CASE REPORT

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An Uncommon Cause Of Dysphagia

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Abstract

Swallowing difficulties are frequently attributed to intrinsic esophageal or neuromuscular disorders. However, in select cases, subtle extrinsic anatomical factors may underlie progressive dysphagia, requiring vigilant clinical and radiological assessment. We present the case of a young adult with non-specific gastrointestinal symptoms whose underlying diagnosis revealed a rare anatomical variant. A 20-year-old man with Type 1 Diabetes Mellitus and congenital scoliosis presented with a three-month history of dysphagia, followed by acute abdominal pain and vomiting. Initial evaluation revealed diabetic ketoacidosis, which was managed conservatively. Further diagnostic workup, including barium swallow and CT angiography, uncovered an unusual cause of esophageal compression. The patient underwent definitive surgical intervention involving a minimally invasive sternotomy, dissection of the arch vessels, and strategic revascularization of the compressing structure. Post-operative recovery was uneventful, and symptoms markedly improved. This case underscores the importance of considering rare congenital anomalies in young patients with unexplained dysphagia. Early imaging and targeted surgical management can lead to excellent outcomes, especially when embedded within a multidisciplinary approach.

Key words: Dysphagia, vascular anomalies, scoliosis.

Introduction

Dysphagia in young adults is often dismissed or attributed to benign gastrointestinal conditions. However, the intersection of subtle symptoms with unexpected anatomical findings can make for a challenging diagnostic journey. Esophageal obstruction secondary to extrinsic compression, while rare, demands a heightened index of suspicion – especially when routine investigations yield inconclusive results.

This case exemplifies one such diagnostic complexity. A young man, with well-documented endocrine and musculoskeletal co-morbidities, developed progressive difficulty swallowing alongside acute gastrointestinal distress. What began as a straight forward metabolic crisis evolved into an intricate investigation, with imaging ultimately revealing a congenital vascular anomaly previously unrecognised in the patient.

Surgery offered resolution for both an anatomical diagnosis and symptomatic relief, reinforcing the need to look beyond conventional causes in cases of persistent dysphagia. The following report delineates the clinical course, diagnostic process, and surgical management of this unusual presentation – reminding clinicians that sometimes, the clue lies just outside the esophageal wall.

Case

A 20-year-old man, previously in good health, presented with a three-month history of progressively worsening difficulty in swallowing, which initially affected solids more than liquids. Over the past three days, he also developed diffuse abdominal pain and recurrent episodes of vomiting occurring within minutes after eating. The vomiting was watery, occurred multiple times, and was not associated with blood. He had type 1 diabetes mellitus for the past six years, well controlled on insulin. His past medical history was also significant for congenital scoliosis and chronic gastritis associated with *Helicobacter pylori* infection. He consumed a mixed diet and had no history of substance abuse, and normal bladder and bowel habits with a preserved sleep cycle.

On examination, the patient's vital signs were stable, with a blood pressure of 102/60 mmHg, pulse rate of 100/min, respiratory rate of 14/min, oxygen saturation of 96% on room air, and a body temperature of 99° F. General and systemic examinations revealed no significant findings. Initial laboratory investigations, including complete blood count, liver and kidney function tests, serum electrolytes, thyroid profile, and coagulation parameters, were within normal limits. Cardiac markers, serum vitamin B12, and folate levels were also normal. However, his HbA1c was elevated at 8.72%, indicating suboptimal long-term glycaemic control. Additional findings included a mildly elevated urine ketone (1+), a metabolic acidosis pattern on arterial blood gas analysis, and a D-dimer level of 173 ng/mL with a fibrinogen level of 304 mg/dL. Echocardiography showed a preserved ejection fraction of 55 - 60% without structural abnormalities.

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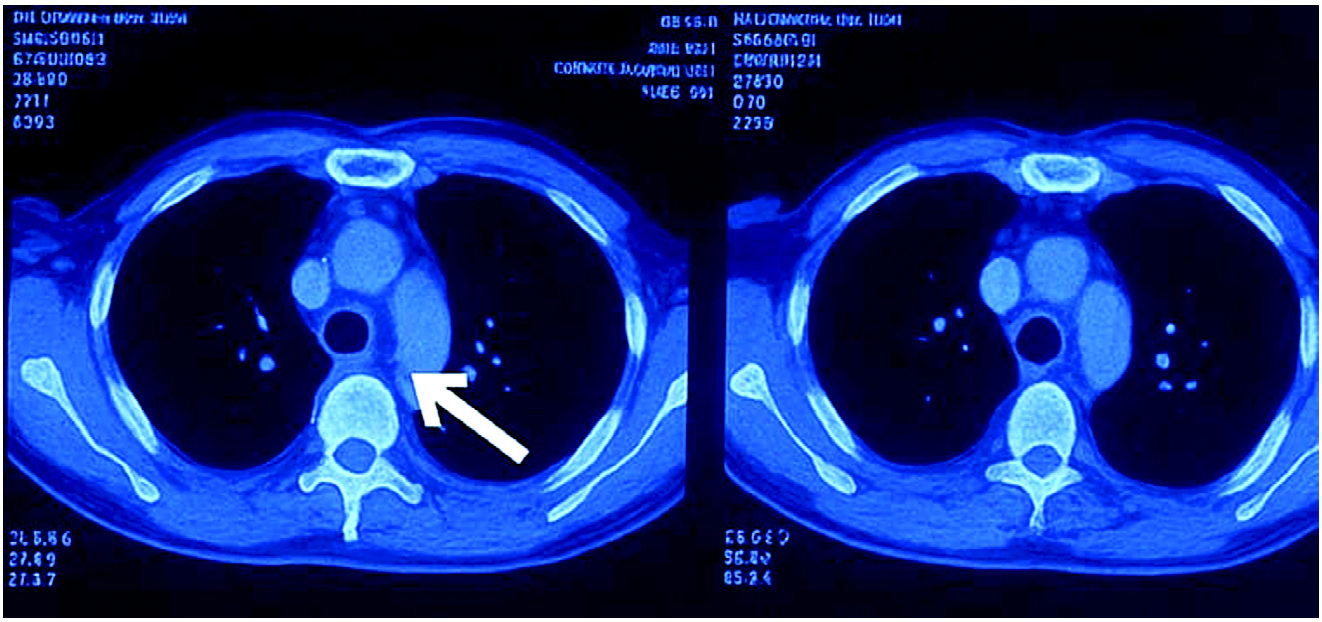

Fig. 1: *CT chest images showing the aberrant right subclavian artery.*

An abdominal ultrasound revealed a liver of normal size and texture, a normal portal vein, spleen, and gallbladder without biliary calculi or dilatation. A barium swallow demonstrated the classic "bird-beak" appearance, raising suspicion of a compressive esophageal pathology. Subsequent contrast-enhanced CT angiography confirmed the presence of an aberrant right subclavian artery (ARSA) coursing posterior to the esophagus, a vascular anomaly known to cause dysphagia lusoria.

Initially, the patient was managed conservatively for diabetic ketoacidosis with intravenous fluids and insulin therapy. Once metabolic parameters stabilised, surgical intervention was planned. Through an upper mini-sternotomy approach, the surgical team dissected the innominate vein and aortic arch

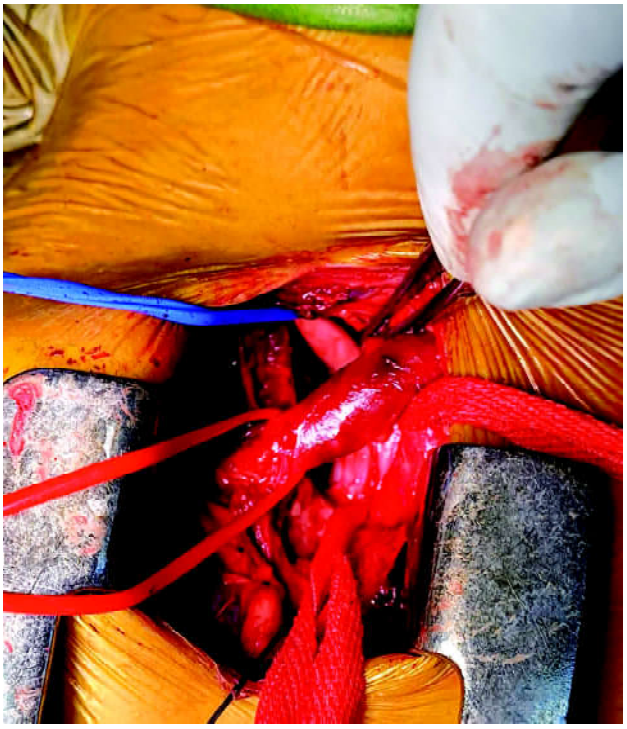

Fig. 2: *Vascular structures separated and aberrant artery identified.*

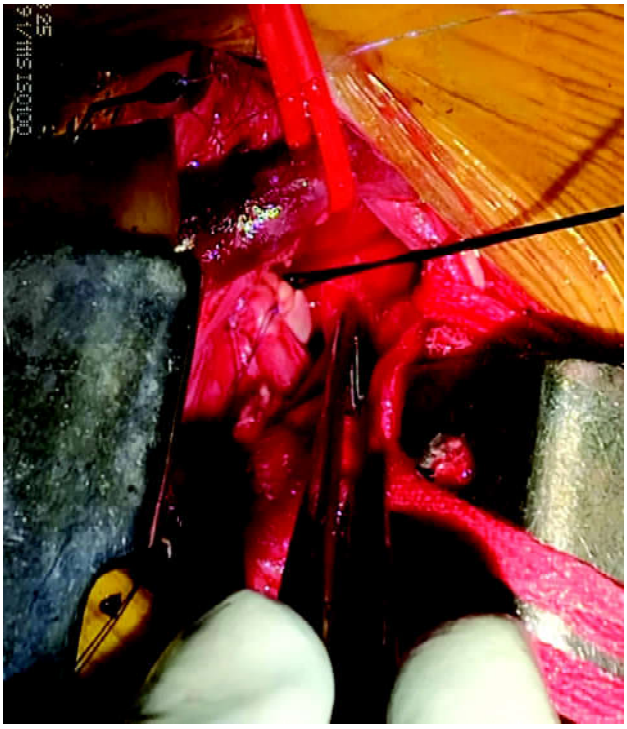

Fig. 3: *Artery was dissected and stump was made.*

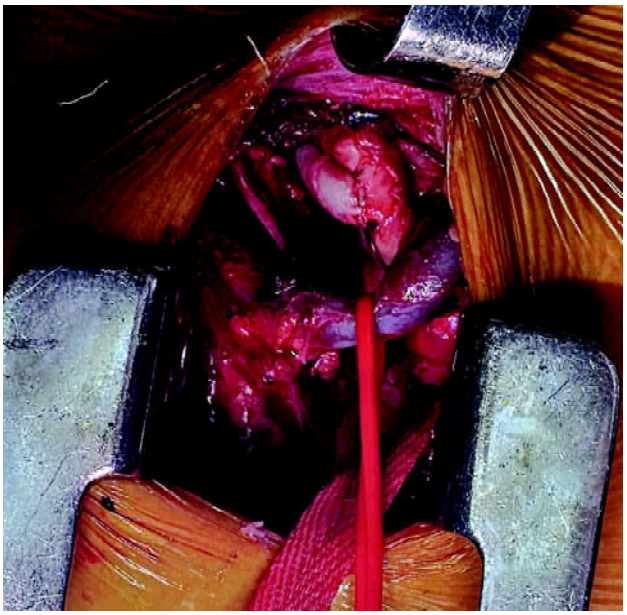

Fig. 4: *Aberrant artery was sutured to the Right Common Carotid Artery.*

vessels. The aberrant right subclavian artery was identified posterior to the esophagus, clipped, transfixed, and reimplanted anteriorly into the right common carotid artery using an end-to-side anastomosis. Hemostasis was secured, and the chest was closed with a right pleural drain in place.

This case exemplifies dysphagia lusoria, a rare but important vascular cause of esophageal compression, which should be considered in patients presenting with unexplained dysphagia. The diagnosis was clinched through characteristic imaging findings, and definitive surgical correction led to resolution of symptoms. The case also highlights the importance of managing co-existing metabolic derangements, such as diabetic ketoacidosis, before undertaking major surgical procedures.

Discussion

Dysphagia lusoria is a rare congenital vascular anomaly that results in difficulty swallowing (dysphagia) due to an aberrant course of the right subclavian artery. The term was first coined by David Bayford in 1761, who described a case of progressive dysphagia due to an unusual vascular structure, referring to it as a *"lusus naturae"* (freak of nature), hence the term "lusoria[1]". Bayford identified an aberrant right subclavian artery (ARSA) that traversed anterior to the esophagus, causing significant compression and ultimately contributing to emaciation and death in the patient he studied. This vascular anomaly is now broadly defined to include any congenital vascular abnormality that compresses the esophagus. The condition has an incidence of approximately 0.4 - 2% in the general population[2]. Symptoms often do not manifest until adulthood, usually triggered by atherosclerotic or aneurysmal changes in the aberrant vessel.

Embryologically, by the fifth week of fetal development, six aortic arches form. The left 4th aortic arch develops into the arch of the aorta and the proximal portion of the left subclavian artery. In contrast, the right 4th aortic arch, right dorsal aorta, and right 7th intersegment artery contribute to the formation of the right subclavian artery. If the distal part of the right dorsal aorta persists and the right 4th arch regresses, an aberrant right subclavian artery can form, typically passing behind the esophagus (retro-esophageal).

Dysphagia lusoria is sometimes associated with other congenital anomalies, often grouped under the acronym VACTERL[3]. This includes:

- V: Vertebral anomalies such as hemivertebrae or spina bifida.
- A: Anal atresia (absence or blockage of the anal opening).
- C: Cardiac defects, including VSD (ventricular septal defect), ASD (atrial septal defect), and PDA (patent ductus arteriosus).
- T: Tracheoesophageal fistula-an abnormal connection between the trachea and esophagus.
- E: Esophageal atresia-where the esophagus ends in a blind pouch.
- R: Renal anomalies like renal agenesis or vesicoureteral reflux (VUR).
- L: Limb anomalies such as radial or thumb aplasia and syndactyly.

Conclusion

Dysphagia lusoria, although uncommon, should remain a consideration in young patients presenting with unexplained dysphagia. Timely diagnosis through appropriate imaging modalities and prompt surgical correction can result in complete symptom resolution. This case underscores the critical role of a multidisciplinary approach and highlights the importance of addressing coexisting co-morbidities to optimise outcomes. The patient's co-existing scoliosis also raises the possibility of underlying syndromic associations, such as VACTERL. Accordingly, a high index of suspicion should be maintained for evaluating additional congenital anomalies in similar presentations.

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