BRIEF COMMUNICATION

Upper gastrointestinal bleeding secondary to an aberrant right subclavian artery-esophageal fistula: A case report and review of the literature

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An aberrant right subclavian artery (ARSA) is a common aortic arch abnormality. A case of a 57-year-old man presenting with melena and hypotension secondary to an ARSA-esophageal fistula is reported. The current report is unique because it is the first reported case of ARSA-esophageal fistula associated with prior esophagectomy and gastric pull-up. A MedLine search was performed for ARSA-esophageal fistula cases, which were then compared with the present case. Because this patient had no vascular conduits, nasogastric or endotracheal tubes, the fistula likely occurred secondary to the previous surgery. This case is unusual because the patient survived the original hemorrhage associated with the ARSA-esophageal fistula. An ARSA-esophageal fistula is a rare, but potentially fatal cause of upper gastrointestinal bleeding. A high index of suspicion is needed to make the diagnosis. This condition should be considered in patients with risk factors combined with hemodynamically significant gastrointestinal bleeding.

Key Words: Aberrant right subclavian artery; Esophagectomy; Gastric pull-up; Gastrointestinal bleeding; Subclavian artery-esophageal fistula

A 57-year-old man with a T4 N0 esophageal adenocarcinoma presented with melena and hypotension secondary to a fistula esophago-ASDCA. He s'agit d’un cas unique en son genre du fait qu’il se rapporte à une fistule esophago-ASCDA associée à une esophagectomie et à une remontée de l’estomac pratiquées antérieurement. Nous avons effectué une recherche dans la base de données Medline sur les cas de fistule oesophago-ASCDA, puis nous avons comparé avec le présent cas. Comme le patient n’avait pas de greffon vasculaire, de sonde nasogastrique ou de tube endotrachéal, la fistule était probablement consécutive à la chirurgie antérieure. Il s’agit également d’un cas inhabituel parce que le patient a survécu à l’hémorragie de départ, associée à la fistule oesophago-ASCDA. Ce genre de fistule est une cause rare mais potentiellement mortelle d’hémorragie digestive haute. La pose du diagnostic exige un indice de suspicion élevé. L'anomalie devrait être envisagée chez les patients qui présentent des facteurs de risque associés à une hémorragie digestive accompagnée de fortes répercussions hémodynamiques.

CASE PRESENTATION

A 57-year-old man with a T4 N0 esophageal adenocarcinoma found at the gastroesophageal junction with no lymph node involvement was treated with an esophagectomy and gastric pull-up. Two years following the surgery, he developed metastases to his scalp and lungs which were treated by resection combined with a chemotherapy regimen of docetaxel (Taxotere, sanofi-aventis Canada Inc) and etoposide (VePesid, Bristol-Myers Squibb, Canada). At the end of therapy, another metastasis was identified in the patient’s left cerebellum. The patient had no other significant medical conditions and was not taking any medications.

Three years following the original surgery, he presented to the emergency department with upper gastrointestinal (GI) bleeding. He was admitted with a one-week history of
nausea, melena and blood-tinged sputum. His blood pressure was 120/70 mmHg, with a heart rate of 84 beats/min. His initial hemoglobin level was 135 g/L.

The gastroenterology service was not consulted initially, because there was a question of whether his melena represented hemoptysis and swallowed blood. By the fourth day of admission, the patient's hemoglobin level had dropped to 111 g/L. The gastroenterology service was consulted and an esophagogastroduodenoscopy (EGD) was performed. A small anastomotic ulcer at a distance of 25 cm from the incisors was visualized (Figure 1A). The ulcer was not actively bleeding and the patient was injected with 5 mL of adrenaline (1:1000 adrenaline in 10 mL of normal saline). After the presumptive diagnosis of the anastomotic ulcer was made, the patient was given lansoprazole 30 mg twice a day. No further rebleeding occurred, and he was later discharged home.

One week following discharge, the patient once again presented to the emergency department with melena, hypotension (blood pressure of 80/30 mmHg), and a hemoglobin level of 67 g/L. A follow-up EGD (Figure 1B) showed the same lesion at a distance of 25 cm from the incisors. There was no overlying clot or active bleeding; however, fresh red blood was visualized in the stomach. There was no evidence of local recurrence of the original tumour.

A contrast-enhanced computed tomography scan demonstrated an ARSA in paper-thin contact with the site of the original gastric pull-up procedure. No contrast medium was visualized in the intestinal lumen. A subsequent thoracic aortic angiogram confirmed the presence of an ARSA-esophageal fistula. Figure 1C shows the ARSA with aneurysmal dilation of its base.

Following the computed tomography scan, the patient was admitted to the intensive care unit for stabilization before surgery. While in the intensive care unit, the patient was transfused three units of packed red blood cells to raise his hemoglobin level to 77 g/L. Two days later, the patient was alert and oriented, with a blood pressure of 140/70 mmHg and a heart rate of 80 beats/min. A left thoracotomy was performed to ligate the ARSA at its origin. This was followed by a right carotid-subclavian artery bypass graft.

Five days postoperatively, the patient developed further upper GI bleeding with hematemesis and hypotension. An EGD showed oozing from the original fistula site. An urgent angiogram was performed. The aortic arch arteriogram revealed recanalization of the aneurysmal stump of the ARSA from a pinhole orifice (Figure 1D). This was thought to represent recanalization of the ARSA due to ligature dehiscence.

The origin of the ARSA was catheterized and embolized with occluding coils, achieving immediate control of the bleeding. Figure 1E shows satisfactory ablation of the aneurysmal stump of the ARSA with the coils.

Ten days postoperatively, the patient began to complain of dysphagia, along with recurring melena. EGD was once again performed, showing coils from the embolization procedure protruding into the esophageal lumen (Figure 1F). In addition to these findings, old blood was visualized in the stomach, suggesting ongoing GI bleeding related to reconstitution of flow in the ARSA. A left thoracotomy was performed, during which the coils from the embolization procedure were removed. The ARSA was resected. On postoperative day 18, the patient bled again, resulting in sudden hypotension, which led to subsequent cardiac arrest and death.

At autopsy, a mycotic aneurysm was identified 1.5 cm distal to the aortic arch. This aneurysm had ruptured into the adjacent left pleural cavity, thereby causing exsanguination. This bleeding site was independent of the original ARSA-esophageal fistula. This mycotic aneurysm was not present in previous studies, suggesting that it had developed in the postoperative period.

In addition to the aneurysm, metastases of the original tumour were documented in the right lung (measuring 1.5 cm $\times$ 1.0 cm), scalp, cerebellum and right kidney (3.5 cm...
× 3.0 cm). However, no local recurrence of the tumour was apparent at the site of anastomosis. Generalized atherosclerosis was also identified on autopsy, including mild atherosclerotic disease in the proximal region of the ARSA. The presence of the atheroma was not believed to play a significant role in the fistula formation.

**DISCUSSION**

The age of patients affected with ARSA-esophageal fistulas varies greatly. A literature search found 13 documented cases. Patients ranged from nine to 82 years of age (Table 1). Among these 13 patients, six had fistulas that occurred secondary to ARSA aneurysm, eight involved NG tubes, six had ET tubes and two of the patients had fistulas with a vascular conduit present. Among the patients with NG tubes, the duration of intubation varied greatly, ranging from nine to 60 days. NG and ET tubes are believed to contribute to fistula formation due to the creation of pressure necrosis where the tubes make contact with the esophageal wall (10,11).

Our case is unique in that it is the first reported case of ARSA-esophageal fistula associated with prior esophagectomy and gastric pull-up. In contrast to many of the previously documented cases of ARSA-esophageal fistula, the patient in the present study had no vascular conduit, NG or ET tubes. Therefore, its is presumed that the fistula was formed secondary to pressure from the gastric pull-up procedure. It is also possible that a foreign body, such as a suture or a staple at the anastomosis site, was partly responsible for the fistula formation.

The present case was also atypical in that the patient survived the hemorrhage associated with the formation of the ARSA-esophageal fistula. Unfortunately, as previously mentioned, he died from subsequent mycotic aneurysmal bleeding.

Aortoesophageal fistulas can develop postesophagogastrectomy in patients with normal aortic anatomy. In a literature review (12) of 500 cases of aortoesophageal fistula, 15 were presumed to be due to prior esophagogastrectomy. All 15 of these cases occurred between 12 and 40 days postoperatively. Therefore, it is unusual that in the present case it was almost three years following surgery before fistula formation occurred. The reason for this delay remains unclear. Among the 500 reported cases of aortoesophageal fistula, three were believed to be due to atherosclerosis. In all of these instances, ulceration of the atheroma was evident (12). This was not the case with our patient, as was evidenced at the time of autopsy.

The present case illustrates the difficulty of diagnosing an ARSA-esophageal fistula. In aortoesophageal fistulas, sentinel bleeding occurs in up to 63% of cases (12). However, only one of the cases of ARSA-esophageal fistula in the literature involved clear sentinel bleeding. Therefore, our case is unusual in that clear sentinel bleeding did occur. Despite the low rate of sentinel bleeding in patients with ARSA-fistulas, in our patient, consideration of a vascular-enteric fistula should have been given at first presentation.

In cases of ARSA-esophageal fistula bleeding, EGD can be used to rule out other causes of GI bleeding. In only two of 13 cases the patients were stable enough to allow EGD. One of these was performed in the operating room in anticipation of surgery. In one of these cases, EGD allowed localization of the bleeding to the esophagus.

**TABLE 1**

*Summary of reported cases of aberrant right subclavian artery (ARSA)-esophageal fistula*

<table>
<thead>
<tr>
<th>References</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Diagnosis</th>
<th>ET Tube</th>
<th>NG Tube</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lynn (4)</td>
<td>M</td>
<td>57</td>
<td>ARSA aneurysm</td>
<td>N</td>
<td>N</td>
<td>Died</td>
</tr>
<tr>
<td>Reyes (5)</td>
<td>F</td>
<td>72</td>
<td>ARSA aneurysm</td>
<td>N</td>
<td>N</td>
<td>Died</td>
</tr>
<tr>
<td>Merchant et al (10)</td>
<td>F</td>
<td>17</td>
<td>Postcaesarean section with NG tube</td>
<td>N</td>
<td>9 days</td>
<td>Died</td>
</tr>
<tr>
<td>Livesay et al (11)</td>
<td>M</td>
<td>25</td>
<td>Post-MVA with ET and NG tubes</td>
<td>Y</td>
<td>13 days</td>
<td>Died</td>
</tr>
<tr>
<td>Belkin (13)</td>
<td>M</td>
<td>27</td>
<td>Metastatic squamous cell CA with NG tube</td>
<td>Y</td>
<td>60 days</td>
<td>Died</td>
</tr>
<tr>
<td>Edwards and Edwards (6)</td>
<td>M</td>
<td>79</td>
<td>ARSA aneurysm</td>
<td>N</td>
<td>N</td>
<td>Died</td>
</tr>
<tr>
<td>Gossot et al (9)</td>
<td>F</td>
<td>72</td>
<td>Aortic graft with ET and NG tubes</td>
<td>Y</td>
<td>30 days</td>
<td>Died</td>
</tr>
<tr>
<td>Kulnig (7)</td>
<td>M</td>
<td>66</td>
<td>ARSA aneurysm</td>
<td>N</td>
<td>N</td>
<td>Died</td>
</tr>
<tr>
<td>Stone et al (2)</td>
<td>M</td>
<td>72</td>
<td>Thoracic aorta graft with ET tube</td>
<td>Y</td>
<td>Unknown</td>
<td>Died</td>
</tr>
<tr>
<td>Ikeda et al (14)</td>
<td>M</td>
<td>9</td>
<td>Post-Fontan with ET and NG tubes</td>
<td>Y</td>
<td>‘Long’</td>
<td>Died</td>
</tr>
<tr>
<td>Miller et al (1)</td>
<td>F</td>
<td>11</td>
<td>Postop for CNS bleeding with ET and NG tubes</td>
<td>Y</td>
<td>17 days</td>
<td>Lived</td>
</tr>
<tr>
<td>Feugier et al (3)</td>
<td>M</td>
<td>24</td>
<td>Trauma with ET and NG tubes</td>
<td>Y</td>
<td>31 days</td>
<td>Lived</td>
</tr>
<tr>
<td>Singh et al (8)</td>
<td>M</td>
<td>82</td>
<td>ARSA aneurysm</td>
<td>N</td>
<td>N</td>
<td>Died</td>
</tr>
</tbody>
</table>

CA Carcinoma; CNS Central nervous system; ET Endotracheal; F Female; M Male; MVA Motor vehicle accident; N No; NG Nasogastric; Postop Postoperative; Y Yes

Only two patients have been reported to have survived bleeding from ARSA-esophageal fistulas. In both cases, the patients survived because a timely diagnosis was made, thereby allowing immediate repair in the operating room.

Surgery should always be considered in any patient diagnosed with an ARSA-esophageal fistula. As evidenced in the present case, embolization may not always provide adequate hemostasis; in some cases it may create further undesirable complications. Endoluminal stent grafts have largely replaced the embolization procedure used in the present case. These are plastic tubes made of a polytetrafluoroethylene resin with a pliable metallic skeleton (stent). They are deployed in the aorta to completely cover the orifice from inside.

**ARSA-esophageal fistula** is a very rare, but potentially fatal cause of upper GI bleeding. Bleeding is often sudden and massive. The fact that this condition is so rare and difficult to identify means that a high index of suspicion is needed to make the diagnosis. An ARSA-esophageal fistula should be considered in patients with risk factors combined with hemodynamically significant GI bleeding. As evidenced in the present case report, prior esophageal surgery should be considered as a risk factor in the development of an ARSA-esophageal fistula.
REFERENCES
