Dysphagia Aortica: Diagnostic Dilemma and Therapeutic Paradigm

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Abstract

Objective: Intrinsic esophageal pathologies constitute prime cause for dysphagia clinically. However, thoracic esophageal domain is prone to extrinsic compression by various vascular afflictions including aneurysms with attendant therapeutic challenges. Herein, we present a case series of dysphagia aortica with emphasis on its appropriate management option based on grade of dysphagia.

Methods: Patients who presented to the vascular division of our tertiary care referral institute between January 2014 and October 2015 with dysphagia due to extrinsic esophageal compression by aneurysmal thoracic aorta form the basis for this report. Prior to referral, all patients were evaluated elsewhere to rule out intrinsic causes and computed tomography angiogram performed delineating aortic aneurysm in four patients and penetrating aortic ulcer in one. Patient cohort included one female and four male patients whose age ranged from 40 to 68 years, with a median of 62 years. Left posterolateral thoracotomy provided access to an aneurysm which was repaired using interposition polyester graft in four patients. Due to severe comorbidities which precluded open surgery, one patient who presented with mild dysphagia was managed by endovascular stent graft repair.

Results: Degree of dysphagia was assessed between grades 0 and 4 as in literature. All patients, including four open conventional and one endovascular, recovered well and left hospital totally symptom free.

Conclusion: Dysphagia due to vascular diseases in the thoracic domain is an uncommon clinical entity. Patients with thoracic aortic aneurysm presenting with severe dysphagia deserve open surgical repair to provide optimal symptomatic relief in addition to saving life. The state-of-the-art endovascular stent grafting may be considered in very elderly patient having severe comorbidities presenting with mild dysphagia.

Key Words: Dysphagia aortica, esophageal compression, penetrating aortic ulcer, thoracic aneurysm, thoracic endovascular aneurysm repair

Introduction

Dysphagia aortica, an uncommon clinical entity, results from mechanical extrinsic compression of the esophagus by dilated/tortuous or aneurysmal thoracic aorta. It is caused by either impingement of thoracic aortic aneurysm on the mobile upper thoracic esophagus or more commonly compression of the lower esophagus sandwiched between the large aortic aneurysm and the heart or rarely the ventral margin of the esophageal hiatus where it is anatomically restricted. The incidence of this presentation is neither known nor the management option standardized. We present a case series of dysphagia aortica with emphasis on its appropriate management option based on grade of dysphagia.

Methods

During the period between January 2014 and October 2015, five patients who reported to Division of Vascular Surgery of our tertiary referral hospital with dysphagia as an exclusive or predominant symptom form the basis for this report. All these patients had undergone extensive evaluation for dysphagia elsewhere including upper gastrointestinal (UGI) endoscopy and barium swallow, which ruled out intraluminal esophageal pathology. Thereafter, the thoracic aortic aneurysm was delineated on contrast-enhanced computed tomography (CECT) of the chest confirming cause-effect relationship of aneurysm and dysphagia.

Case reports

Case 1

A 68-year-old hypertensive male presented with progressive dysphagia associated with chest pain for 3 months. He was able to swallow only liquids. Investigations were suggestive of extrinsic compression of lower end of the esophagus by fusiform Type B descending thoracic

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aortic (DTA) aneurysm [Figure 1a]. Preoperative evaluation including coronary angiogram (CAG) was done, which was normal.

**Case 2**
A 62-year-old chronic smoker presented with severe chest pain and absolute dysphagia for 1 week. Non-CECT of the chest showed a large fusisaccular Type V thoracoabdominal aortic aneurysm (TAAA) causing severe narrowing of the lower end of the esophagus [Figure 1b]. He underwent an emergent repair due to impending rupture state of the aneurysm.

**Case 3**
A 62-year-old diabetic and hypertensive male presented with complaints of progressive dysphagia for 1 month. Preoperative UGI endoscopy and barium swallow were suggestive of extraluminal compression of the esophagus. CECT of the chest showed a penetrating aortic ulcer (PAU) at D9 vertebra level, with surrounding thrombus compressing and displacing the lower end of the esophagus [Figure 2]. The patient underwent preoperative evaluation including CAG which was normal.

**Case 4**
A 40-year-old female with no known comorbidities presented with complaints of dysphagia for solids for 2 months. On evaluation, a 6 cm sized saccular Type B DTA aneurysm compressing the esophagus was detected. She also had multiple hilar and cervical lymphadenopathy, biopsy of which was suggestive of tuberculosis. She was started on anti-tubercular therapy and surgery for an aortic aneurysm performed 3 months thereafter.

**Case 5**
A 59-year-old hypertensive male presented with chest pain and dysphagia for solids for 2 months. On evaluation, he was diagnosed to have double vessel coronary artery disease (CAD) and chronic liver disease (CLD) with deranged liver enzymes. CT of the chest showed Type B DTA aneurysm compressing the esophagus. Due to high risk for open surgery, he received endovascular aneurysm repair (thoracic endovascular aneurysm repair [TEVAR]).

**Management**
Although open surgery and endovascular procedures are feasible, we opted for open repair in all, except in one patient who had symptomatic DTA aneurysm and mild dysphagia associated with high comorbidity index including CAD and liver dysfunction (Case 5 [Table 1]) and was treated successfully by TEVAR. In open repair, all patients had a cerebrospinal fluid drain as a critical component of spinal cord protection strategy. Distal aortic perfusion was provided by custom made perfusion circuitry using temporary aorto femoral bypass and cell saver was used to retrieve the shed blood. After the proximal and distal clamps had placed, the distal flow was established by cannulating proximal aorta and left femoral artery. The flow was regulated by optimizing brachial pressure around 90 mm Hg mean pressure so that the mean arterial pressure in the femoral artery was around 60 mm Hg. Aneurysm was opened between clamps and the laminated thrombus within the aneurysmal sac was removed, except that near the esophagus to avoid its inadvertent injury. Thereafter, endoaneurysmorrhaphy was completed using appropriate sized collagen coated woven polyester graft (Intergard®, Maquet). Endoluminal repair in the patient with mild dysphagia with significant yet asymptomatic CAD/CLD was achieved by retrograde transfemoral deployment of 36 mm × 160 mm Medtronic Valiant® Captivia® stent graft successfully.

![Figure 1](image1.png)

**Figure 1:** (a) Computed tomography angiography axial section showing esophagus being sandwiched between the heart and descending thoracic aneurysm (white arrow). (b) Sagittal section of computed tomography angiography showing a large thoracoabdominal aortic aneurysm compressing and anteriorly displacing esophagus in a patient who presented with absolute dysphagia (yellow arrow).

![Figure 2](image2.png)

**Figure 2:** (a) Computed tomography angiography axial section showing a penetrating aortic ulcer of descending thoracic aorta with surrounding thrombus compressing the esophagus which appears slit-like (white arrow). (b) Preoperative barium swallow showing diffusely narrowed lower end of the esophagus due to extrinsic compression and is displaced anteriorly (white arrowhead). (c) Postoperative computed tomography angiography picture shows widely opened up esophagus (yellow arrow). (d) Postoperative barium swallow shows relief extrinsic compression with free passage of contrast into stomach without holdup (black arrowhead).
Pitchai, et al.: Dysphagia aortica

causes significant compression

Comorbidities

This entity was reported to be primarily seen in

Conventional

[16,17]

HTN

DM, HTN

[4,5]

[10,11]

[3]

[1]

Diagnosis

Type B DTA aneurysm

Penetrating aortic ulcer

Type B DTA aneurysm

Type B DTA aneurysm

[14,15]

Type B DTA

Open repair

Open repair

Open repair

TEVAR

Table 1: Clinical profile of patients

| Age | Sex | Comorbidities | Diagnosis* | Dysphagia Management grade* |
|-----|-----|---------------|------------|----------------------------|
| 68  | Male| HTN           | Type B DTA aneurysm | 3 - Open repair |
| 62  | Male| NIL           | Type V TAAA    | 4 - Open repair |
| 62  | Male| DM, HTN       | Penetrating aortic ulcer | 3 - Open repair |
| 40  | Female| NIL         | Type B DTA aneurysm | 2 - Open repair |
| 59  | Male| HTN, CAD, CLD | Type B DTA aneurysm | 1 - TEVAR |

*DTA aneurysm extent: Type A - LSCA to T6, Type B - T6 to diaphragmatic hiatus, Type C - LSCA to diaphragmatic hiatus. *Grades of dysphagia: 1 - Able to swallow solid foods with some difficulty, 2 - Able to swallow soft or semi-solid foods only, 3 - Able to swallow liquefied foods and liquids only, 4 - Unable to swallow liquids/saliva. LSCA: Left subclavian artery, HTN: Hypertension, DM: Diabetes mellitus, CAD: Coronary artery disease, CLD: Chronic liver disease, TAAA: Thoracoabdominal aortic aneurysm, DTA: Descending thoracic aortic, TEVAR: Thoracic endovascular aneurysm repairs

Results

All the patients made a satisfactory postoperative recovery and left the hospital in a weeks’ time. One patient developed surgical site infection and was managed conservatively. On follow-up which ranged between 6 and 20 months, all the patients remained symptom-free. Check CT angiogram performed during follow-up showed intact repair and well opened up the esophagus.

Discussion

Dysphagia aortica was first described by Pape in 1932 resulting from mechanical extrinsic compression of the esophagus by dilated/tortuous or aneurysmal thoracic aorta.[2] It was also observed that the esophageal compression by the aortic arch pathologies was relatively rare.[1] This entity was reported to be primarily seen in women and more associated with short stature, old age, and kyphosis.[3] Our cohort composed of predominantly male population who were relatively younger than the reported literature.

Etiology of dysphagia aortica ranges from an ectatic, tortuous aorta, and more importantly a large aneurysm involving the thoracic aorta like in our report. Esophagus being a hollow viscus within the thoracic cavity, it easily gets displaced by the enlarging aneurysm. However, in small subset of the patients, when it gets sandwiched between the large thoracic aortic aneurysm and heart, dysphagia occurs. Rarely, a PAU with surrounding thrombus can compress esophagus.[4,5]

Most of the time dysphagia aortica is not considered as a routine differential diagnosis for dysphagia, and hence, there is often a delay in diagnosis and treatment. If not treated on time, there is always a potential danger for aneurysmal rupture into the esophagus or pleural space since most of these patients have relatively larger aneurysm and an expeditious treatment is mandated.[6,7]

Due to rare incidence of dysphagia aortica with only a few cases reported in published literature, there is no standard treatment protocol described. Conservative management such as dietary modification (e.g., chewing small amounts of soft diet, avoiding food likely to stick into esophagus) and treatment of underlying heart failure and hypertension has been advised in mild dysphagia.[8,9] However, conservative approach is to be followed only in elderly patients with short life expectancy who are otherwise unfit for any intervention.

Open surgical repair of TAAA remains the gold standard treatment, and with advent of better surgical and anesthetic management, surgery could be performed with acceptable mortality and morbidity.[10,11] Conventional surgical repair entails aortic reconstruction with a graft of 20–26 mm appropriately (in our experience) along with removal of large amount of laminated thrombus relieving the extrinsic esophageal compression in toto. In this regard, special attention needs to be employed not to attempt total removal of laminated thrombus so as to avoid injury to already thinned out esophagus.

Even though TEVAR is largely replacing the open surgery due to its less invasiveness and low perioperative morbidity rates, its role in dysphagia aortica needs to be evaluated.[12] Two contemporary case reports of TEVAR in treating dysphagia aortica in literature suggested its safety. Kische et al. reported successful treatment of a 75-year-old patient with a giant PAU, managed endovascularly following which the patient had complete relief of the symptom.[4] Similarly, Siddiqui and Hughes reported on management of DTA aneurysm causing dysphagia in a patient by TEVAR.[13] The patient was completely relieved of his symptom only after 4 months following surgery. This is a particular issue with TEVAR because the aortic remodeling takes time after the procedure and the patient may not get immediate relief from the symptoms following TEVAR even though the direct pressure over the esophagus may decrease.[14,15] We have treated only one patient in our cohort reporting mild dysphagia by TEVAR, as asymptomatic double vessel CAD and CLD precluded open surgery.

However, TEVAR imposes significant risk of esophageal erosion by stent graft; the treatment whereof carries prohibitive mortality. One of the important proposed mechanisms of secondary aortoesophageal fistula is due to pressure necrosis caused by continuous expansile forces of oversized self-expanding stent graft on esophagus.[16,17] When aneurysm per se causes significant compression over esophagus with potential for luminal erosion sooner than later, an oversized stent graft expedites impending fatal hazard. We have had previous experience of fatal stent graft erosion into esophagus in a patient with severe
dysphagia who had a large DTA aneurysm in the past. Hence, we believe the optimal treatment remains open surgical which allows removal of laminated thrombus and thereby relieving extrinsic compression instantaneously.

We have tried to standardize the mode of managing these patients based on the clinical grade of dysphagia. For patients presenting with severe dysphagia (Grade 3/4), we suggest open surgical repair in most instances. For mild to moderate form of dysphagia (Grade 1/2), we suggest that either of the modality could be chosen in the context of patient’s fitness. In a surgically unfit patient and with mild dysphagia, TEVAR offers the best choice of treatment. However, we believe severe dysphagia due to thoracic aortic aneurysm precludes aortic stent graft procedure unlike contemporary clinical practice world over.

**Conclusion**

Dysphagia aortica is a rare clinical presentation of thoracic aortic aneurysm and prompt diagnosis with expeditious management is mandatory to avoid catastrophe. Our report is the first attempt to describe management option based on degree of dysphagia. Severe form of dysphagia aortica should be preferentially managed open surgically to relieve symptoms and prevent aneurysmal rupture. In very elderly patients with high morbidity index having mild dysphagia, TEVAR would be the judicious therapeutic strategy.

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**Conflicts of interest**

There are no conflicts of interest.

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