all parameters

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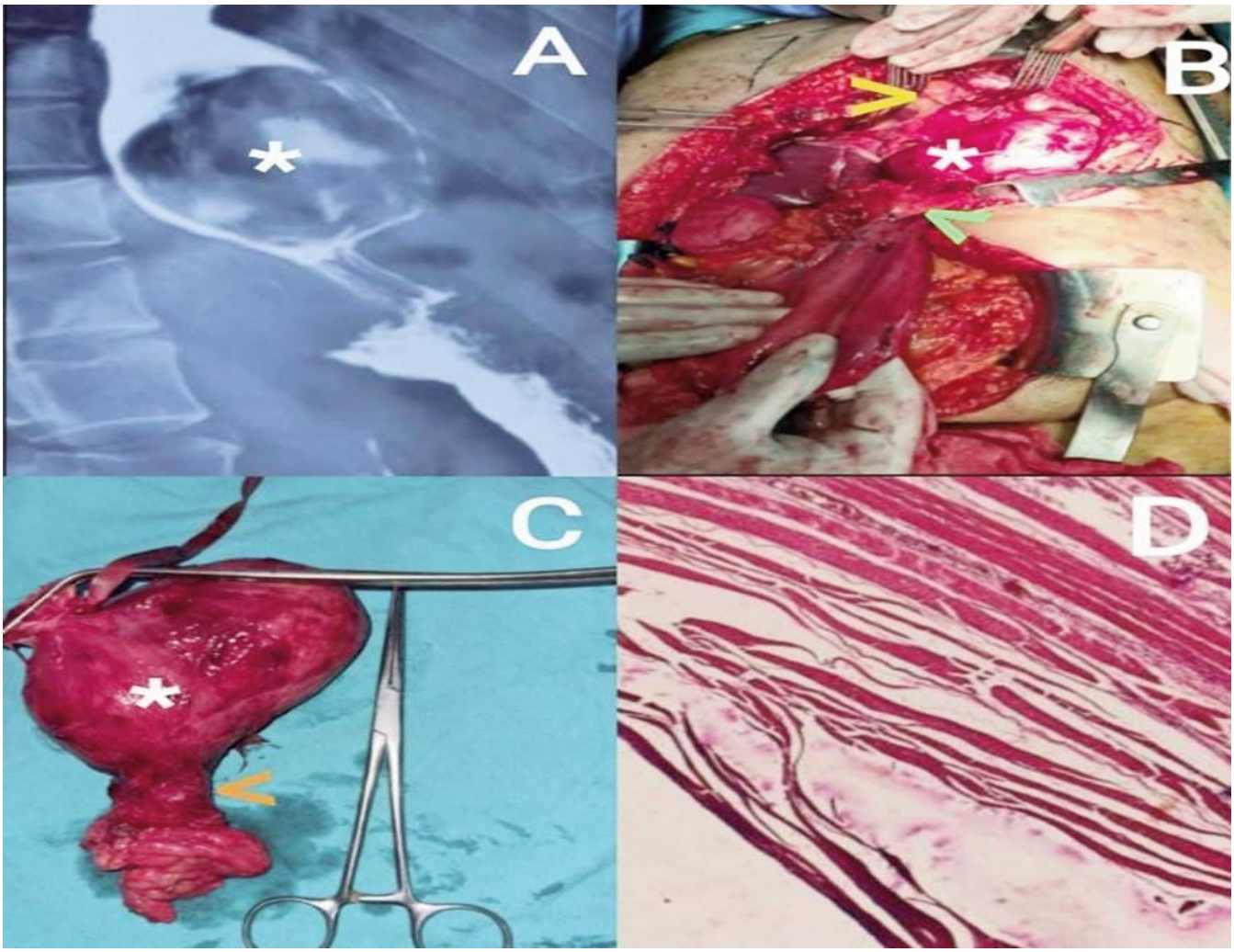

Figure-1: A. Oesophageal cyst visible on radiograph post-Barium swallow, marked in a white Asterisk (*); B. Intra-operative picture of oesophageal cyst, retracted left lung and left dome of diaphragm, marked in a white Asterisk (*), yellow arrow (>) and green arrow (<), respectively; C. Excised oesophageal cyst and lower oesophageal junction, marked in a white Asterisk (*) and orange arrow (<), respectively; and D. Wall of the cyst with low cuboidal to flat epithelium and underlying muscle layer.

in normal ranges, except for severe anaemia with haemoglobin (Hb) 5.8g/dL (reference range 13.5-16.5g/dl). Chest X-ray (CXR) illustrated a vague mass in lower mediastinum. Barium swallow demonstrated a well-rounded defect in distal oesophagus (Figure 1A). Computed tomography with contrast scan of the chest, abdomen, and pelvis (CT CAP) confirmed "a well-defined, thin-walled hypo-dense lesion, measuring 45x130x92mm in posterior mediastinum at the level of T6-T10 vertebrae, extending proximally from subcarinal region, stretching the carina and widening the angle". Upper gastrointestinal endoscopy (UGIE) with endoscopic ultrasound (EUS) was performed under sedation, which revealed a benign-looking swelling at a distance of 25cm from the incisor teeth. The swelling was about 7-8 cm in length, partially obstructing the oesophagus, however, the scope passed easily.

A multidisciplinary team meeting was conducted to assess the operative risk for further management. Upfront surgical approach was planned with cardiothoracic surgeons on board. The patient was optimised with units of blood achieving a preoperative Hb 9.7g/dL (reference range 13.5-16.5g/dl). In the same month, the patient underwent subtotal oesophagectomy with gastric pull-up, subaortic oesophagogastrostomy, and left diaphragmatic repair via upper midline and left anterolateral thoracotomy. A bilateral thoracostomy was done to access distal oesophagus. The surgical specimen was suggestive of "an 8x8cm, cystic, oval, well-circumscribed lesion in the muscularis propria of mid and distal oesophagus, about 3cm proximal from lower oesophageal sphincter, which was not adherent to the

surrounding structures''. The rest of the visualised organs appeared normal (Figure 1B). The lesion was excised and oesophagogastrostomy anastomosis hand sewn in an interrupted fashion was performed (Figure 1C). Feeding jejunostomy was done at the end of the procedure to ensure early post-operative enteral feeding.

The patient's recovery from general anaesthesia was smooth and there were no immediate post-operative complications. He was shifted to the intensive care unit (ICU) for strict haemodynamic monitoring and nutritional support. However, the patient's seven-day ICU stay was affected by left basal atelectasis, which was relieved with the course of broad-spectrum antibiotics and aggressive chest physiotherapy. Liquid-based feed via feeding jejunostomy was started on the third post-operative day.

Oral intake was started on the seventh post-operative day and he was stepped down to the surgical high dependency unit (HDU) on the same day. Right and left chest drains were removed on the seventh and tenth post-operative day, respectively. He was discharged 48 hours after removal of chest tube without any significant complications. Histopathology was consistent with adult ODC (Figure 1D). He was followed-up at the surgical outdoor clinic on a biweekly basis for one month, on a monthly basis for the next two months, and telephonically on the sixth month of discharge. Normal wound healing was achieved and the feeding jejunostomy was removed on his second follow-up visit after the resumption of normal oral feeding. The patient is living a healthy life.

Discussion

It has been estimated that ODC occurs in one in 8,000 live births. ODC is twice as common in males as in females.[1,4] Gonzalez-Urquijo et al. have recently conducted a systematic review of adult ODC concluding the average age of presentation to be 42.3 years. The current patient was in his sixth decade at presentation.[1,4]

ODC can become symptomatic in adults when they become large. An old Finnish study documents dysphagia (70%), epigastric pain (20%), retrosternal pain (10%), and respiratory symptoms including cough, stridor, and wheezing to be the most frequent clinical manifestations at presentation.[6] However, the current patient neglected his symptoms and presented at a complicated stage with haematemesis, severe anaemia, significant weight loss, and dysphagia. These manifestations were indicative of malignancy.

Tunisian authors conducted a review of the existing literature from 2000 to 2023, reporting that a surprising 76% of patients were adults in the age range 18 – 70 years. However, 80% of children with foregut duplication cysts are diagnosed before the age of two.[2] About 48% of patients had their duplication cysts discovered incidentally.[2] There is scarcity of data to support the reason behind incidental presentation among adults. Systematic review by Mexican doctors also advocates that the size of ODC influences presenting symptoms, rather than the location of the lesion.[4] This is reinforced by Balakrishnan et al. in their unicentric study showing that lesions located in the distal oesophagus tend to remain undetected for many years.[5] In the current patient, it was not symptomatic until the lesion was about eight centimetres in size. Therefore, gradual increase in size and location of duplication cyst could be the contributing factors for late presentation in this patient.

Thorough investigation is important as the patient can be challenging during surgery and anaesthesia, with special regard to risk of pneumothorax, pneumo-mediastinum, vascular injuries during airway management and central venous access.[1] Chest radiography with or without Barium swallow may be helpful in preliminary diagnostic evaluation for relatively larger lesions. Smaller lesions may not be apparent on an X-ray.[7] Some reports mention the use of ultrasonography, probably because of wider availability and better safety than ionic radiation. However, it is operator-dependent and smaller lesions are usually not identified, especially in obese patients. In the current patient, CT CAP delineated the site, location, and proximity of the surrounding structure.[1]

It is inevitable to mention the importance of EUS to delineate the origin of pathology.[8,9] EUS can be useful to differentiate between ODC and a malignant mass. EUS uses the function of direct lumen visualisation in real time coupled with highlighting the layers of the cyst wall; it is superior in the diagnosis of subepithelial intraluminal lesion.[8] In the present case, both CT CAP and EUS had contributed to pre-operative diagnosis and ruling out other possibilities.

The open versus minimally invasive approach has been debated for a long time. There are proven benefits of the minimally invasive/laparoscopic/video-assisted thoracoscopic surgery, but currently, there is insufficient data on long-term follow-up. Open approaches are associated with longer hospital stay and higher risk of post-operative complications.[7,10] However, for resection of large duplication cysts (especially greater than 5cm) and to restore the continuity of the alimentary canal post-resection, an open conventional approach was favoured as a safe option in this patient.

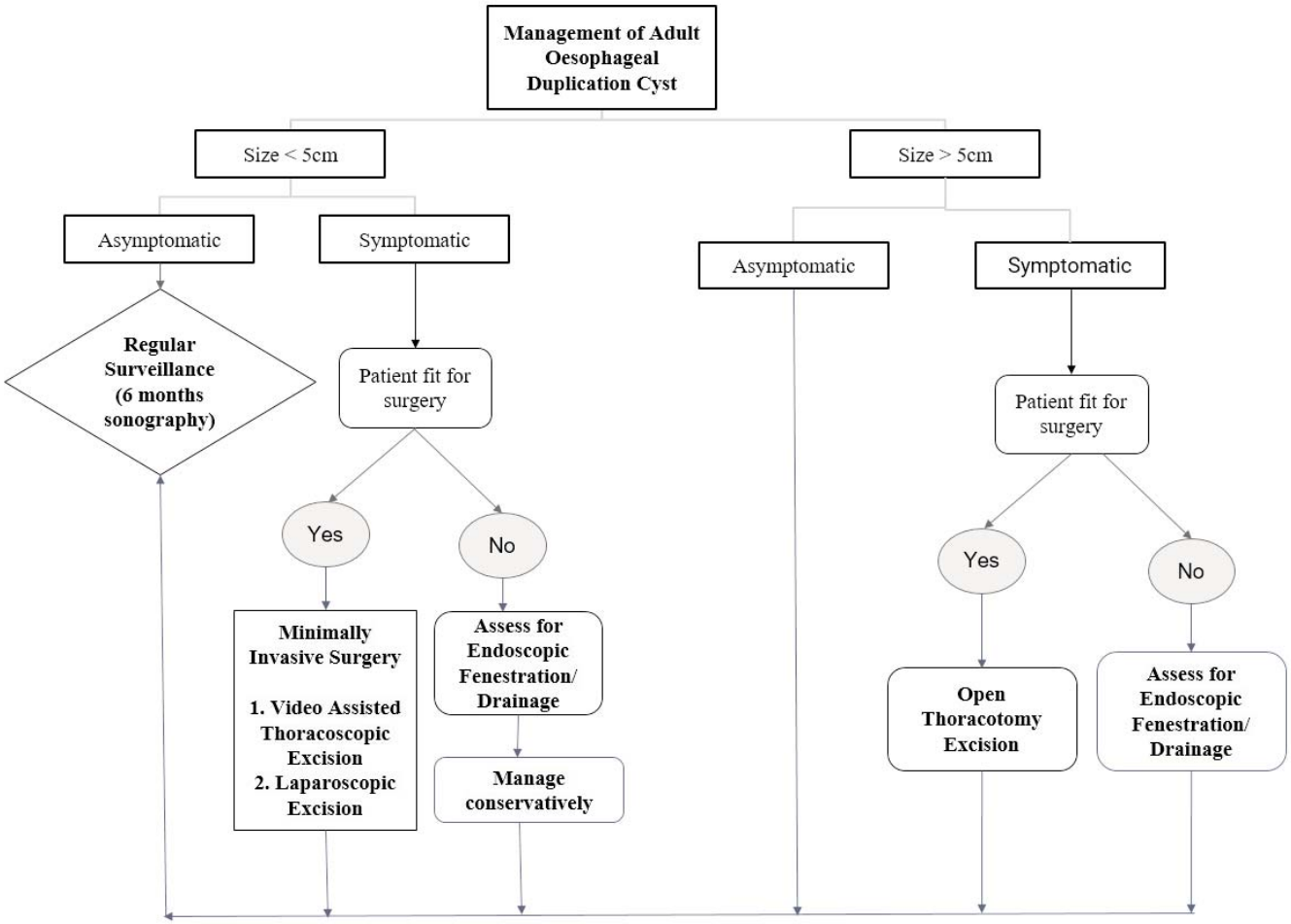

Figure-2: Management Algorithm of Adult Oesophageal Duplication Cyst (ODC).

Endoscopic fenestration, although a possible intervention, is a double-edged sword.[8] Endoscopic fenestration can present with complications of infections, spontaneous lumen closure, and oesophageal perforation.[1] It was concluded that as per management of complicated oesophageal duplication cyst, the open surgical approach is relatively safer and suited to a resource-limited setting. The present patient was symptom-free and gained weight due to the restoration of enteral nutrition. There was no cyst recurrence at three-month follow-up, similar to the findings documented in the case studied by Sghaier et al.[2] Figure 2 illustrates our prototype management algorithm for adult oesophageal duplication cysts based on a literature review of the cited articles.

Conclusion

Sequential diagnostic work-up and meticulous consideration of differential diagnoses helped us reach the pathology of oesophageal duplication cyst in the adult patient. It is difficult to suspect adult ODC based on patient presentation and clinical examination alone. Large, symptomatic ODC needs urgent attention. The open approach offers a complete and safe surgical excision and the restoration of gut continuity in a large, complicated ODC.

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AUTHOR'S CONTRIBUTION:

AAK: Concept, drafting, revision, final approval and agreement to be accountable for all aspects of the work.

AJ: Design, drafting, final approval and agreement to be accountable for all aspects of the work.

HMQ: Supervision, concept, design, data analysis, interpretation, drafting, revision, final approval and agreement to be accountable for all aspects of the work.

NFH & PY: Data acquisition, revision, final approval and agreement to be accountable for all aspects of the work.