

A Fatal Aorto-esophageal Fistula Caused by Critical Combination of Double Aortic Arch and Nasogastric Tube Insertion for Superior Mesenteric Artery Syndrome

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Key Words

Aorto-esophageal fistula · Double aortic arch · Vascular ring · Nasogastric tube · Superior mesenteric artery syndrome

Abstract

Double aortic arch (DAA) is a rare vascular congenital abnormality. Since a vascular ring surrounds bronchus and esophagus, any oral or nasal intubation can physically cause fatal aorto-esophageal fistula (AEF). We report herein the first case of association of DAA and superior mesenteric artery (SMA) syndrome and the second case of AEF caused by nasogastric intubation in an adult with DAA. A 19-year-old woman visited our hospital for nausea and vomiting. She was diagnosed with SMA syndrome by computed tomography (CT). Nasogastric intubation relieved her symptoms in 4 days. Extramural compression with top ulceration was found in esophagogastroduodenoscopy on the 5th hospital day. She suddenly showed massive hematemesis on the 12th hospital day. AEF was found by CT. Soon, she died despite of intensive care. Retrospective interview disclosed the fact that DAA was pointed out in her childhood. We conclude that intubation must be avoided in DAA and a detailed clinical interview about DAA is mandatory to avoid AEF.

Introduction

Nasogastric tube is one of the most useful devices in medical practice. It is useful in feeding, drainage, etc. But the combination of nasogastric tube and double aortic arch (DAA), a rare congenital vascular abnormality, is known to cause fatal aortoesophageal fistula (AEF) [1–9]. Moreover, DAA had never been reported as a cause of superior mesenteric artery (SMA) syndrome.

Case Report

A 19-year-old woman was admitted to our hospital for nausea and vomiting in April, 2009. She had a past history of bronchial asthma in her childhood. As she was emaciated due to dysphagia and her unbalanced diet for 1 year, her body indices became low as follows: body height 153 cm, body weight 29.8 kg and body mass index 12.7.

Computed tomography (CT) revealed that her stomach and duodenum were expanded to their limit, being filled with food, and that the third portion of the duodenum was stenotic between the SMA and the abdominal aorta. Based on the above-mentioned findings, she was diagnosed with SMA syndrome. A nasogastric tube was immediately inserted and over 5,600 ml of gastric contents were drained during the consecutive 4 days, and she became symptomless. The nasogastric tube was removed on the 4th hospital day. Esophagogastroduodenoscopy on the 5th hospital day disclosed extramural compression, smooth-surfaced stenosis with top ulceration that might be caused by the physical pressure of the nasogastric tube at the upper esophagus (fig. 1). The stomach was extremely dilated but empty. A liquid diet was started on the 6th hospital day and the patient became better. Suddenly, massive hematemesis appeared and she fell into shock on the 12th hospital day. Contrast enhanced CT showed that her esophagus and stomach were filled with blood. Blood pressure gradually decreased, although blood transfusions and intensive resuscitation were carried out. She died after a short while. Later, the AEF formation was found by CT (fig. 2a, b), demonstrating the cause of her death. A retrospective detailed clinical interview disclosed the fact that DAA was pointed out in her childhood, although she quit visiting a clinic since she had no symptoms.

Discussion

DAA is a rare vascular congenital abnormality and forms a complete vascular ring tightly surrounding the trachea and esophagus [10, 11]. Complete vascular ring is composed of a double aortic arch comprising the right aortic arch with the left ductus arteriosus and the aberrant left subclavian artery [11]. Our case is classified as Type IA according to Stewart and Edwards [11]. Most cases are diagnosed in infancy with only a few exceptions [1, 10, 11]. As symptoms are usually due to compression pressure to the surrounding organs such as the bronchus and esophagus, wheezing, stridor, noisy breathing and dysphasia are usual symptoms. Thus, DAA tends to be misdiagnosed as bronchial asthma or bronchitis in connection with wheezing and stridor, and as psychosomatic disorders in connection with dysphagia [5]. Our case is just such an example, as she was diagnosed with bronchial asthma in childhood and was strongly suspected of anorexia nervosa according to the Diagnostic and Statistical Manual IV (DSM-IV) criteria [12] at admission. Tracheal development when growing up had gradually hidden the symptoms due to the compression of the trachea and bronchus. Actually, adult cases of DAA complain of swallowing difficulty rather than respiratory symptoms [13]. Both dysphasia due to DAA and anorexia nervosa might have made our patient emaciated and might have resulted in SMA syndrome. DAA is diagnosed by means of CT, magnetic resonance imaging and aortography. Recently, 3D-CT is reported to be more useful for the diagnosis of DAA than other modalities [14]. The only therapy for DAA is operation, decompressing bronchus and esophagus by dividing the

non-dominant arch [10, 15]. The outcome of operation is good and it provides symptomless status in almost all cases [10, 16]. Atherosclerosis usually becomes worse by aging. Thus, symptomatic adult patients have a good indication for operation [10, 15].

AEF that is caused by intubation into the esophagus is reported to be a fatal complication in DAA patients [1–9], although only 1 case has been reported in adults [1]. As both an inserted tube and the vascular ring together compress the esophageal wall, necrosis and ulceration develop and often result in AEF accompanied with massive hematemesis [5]. Once exsanguine hemorrhage occurs in DAA patients under intubation, AEF is a most probable complication, and a Sengstaken-Blakemore tube (SB tube) is only a choice of emergency actions in such instances, controlling hemostasis through its balloon effects, and emergency operation should immediately be taken into consideration [5]. Thirteen cases of AEF caused by nasogastric tube insertion in DAA have been reported so far (table 1), in which all but 1 case were non-adults. Almost all cases had respiratory symptoms that might be related to weakness and immaturity in the bronchus. The duration of intubation with nasogastric tubes spanned 4 to 60 days, which means that intubation periods have no relation to an occurrence of AEF. It is crucial to start the treatment quickly, since 3 cases in whom the SB tubes were inserted immediately after the diagnosis of AEF were alive. Endoscopic observation was performed in 4 cases, but the bleeding point of AEF could be revealed in only 1 patient. Based on these observations, it may be concluded that endoscopy is less effective in both diagnosis and therapy of AEF.

SMA syndrome is a common gastrointestinal disease [17]. The compression of the third portion of the duodenum by the aorta and SMA causes the syndrome. Drainage of the stomach, nutritional support or operation is eventually needed. As our case showed an extremely dilated full stomach, a nasogastric tube was inserted to drain the gastric content. The patient immediately became symptomless; thus, the nasogastric tube was removed after 4 days. As a result, AEF was abruptly revealed. In this context, our case is extremely special because DAA indirectly causes SMA syndrome in adults, since dysphagia due to DAA may decrease the amount of oral intake and it may in turn result in a marked emaciation that is a major cause of the syndrome. Our case is the first that demonstrated the relationship between DAA and SMA syndrome. The nasogastric tube insertion for SMA syndrome due to DAA finally caused AEF. In fact, a dilemma exists. Firstly, drainage is apparently needed for SMA syndrome, although intubation is a contraindication in DAA. Secondly, an emergency operation should always be considered whenever AEF due to esophageal ulceration is found because endoscopic hemostasis is very difficult, or even impossible. Only the SB tube insertion may temporarily be effective for a short duration. In our case, operation would have been indicated immediately after the esophageal ulceration was found by esophagogastroduodenoscopy, although in fact we could not imagine in how far it was important. In this context, our alternative proposal is to perform intubation and drainage of the gastric content as soon as possible, and thereafter the repair operation for DAA should be indicated, since the operation is inevitable because the symptoms due to DAA were already present, one of which was SMA syndrome, which must be treated by nasogastric tube insertion.

In conclusion, all of us should pay attention to the fact that nasogastric tube insertion is a contraindication in SMA syndrome that is caused by DAA, since it may cause very serious complications such as AEF. Whenever massive hematemesis occurs in DAA patients under nasogastric tube intubation, SB tube is a possible temporary choice of action, and following emergency operation is mandatory. Thus, we believe that before intubation for SMA syndrome a detailed clinical interview clarifying whether a past history of DAA exists is mandatory to avoid AEF.

Table 1. Literature review of AEF caused by nasogastric intubation for DAA

First author Ref.	Age	Symptom	Diagnosis	Period of nasogastric tube (day)	Therapy	Outcome
Massaad [1]	38 years	–	N.D.	5	operation	alive
Angelini [2]	3 months	wheezing, stridor	aortography	23	–	dead
	3 months	respiratory distress	autopsy	>60	endoscopy, operation	dead
Chaikitpinyo [3]	2 months	difficulty of weaning off	during operation	56	Operation	alive
D'Angelis [4]	3 months	hypoxemia	echocardiography, aortography	N.D.	endoscopy, SB tube, operation	alive
McKeating [5]	3 months	wheezing, ventilator failure	during operation	17	endoscopy, operation	dead
Ohtersen [6]	5 weeks	stridor	MRI	N.D.	SB tube, operation	alive
	2 months	difficultly of weaning off	MRI	48	SB tube, operation	alive
van Woerkum [7]	9 weeks	stridor	echocardiography, CT	22	endoscopy, operation	alive
Yahagi [8]	9 days	–	during operation	8	nasogastric tube, reintubation, operation	alive
Heck [9]	6 weeks	difficulty of respiration and feeding	aortography	25	operation	dead
	3 weeks	difficulty of respiration and feeding	aortography	28	operation	alive
Miura [this case report]	19 years	dysphagia, SMA syndrome	CT	4	–	dead

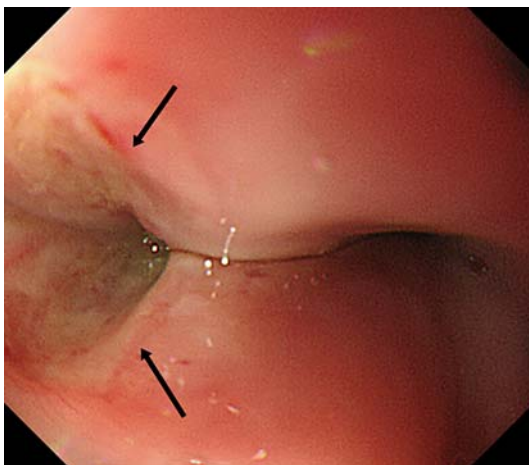
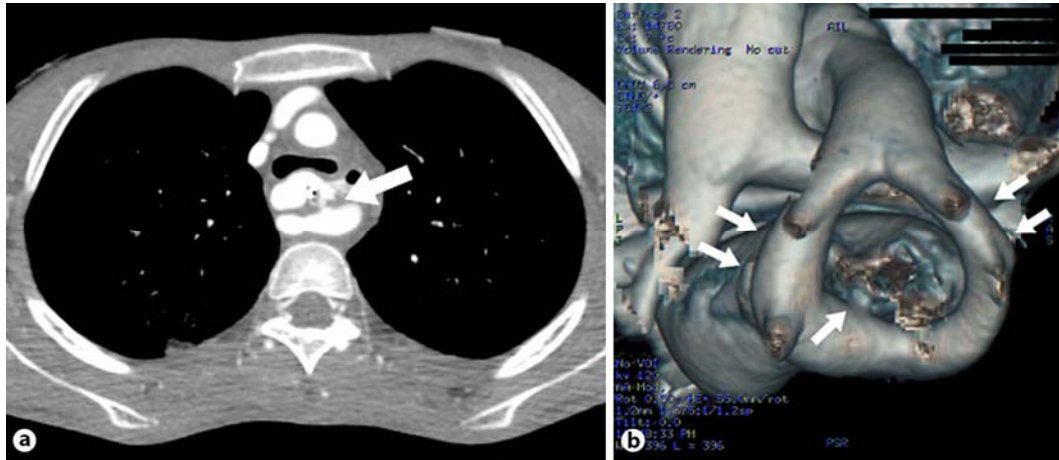
Fig. 1. Esophagogastroduodenoscopy. Esophagoduodenogastroscopy revealed extramural compression and stenosis at the upper thoracic esophagus. Esophageal ulcers caused by a nasogastric tube are discernible at the left upper corner (arrows). It is hard to pass through this compression by a routine transoral endoscope.

Fig. 2. Contrast enhanced CT at the time of hematemesis. **a** An aortoesophageal fistula is visualized by contrast medium extravasation (arrow). **b** The vascular ring (arrows) and an aortoesophageal fistula (arrow at the lower center) are clearly visible in 3D-CT images.



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