A Fatal Case of Ruptured Aberrant Left Subclavian Artery into the Oesophagus

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Abstract

A right-sided aortic arch is a rare congenital abnormality and could be sometimes associated with an aberrant left subclavian artery which runs behind the oesophagus. It is usually incidentally diagnosed during adulthood unless it gives pressure effects on the adjacent structures. Here we present a case of 35-year-old mother with a period of amenorrhea of 9 weeks who scammed to death due to massive bleeding following ruptured aberrant left subclavian artery into the oesophagus.

Keywords: Aberrant left subclavian artery, oesophageal fistula, massive bleeding

Introduction

A right sided aortic arch is a rare congenital abnormality with an incidence of 0.1% and could be sometimes associated with an aberrant left subclavian artery which runs behind the oesophagus.[1] It is assumed to be due to the persistence of the right fourth embryologic aortic arch complicated with involution of the left aortic arch. Usually, the left subclavian artery (LSA) arises directly from the aortic arch, but it could arise from a diverticulum known as 'Kommerel’ giving rise to an aberrant LSA (Figure 1).

Case report

A 35-year-old mother with a period of amenorrhoea (POA) of 9 weeks presented with vomiting and sleepiness for 2 days. She had mild neck stiffness with low-grade fever, but both computed tomography of brain and lumbar puncture were normal. Her white cell count was 9×10³ with lymphocytic predominance. A clinical diagnosis of viral encephalitis was made. On the 5th day of admission, she developed a prolong seizure which led into intubation and mechanical ventilation. On the 9th day of admission, she was extubated, and clinical improvement was remarkable. While she was continuing to receive her antiviral therapy on the 11th day of admission, she developed profound haematemesis. A massive transfusion protocol was activated. Despite vigorous resuscitation she succumbed to death.

Autopsy revealed an oedematous body, pale conjunctivae, and multiple cutaneous ecchymosis with evidence of bleeding from orifices. No external or internal injuries were noted. Oesophagus was...
grossly swollen with clotted blood up to the pharynx. The stomach and bowels were grossly distended with blood (Fig. 2). Approximately 2 Liters of blood were recovered from the stomach alone.

A right-sided aortic with aberrant LSA behind the oesophagus at the level of 4th thoracic (T4) vertebrae noted making a band (Fig. 3).

A tear at its origin complicated with a fistula formation to the mid-oesophagus was identified (Fig. 4). Sub-endocardial haemorrhages and swollen kidneys with congested medulla noted. Histology revealed perivascular cuffing with lymphocytes associated with oedema. A foetus compatible with POA was observed without any gross abnormality.

Discussion
The aorta begins to develop during week four of gestation and its arches undergo sequential maturation with fusions, migrations, and involutions. Failure in this complex process give rise
to congenital anomalies as seen in this case.[4] Here the left subclavian, which was aberrant in nature has ruptured into the esophagus causing massive bleeding. Pallor organs, pools of internal blood, shock hemorrhages in the left ventricular myocardium supports the diagnosis of hemorrhagic shock.[5]

Several risk factors for the rupture of aberrant left subclavian artery have been identified in previously published cases. Aberrant nature and aortic arch being right sided are strongly associated with rupture and both factors are seen here. Other than those, coarctation of aorta, hypertension, neurofibromatosis, and advanced pregnancy have been described in the literature which were not evident in this case.[6,7]

The mechanism of formation of fistula between aberrant LSA and esophagus has been attributed to conditions that induce chronic irritation. Commonly recognized factors include prolong use of the nasogastric tube and endotracheal tube.[8,9] However in this case the woman was not on a nasogastric tube throughout her hospital stay and although she was intubated, it was only for a period of 4 days which unlikely to exert chronic pressure leading to necrosis and ulceration. Furthermore, there was no recent history of previous hospital admissions, and no history of chronic medical disease was reported. Therefore, the etiology of the formation of the fistula in this case is not clearly understood.

The manner of rupture appears to be spontaneous with negative external and internal injuries or circumstantial evidence of trauma. Evidence of viral encephalitis could have been contributory, which is the likely cause for sleepiness and vomiting, but the pregnancy is unlikely since it is during the peripartum that typically gives rise to hyperdynamic circulation and elevated blood pressure due to the gravid uterus.[10]

**Conclusions**

An aberrant left subclavian artery with a right-sided aortic arch, an extremely uncommon congenital vascular anomaly, may remain entirely asymptomatic unless it exerts significant pressure effects on the surrounding anatomical structures. Once rupture occurs, it can result in calamitous bleeding, potentially leading to exsanguination and death, even with aggressive management.

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**References**