International Journal of Surgery Case Reports 93 (2022) 107014

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Esophageal stenosis: A case of delayed dysphagia in an infant from northern Tanzania

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ARTICLE INFO

Keywords:
Dysphagia
Esophagogram
Esophageal malformation
Esophageal stricture
Regurgitation
Malnutrition

ABSTRACT

Introduction and importance: Congenital stenosis of the esophagus is often overlooked in children with feeding difficulties leading to delayed diagnosis and treatment. This can have detrimental effects on the child's growth and general health, ranging from malnutrition to life-threatening complications such as aspiration pneumonia.
Case presentation: We present a case diagnosed at the age of 3 years when the child already had developed moderate malnutrition and offer the diagnostic and surgical findings in the hope of helping others suspect and clench the diagnosis early.
Clinical discussion: With an incidence of up to 1 in 25,000, congenital stenosis of the esophagus is definitely a congenital malformation worth knowing about, particularly since misdiagnosis often leads to delayed intervention as was in our case. History of feeding difficulty and failure to thrive are often the triggers to seeking medical attention. The confirmation can be easily made with contrast imaging opening up the possibility for surgical and non-surgical management.
Conclusion: With a high index of suspicion, diagnosis can be made relatively easily, allowing for early intervention, which can prove to be lifesaving.

1. Introduction

Congenital esophageal stenosis (CES) is a rare pathology, and the diagnosis is usually delayed. The first clinical signs and symptoms are abnormalities in the swallowing mechanism caused by the intrinsic narrowing of the esophagus [1]. The challenges with feeding infants often lead to the symptoms being overlooked and diagnosis being delayed. It requires a detailed history and a high level of suspicion, aided with basic radiologic investigations. Treatment usually involves surgery depending on the characteristic of the stenosis [2]. Herein we report of a three-year-old male child with difficulty swallowing and a characteristic esophagogram.

This work has been reported in line with the SCARE 2020 criteria [3].

2. Case presentation

A 3-year-old male presented with progressive difficulty in swallowing solid food. The mother noticed the child struggled to swallow solids from weaning, progressing to choking and vomiting when fed solids at the time of presentation. Mothers' concern was heightened by the child's failure to gain weight.

The child had been delivered by cesarean section at a gestational age of 33 weeks due to the mother having vaginal bleeding and pregnancy-induced hypertension. The child weighed 2300 g at birth and was not noted to have any malformations or illnesses. At the time of admission, the child appeared to be alert, not in distress, not cyanotic, and neither pale nor jaundiced. He was small for age with a height of 77 cm and weight of 9 kg (Weight for height −2 z-score).

On his systemic examination, he had nasal congestion with grade-2 tonsilar hypertrophy. Other systems were unremarkable on examination. His labs revealed a normal leukocyte count, hemoglobin of 10 g/dL, and platelet count of 574 × 10^9. His serum sodium and potassium were in range, and he had a serum creatinine of 24 μmol/L, BUN of 2.71 mmol/L, and an albumin level of 40.3 g/L. Barium swallow showed dilated proximal two-thirds of the esophagus with distal rat-tailing (Fig. 1). Nasal bone X-ray showed enlarged adenoids narrowing the posterior pharyngeal airway by 90%. His plain chest X-ray and

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https://doi.org/10.1016/j.ijscr.2022.107014
Received 16 February 2022; Received in revised form 27 March 2022; Accepted 31 March 2022
Available online 2 April 2022

echocardiogram were normal, with an ejection fraction of 67%.

He underwent an esophagoscopy that showed dilated esophageal lumen with residual food particles. At 18 cm from the upper premolar, the esophageal lumen was stenotic, and the scope couldn't pass beyond this, but the lower esophageal sphincter could be visualized beyond the stricture. Multiple biopsies were taken showing features of acute esophagitis.

The child was given a high protein diet and plumpy nuts (ready to use therapeutic food) for two weeks. His control labs showed a total protein of 65 g/L, albumin of 38 g/L, hemoglobin of 10.9 g/dl, and platelets of 774×10^9. He was scheduled for thoracotomy which was performed by the consultant cardiothoracic surgeon, whereby the esophagus was approached through the sixth intercostal space. There was a congenital stricture near the diaphragm approximately 2 cm long which was excised (Fig. 2), and the esophagus was repaired primarily. The child received 200mls of whole blood during surgery, and post-operatively, was taken to intensive care for close monitoring. Unfortunately, ten hours post-operatively, the child sustained a sudden cardiac arrest, and resuscitation attempts were unsuccessful. Unfortunately post mortem was not done as was not consented for.

3. Discussion

CES is a relatively rare entity with an incidence of 1 in 25,000 to 50,000 live births with a slight male predominence [1]. Depending on the severity of the stenosis, the age of presentation can be highly variable, with some being incidental findings during imaging in adulthood, while others have severe symptoms early on [4]. Often symptoms appear when the child starts to take solids [5]. Initial presentation is vomiting and regurgitation [5], which in an infant being introduced to feeds is not uncommon, delaying the diagnosis [5]. The persistence of the vomiting, particularly when taking solids, is key to the diagnosis. Esophageal atresia should be included as a differential to suspected foreign body impaction and failure to thrive, accompanied by vomiting.

Esophageal stenosis can either be congenital (CES) or acquired. CES is categorized into three types: fibromuscular stenosis (FMS), esophageal membranes or web, and tracheobronchial remnants (TBR) [1,5]. TBR is the most common of these, and FMS is the least occurring [6]. Acquired esophageal stenosis can be divided into traumatic, inflammatory, peptic, and iatrogenic [5]. Ingestion of corrosives (accidental or intentional) remains an important cause of acquired esophageal stenosis [4].

The symptomatology of CES is also influenced by the location of the stenosis, which is itself influenced by the etiology. CES of the distal third esophagus is commonly due to trachea-bronchial remnants and is

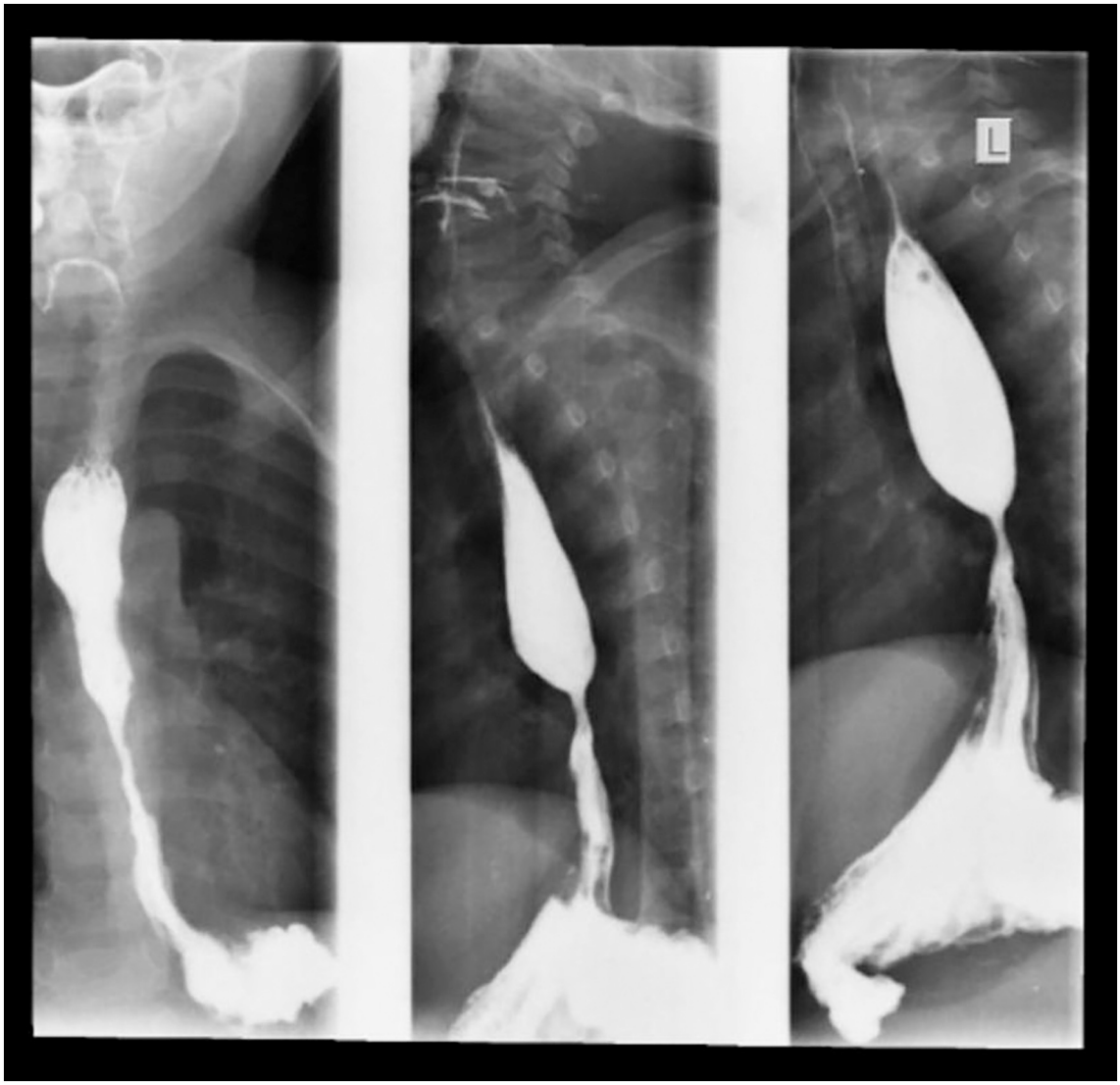

Fig. 1. Barium esophagogram showing proximal two-third esophageal dilatation with distal narrowing.

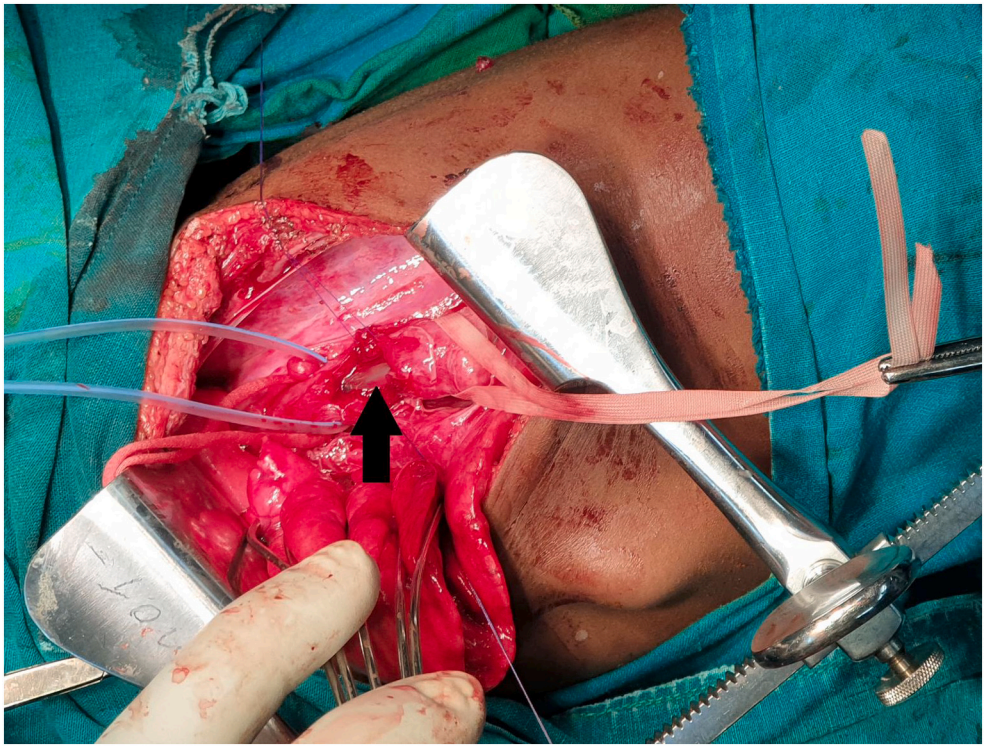

Fig. 2. Excised stenotic esophagus (arrow) with nasogastric tube in situ.

associated with vomiting. In contrast, middle CES is commonly due to web/membrane and fibromuscular etiologies and are commonly associated with respiratory symptoms [7]. Other congenital malformations (e.g., esophageal atresia, duodenal web, congenital heart defects) and secondary symptoms stemming from complications such as aspiration may also be present [2,8].

Esophagogram in CES shows sharp, circumferential luminal narrowing with smooth and tapering margins associated with proximal esophageal dilatation without obvious signs of shouldering as this was evident in the index case [7]. Endoscopic examination of the esophagus can also be helpful to visualize and rule out mucosal causes of the stenosis [4]. This is often not accessible in many resource-limited settings, and we have to rely on radiological studies.

CES can be managed in a surgical or non-surgical (conservative) manner. The main considerations for the management modality are the length, aspect, location, and the number of stenoses [9]. Conservative management through endoscopic balloon dilatation of the stenosis often gives temporary relief of symptoms, so surgery remains the definitive management for most cases of CES[9]. The surgical approach depends on the site of the lesion. Stenosis close to the gastro-esophageal junction can be approached abdominally, while thoracic lesions can be managed using the right or left thoracotomy incisions [10]. Lesions higher-up around the cervical esophagus can be approached through the neck [10]. Though surgery is stated to be definitive, it poses various risks such as anastomotic leaks, restenosis, gastro-esophageal reflux due to the alternation of esophago-gastric angle. Esophageal dilatation procedures are not without risk and can cause perforation, aspiration pneumonia, or cardiac arrest [10].

4. Conclusion

Although relatively rare, congenital esophageal stenosis should always be suspected in children who present with dysphagia. Contrast radiological studies such as barium esophagogram are affordable and accessible means of making the diagnosis even where resources are relatively limited and can allow for early intervention sparing the child of secondary comorbidities, which could be life-threatening.

Consent

Written informed consent was obtained from the patient's mother for publication for this case report; additionally, accompanying images have been censored to ensure that the patient cannot be identified. A copy of the consent is available on record.

Ethical approval

Ethical approval was obtained from the Department of General surgery, Kilimanjaro Christian Medical Centre.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Authors' contributions

- Jay Lodhia – conceptualization and writing of the script
- Samwel Chugulu – lead surgeon
- Joylene Tendai – reviewed medical records
- Rune Philemon - conceptualization and writing of the script

Research registration

N/A.

Guarantor

Jay Lodhia – Corresponding author.

Declaration of competing interest

The authors declare they have no competing interests.

Acknowledgement

The authors would like to thank the child's mother for permission to share her child's medical history for educational purposes and publication.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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