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.

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 showed ARSA with retroesophageal course and 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. 1). 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...
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. 1](https://example.com/f1.png) \(\text{Fig. 1} \) (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.

![Fig. 2](https://example.com/f2.png) \(\text{Fig. 2} \) 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>
<thead>
<tr>
<th>Sr. no.</th>
<th>Study (year)</th>
<th>Age (y)/sex</th>
<th>Basic disease</th>
<th>ET/ tracheostomy duration (d)</th>
<th>NGT duration (d)</th>
<th>Proposed risk factor for fistula</th>
<th>Surgical treatment</th>
<th>Endovascular treatment for fistula</th>
<th>Follow-up</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Livesay et al (1982)</td>
<td>25/M</td>
<td>Trauma</td>
<td>13</td>
<td>7</td>
<td>Inflated tracheostomy balloon + NGT</td>
<td>NA</td>
<td>NA</td>
<td>2 weeks</td>
<td>Died</td>
</tr>
<tr>
<td>2</td>
<td>Jungck and Puschel (1983)</td>
<td>6/M</td>
<td>Trauma</td>
<td>28</td>
<td>42</td>
<td>Inflated tracheostomy balloon + NGT</td>
<td>NA</td>
<td>NA</td>
<td>Same day</td>
<td>Died</td>
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<tr>
<td>4</td>
<td>Edwards et al (1984)</td>
<td>36/F</td>
<td>Cerebral aneurysm</td>
<td>Yes/NA</td>
<td>27</td>
<td>Prolonged NGT/steroid use/secondary infection</td>
<td>NA</td>
<td>NA</td>
<td>Same day</td>
<td>Died</td>
</tr>
<tr>
<td>5</td>
<td>Cours et al (1985)</td>
<td>72/F</td>
<td>Aortic repair</td>
<td></td>
<td>30</td>
<td>Prolonged NGT/ET</td>
<td>NA</td>
<td>NA</td>
<td>Died</td>
<td>Died</td>
</tr>
</tbody>
</table>

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

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<th>NGT duration (d)</th>
<th>Proposed risk factor for fistula</th>
<th>Endovascular treatment</th>
<th>Surgical treatment/esophageal balloononing</th>
<th>Follow-up</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>7</td>
<td>Ikeda et al(^10) (1991)</td>
<td>9/M</td>
<td>Congenital heart disease</td>
<td>Yes/NA</td>
<td>Yes/NA</td>
<td>Prolonged NGT</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>Died</td>
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<tr>
<td>8</td>
<td>Hirakata et al(^11) (1991)</td>
<td>55/M</td>
<td>Esophagus carcinoma surgery</td>
<td>(NA)</td>
<td>44</td>
<td>Prolonged NGT, Radiation enteritis, surgical trauma</td>
<td>Ballooning</td>
<td>Ligation</td>
<td>NA</td>
<td>Survived</td>
</tr>
<tr>
<td>9</td>
<td>Miller et al(^12) (1996)</td>
<td>11/F</td>
<td>Intraventricular bleed</td>
<td>14</td>
<td>17</td>
<td>Prolonged NGT/ET</td>
<td>None</td>
<td>Esophagus balloon Ligation</td>
<td>2 years</td>
<td>Survived</td>
</tr>
<tr>
<td>10</td>
<td>Minyard and Smith(^13) (2000)</td>
<td>39/F</td>
<td>Head trauma</td>
<td>NA</td>
<td>6</td>
<td>NGT</td>
<td>None</td>
<td>None</td>
<td>6 days</td>
<td>Died</td>
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<tr>
<td>11</td>
<td>Feugier et al(^14) (2003)</td>
<td>24/M</td>
<td>Polytrauma</td>
<td>31</td>
<td>31</td>
<td>Prolonged NGT and ET</td>
<td>Ballooning</td>
<td>Ligation</td>
<td>4 month</td>
<td>Survived</td>
</tr>
<tr>
<td>12</td>
<td>Chapman et al(^15) (2010)</td>
<td>34/F</td>
<td>NA</td>
<td>Yes/NA</td>
<td>Yes/NA</td>
<td>Prolonged NGT</td>
<td>Ballooning</td>
<td>Ligation</td>
<td>NA</td>
<td>Died</td>
</tr>
<tr>
<td>13</td>
<td>Jain et al(^16) (2012)</td>
<td>57/M</td>
<td>Scimitar syndrome</td>
<td>18</td>
<td>18</td>
<td>Prolonged NGT and ET</td>
<td>Coiling</td>
<td>Esophageal balloon Ligation</td>
<td>3 weeks</td>
<td>Survived</td>
</tr>
<tr>
<td>14</td>
<td>Oliveira et al(^3) (2016)</td>
<td>20/M</td>
<td>Trauma</td>
<td>22</td>
<td>22</td>
<td>Prolonged NGT and ET</td>
<td>None</td>
<td>Ligation</td>
<td>6 weeks</td>
<td>Survived</td>
</tr>
<tr>
<td>15</td>
<td>Kudose et al(^17) (2017)</td>
<td>20/M</td>
<td>VATER Status lung transplant</td>
<td>Yes/NA</td>
<td>Yes/NA</td>
<td>Prolonged NGT and ET</td>
<td>None</td>
<td>None</td>
<td>Same day</td>
<td>Died</td>
</tr>
<tr>
<td>16</td>
<td>Shires and Rohrer(^18) (2018)</td>
<td>41/M</td>
<td>Pneumonia</td>
<td>Yes/NA</td>
<td>16</td>
<td>NGT and ET</td>
<td>Stenting</td>
<td>None</td>
<td>Same day</td>
<td>Died</td>
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<tr>
<td>17</td>
<td>Kim et al(^19) (2021; ALSA)</td>
<td>63/M</td>
<td>Intracranial Bleed</td>
<td>NA</td>
<td>Yes/NA</td>
<td>NGT, biopsy</td>
<td>TEVAR Coiling</td>
<td>None</td>
<td>2 months</td>
<td>Died</td>
</tr>
<tr>
<td>18</td>
<td>Index case (2021)</td>
<td>14/F</td>
<td>Porphyria COVID-19 pneumonia</td>
<td>18</td>
<td>20</td>
<td>NGT/ET/COVID-19</td>
<td>None</td>
<td>None</td>
<td>Same day</td>
<td>Died</td>
</tr>
</tbody>
</table>

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.
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fistula formation. The index patient did not receive 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. In few cases, initial “alarming” episodes of minimal bleeding have also been reported.

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. Surgical options include ligation of the subclavian artery with revascularization of the right arm. In endovascular approach, angioplasty balloon can be inflated across the fistulous segment as a temporary measure before the definitive surgery. More recently, successful usage of covered stents as a definitive solution has been reported. Despite all attempts at management, the reported overall survival rate of ARSA–esophageal fistula is only 35.7%. 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. 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 R.S.

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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References