An Aberrant Right Subclavian Artery–Esophageal Fistula—A Fatal Complication of a Common Anomaly: A Case Report and Review of Literature

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Abstract

An aberrant right subclavian artery (ARSA), also called as arteria lusoria, is one of the most common aortic arch anomalies. ARSA–esophageal fistula is a rare, life-threatening complication, with only 37 cases reported in literature. We describe a case of a young girl who developed acute episode of massive hematemesis after the recovery from novel coronavirus disease 2019 (COVID-19) pneumonia. Computed tomography (CT) angiography showed ARSA with retroesophageal course and active contrast leak in esophagus. Digital subtraction angiography confirmed the site of active contrast extravasation from the ARSA. However, the patient succumbed to hypovolemic shock even before the endovascular or surgical interventions could be done.

Keywords
- ARSA
- aberrant right subclavian artery
- esophageal bleeding
- nasogastric tube

Introduction

An aberrant right subclavian artery (ARSA) with retroesophageal course is a common anomaly of the aortic arch.[1] Erosion of the esophagus due to pressure necrosis from prolonged indwelling objects (nasogastric tube and metallic stents), radiation therapy, esophageal carcinoma, or iatrogenic injury lead to the development of a fistula between the ARSA and the esophagus. Endovascular interventions, such as balloon occlusion, covered stent deployment, and coil embolization, can be life-saving, as hypovolemic shock precludes surgery in most patients.

Case Presentation

A 14-year-old girl was referred to a radiology department with sudden onset of massive hematemesis. She had history of quadriparesis and respiratory failure a month ago and was reverse-transcription polymerase chain reaction (RT-PCR)-positive for novel coronavirus disease 2019 (COVID-19). She had been intubated 18 days back with placement of nasogastric tube (NGT) 20 days before the hematemesis episode. Computed tomography (CT) angiography (►**Fig. 1**) showed ARSA with retroesophageal course with active extravasation of contrast into the esophagus.

The selective cannulation of ARSA was done using 5F Vert catheter (Cook Medical, Ireland) which revealed active, rapid extravasation of contrast into the esophagus, suggesting ARSA–esophageal fistula (►**Fig. 2**). A plan was made to perform a balloon occlusion followed by deployment of an covered stent across defect. Unfortunately, she went into cardiopulmonary arrest and could not be revived.

Discussion

The prevalence of an ARSA is 0.5 to 2% in the general population.[1] The occlusion of vasa vasorum of pressure necrosis and erosion of the esophagus lead to the formation of a fistula with the ARSA.[2] Prolonged endotracheal and/or

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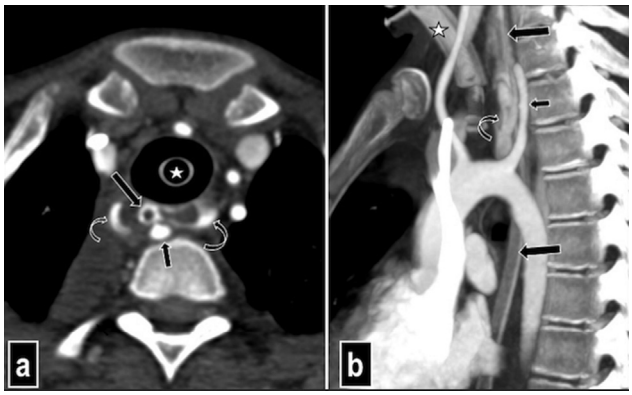

Fig. 1 **(a)** Axial and **(b)** sagittal CT angiography images showing the retroesophageal aberrant right subclavian artery (small arrows). Nasogastric tube (larger arrows) and tracheostomy tube with bulb (asterix) is seen in situ. There is contrast extravasation into the esophagus (curved arrows). CT, computed tomography.

nasogastric intubation is the most common predisposing factor for fistula formation. On literature review, we found a total of 17 cases of aberrant subclavian artery–esophageal fistula in which the prolonged endotracheal and/or nasogastric intubation has been listed as a causative factor of fistula formation[3–19] (►**Table 1**). The average duration between NGT and endotracheal tube (ET) placement and first episode of bleeding is of 29 (6–56) and 23 (13–31) days, respectively. In index case, NGT and ET placements were done 20 and 18 days back, respectively. The steroid use and secondary infection have also been reported as risk factors for the

Fig. 2 A digital subtraction angiogram performed in antero-posterior oblique projection showing the aberrant right subclavian artery (asterix) with arterioesophageal fistula (larger arrow) and contrast extravasation along the esophagus (curved arrows) close to nasogastric tube (arrow heads). Left subclavian artery is also visualized in same image (dashed arrow).

Table 1 Reported cases of aberrant subclavian artery–esophageal fistula due to prolonged use of nasogastric or endotracheal tube

Sr. no.	Study (year)	Age (y)/sex	Basic disease	ET/tracheostomy duration (d)	NGT duration (d)	Proposed risk factor for fistula	Endovascular treatment	Surgical treatment/esophageal ballooning	Follow-up	Outcome
1	Livesay et al[4] (1982)	25/M	Trauma	13	7	Inflated tracheostomy balloon + NGT	None	Repaired	2 weeks	Died
2	Jungck and Püschel[5] (1983)	6/M	Trauma	28	42	Inflated tracheostomy balloon + NGT	None	Esophageal balloon Thoracotomy	Same day	Died
3	Belkin et al[6] (1984)	27/M	Right retromolar carcinoma	No	56	Prolonged NGT	None	Esophageal balloon Ligation	10 days	Died
4	Edwards et al[7] (1984)	36/F	Cerebral aneurysm	Yes/NA	27	Prolonged NGT/steroid use/secondary infection	None	None	Same day	Died
5	Gossot et at[8] (1985)	72/F	Aortic repair	30	30	Prolonged NGT/ET/secondary infection	NA	NA	NA	Died

Table 1 (*Continued*)

Sr. no.	Study (year)	Age (y)/ sex	Basic disease	ET/ tracheostomy duration (d)	NGT duration (d)	Proposed risk factor for fistula	Endovascular treatment	Surgical treatment/ esophageal ballooning	Follow-up	Outcome
6	Guzzetta et al[9] (1989)	4 mo/F	Congenital heart disease and its repair	28	56	Prolonged NGT	None	Ligation	14 weeks	Died
7	Ikeda et al[10] (1991)	9/M	Congenital heart disease	Yes/NA	Yes/NA	Prolonged NGT	NA	NA	NA	Died
8	Hirakata et al[11] (1991)	55/M	Esophagus carcinoma surgery	(NA)	44	Prolonged NGT, Radiation enteritis, surgical trauma	Ballooning	Ligation	NA	Survived
9	Miller et al[12] (1996)	11/F	Intraventricular bleed	14	17	Prolonged NGT/ET	None	Esophagus balloon Ligation	2 years	Survived
10	Minyard and Smith[13] (2000)	39/F	Head trauma	NA	6	NGT	None	None	6 days	Died
11	Feugier et al[14] (2003)	24/M	Polytrauma	31	31	Prolonged NGT and ET	Ballooning	Ligation	4 month	Survived
12	Chapman et al[15] (2010)	34/F	NA	Yes/NA	Yes/NA	Prolonged NGT	Ballooning	Ligation	NA	Died
13	Jain et al[16] (2012)	57/M	Scimitar syndrome	18	18	Prolonged NGT and ET	Coiling	Esophageal balloon Ligation	3 weeks	Survived
14	Oliveira et al[3] (2016)	20/M	Trauma	22	22	Prolonged NGT and ET	None	Ligation	6 weeks	Survived
15	Kudose et al[17] (2017)	20/M	VATER Status lung transplant	Yes/NA	Yes/NA	Prolonged NGT and ET	None	None	Same day	Died
16	Shires and Rohrer[18] (2018)	41/M	Pneumonia	Yes/NA	16	NGT and ET	Stenting	None	Same day	Died
17	Kim et al[19] (2021; ALSA)	63/M	Intracranial Bleed	NA	Yes/NA	NGT, biopsy	TEVAR Coiling	None	2 months	Died
18	Index case (2021)	14/F	Porphyria COVID-19 pneumonia	18	20	NGT/ET/COVID-19	None	None	Same day	Died

Abbreviations: ALSA, aberrant left subclavian artery; COVID-19, novel coronavirus disease 2019; ET, endotracheal tube; F, female; M, male; NA, data not available; NGT, nasogastric tube; VATER (VACTERL), vertebrae, anus, heart, trachea, esophagus, kidney and limbs.

fistula formation.[7] The index patient did not received steroids in her course in the hospital. The COVID-19 infection has been shown to have more risk of development and rupture of pseudoaneurysms due to endothelial inflammation.[20] In few cases, initial "alarming" episodes of minimal bleeding have also been reported.[7,9,10,19,21]

ARSA can be visualized on CT angiography and can be confirmed on conventional angiography. Placement of esophageal Sengstaken–Blakemore tube can help in temporary control of bleeding.[6] Surgical options include ligation of the subclavian artery with revascularization of the right arm.[14] In endovascular approach, angioplasty balloon can be inflated across the fistulous segment as a temporary measure before the definitive surgery.[14] More recently, successful usage of covered stents as a definitive measure has been described.[18] Despite all attempts at management, the reported overall survival rate of ARSA–esophageal fistula is only 35.7%.[17] In present literature review of NGT or ET, the overall survival found to be 29.4% as a cause of the fistula formation. Out of these 17 cases, four patients died on the same day of bleeding episode.[5,7,17,18] Therefore, it is very important to recognize and manage this fatal condition as soon as possible. The authors also recommend to avoid prolonged nasogastric tube placement in patient with aberrant subclavian artery.

Conclusion

The fistulization of ARSA into the esophagus is a rare and lethal complication and may be seen in patients with prolonged nasogastric or endotracheal intubation. A high index of suspicion and careful evaluation of radiological imaging is required in its timely recognition and treatment.

Authors' Contributions

Conception and design, acquisition of data, and analysis and interpretation of data: P.C.S., N.C., R.S., H.B., and N.P.

Literature search, drafting the manuscript, and revising it critically for important intellectual content: R.S., H.B., N. P., and N.C.

Manuscript editing and final approval of the versions to be published: H.B., N.P., and R.S.

Conflict of Interest

None declared.

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