Aortoesophageal Fistula: A Life-Threatening Cause of Upper Gastrointestinal Hemorrhage in Double Aortic Arch, a Case Report

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The authors present the case of a 2 -month- old infant with double aortic arch that developed massive bright red upper gastrointestinal hemorrhage from aortoesophageal fistula (AEF) after prolonged endotracheal and nasogastric intubation. Emergency thoracotomy with AEFand double aortic arch repaired were done successfully under cardiopulmonary bypass. Due to tracheomalacia and left phrenic nerve injury, tracheal extubation could not be done until 1 month after correction of the vascular ring. The endotracheal and nasogastric tube led to fistula formation by compression of the esophageal wall against an abnormal double aortic arch. When a double aortic arch is suspected, prolonged nasogastric intubation should be avoided.

Keywords : *Upper gastrointestinal hemorrhage, Aortoesophageal fistula, Double aortic arch*

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Aortoesophageal fistula (AEF) is an abnormal communication between the aorta and the esophagus. It is a rare but life-threatening cause of upper gastrointestinal hemorrhage in infants and children[1]. In adults, it may result from thoracic aortic aneurysm, foreign body ingestion, esophageal malignancy, post surgical esophageal procedures, etc[2]. Nowadays, it is reported with increasing frequency especially in infants with a vascular ring (Table 1). Clinical diagnosis of aortoesophageal fistula is considered when an infant with double aortic arch who has tracheal and esophageal intubation develops massive bright red hematemesis. An immediate operative intervention should be undertaken. The Sengstaken-Blakemore (SB) tube is effective for temporary controlling the bleeding associated with an aortoesophageal fistula before the definitive surgical repair under cardiopulmonary bypass can be performed[3]. The authors present the case of 2 -month- old infant with double aortic arch who developed massive upper gastrointestinal hemorrhage on the 14th day of hospitalization.

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Case Report

A 17 day old boy with an unremarkable history in the perinatal period developed respiratory distress symptoms for 3 days and needed to be intubated before being transferred to a provincial hospital. Two days after he was admitted and treated as pneumonia, the boy had bronchospasm and CO2 retention with partially response to ventilatory and pharmacological interventions. The chest radiography revealed hyperinflation. The vascular ring was suspected and further investigations were performed. Barium esophagography demonstrated the extrinsic indentation at the posterior wall of the esophagus. Computed tomography of the chest was done but it was difficult to delineate the vascular structure,

Table 1. Case report of aortoesophageal fistula and vascular ring in infants and children

Author/Location/Date	Number of case	Age/Type of vascular ring	Day of massiveUGI hemorrhage	Result
Angelini [5]Italy2002	2	Infants *DAA	*	Dead
Othersen [3]USA1996	2	5 weeksDAA	10 days post aortoplexy at 9 week old	Alive
		2 monthsDAA	48th day of hospitalization	Alive
Mizushima [6]Japan1995	1	3 monthsDAA	9 days postV-P shunt operation	Dead
Sigalet [7]Canada1994	2	3.5 monthsDAA	59 days postoperative	Dead
		3 monthsDAA	*	Alive
McKeating [8]USA1990	1	3 monthsDAA	17th day of hospitalization	Dead
Arciniegas [9]*1979	1	4 monthsDAA	19 days postDAA repair	Alive

DAA = Double aortic arch; * No data

only dilatation of the upper and lower thoracic esophagus was seen. Because of the questionable definite diagnosis and failure of weaning from the ventilator, the boy was referred to our institution at 8 weeks old. Both endotracheal and nasogastric intubation were continued . Reevaluation with chest radiography, echocardiography, barium esophagography (Fig.1) and computed tomography of chest (Fig. 2) confirmed the diagnosis of double aortic arch that caused posterior compression over the esophagus and adjacent trachea above the carina level. The cardiothoracic surgeon was consulted for surgical correction of this vascular ring. The day before surgery (the 14th day after admission to our institution), the boy developed massive bright red upper gastrointestinal hemorrhage and required 380 ml of packed red cells and 280 ml of fresh frozen plasma transfusion to stabilize the hemodynamic status. Emergency surgical exploration was done. The posterior esophageal wall was compressed between the nasogastric tube and posterior arch of aorta .The aortoesophageal fistula formation, 2 mm in diameter, with bleeding was found. Double aortic arch was widely patent and equal in size of about 1 cm. The right subclavian artery originated from the posterior arch. Surgical resections of the posterior aortic arch and aortoesophageal fistula repair were performed successfully under cardiopulmonary bypass. The boy had methicillin resistance staphylococcus aureus (MRSA) septicemia and developed cardiac tamponade from massive pericardial effusion on the 11th day postoperatively and required pericardial drainage. Culture from his pericardial fluid yielded MRSA too. He received vancomycin initially then changed to rifampicin plus fosfomycin due to nephrotoxicity related to vancomycin. Although the double aortic arch was completely corrected, adequate ventilation was possible only when the tip of the tracheal tube was located just above the carina. Bronchoscopy confirmed that there was tracheomalacia at 1- 1.5 cm above the carina. Because of tracheomalacia and left phrenic nerve injury, tracheal extubation could not be done until 1 month after correction of the vascular ring. The boy continued to do well 5 months later.

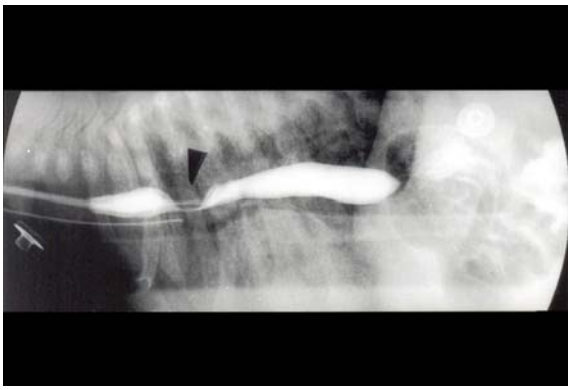

Fig. 1 Esophagogram revealed extensive posterior indentation (arrowhead). The patient had retained endotracheal tube and nasogastric tube

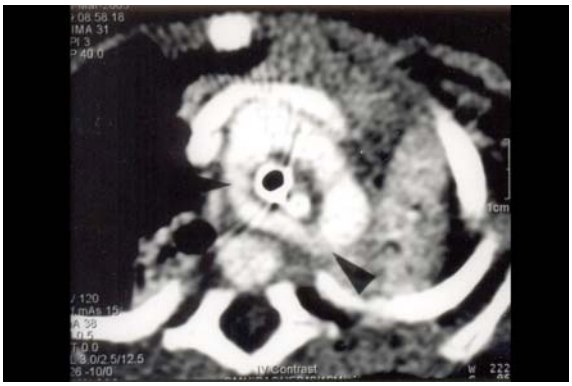

Fig. 2 Contrast enhahanced CT,axial view, demonstrated the ring (arrowhead) surrounding the central trachea and esophagus

Discussion

Aortoesophageal fistula (AEF) was the first report as the cause of death in a French sailor who had swallowed a chicken bone in 1818. Since then more than 500 adult cases of AEF have been described[2]. In 1967 Sloop and Thompson reviewed 80 cases of AEF, only 10 of them were in infants or children[1]. Now it is seen with increasing frequency especially in infants with a vascular ring. Othersen reported 2 of 30 children with double aortic arch who had AEF[3]. The present case demonstrated the typical characteristics of AEF in the double aortic arch. The clinical manifestation of double aortic arch is related to the tightness of the ring[4]. With both arches widely patent, as in the present case, the ring is very tight. The patients present with respiratory distress symptoms in the first few weeks of life. The chest roentgenogram reveals hyperinflation. Barium esophagogram shows an extrinsic indentation at the posterior wall of the esophagus. Echocardiography, angiography, computed tomography angiography or magnetic resonance imagings are needed for confirmation because the two arches may be unequal in caliber, and it is important to identify the hypoplastic segment in order to divide it. The nasogastric tube may have led to fistula formation by compression of the esophageal wall against the posterior aortic arch. When the double aortic arch is demonstrated, any esophageal tubes should be removed to prevent fistula formation. Angelini reported 2 cases of AEF, in which the fistula had sharp edges that could be iatrogenic laceration caused by manipulation of the nasogastric tubes[5]. Bright red arterial bleeding suggests that the gastrointestinal bleeding arises from the aorta and a presumptive bedside diagnosis of AEF is most likely. Gastric bleeding is less in volume and darker due to the exposure of blood to gastric acid. Typically, massive dark gastrointestinal bleeding, as the hemorrhage is venous in origin, suggests esophageal varices. Blood transfusion and pediatric Sengstaken-Blakemore (SB) tube were the temporary maneuver while waiting for surgical intervention. Endoscopy after the onset of massive hemorrhage will delay the treatment. The present case had a successful repair of both AEF and double aortic arch. Eventhough he had a difficult postoperative period, he is alive and doing well.

The authors emphasize that AEF must be diagnosed clinically from the history of double aortic arch and nasogastric intubation that develops massive bright red upper gastrointestinal hemorrhage. Prompt surgical intervention should begin as soon as possible.

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ภาวะเลือดออกรุนแรงในทางเดินอาหารส่วนต้นจาก Aortoesophageal fistula ในผู้ป่วย double aortic arch: รายงานผู้ป่วย 1 ราย

อรรฆิศา ไชกิจภิญโญ, มนัส ปะนะมณฑา, สุมิตร สุตรา, เชิดชัย ตันติศิรินทร์, จิราภรณ์ ศรีนัครินทร์, ยุทธพงษ์ วงศ์สวัสดิวัฒน์

รายงานผู้ป่วยอายุ 2 เดือนซึ่งได้รับการวินิจฉัยว่ามี double aortic arch จำเป็นต้องได้รับการรักษาด้วยการใส่ท่อช่วยหายใจและให้อาหารทางสายยางผ่านจมูกเป็นเวลานานเนื่องจากการอุดตันของทางเดินหายใจตั้งแต่อายุ 17วัน เกิดภาวะเลือดออกรุนแรงในทางเดินอาหารส่วนต้นจาก Aortoesophageal fistula (AEF) ได้รับการผ่าตัดซ่อม AEF และแก้ไข double aortic arch โดยใช้ปอดและหัวใจเทียม เนื่องจากภาวะ tracheomalacia และการบาดเจ็บของ phrenic nerve ข้างซ้าย ต้องใช้เวลา 1 เดือนจึงถอดท่อช่วยหายใจได้ ท่อช่วยหายใจและสายยางให้อาหารผ่านจมูกทำให้เกิด AEF จากการกดผนังหลอดอาหารกับหลอดเลือดที่ผิดปกติ ในกรณีที่สงสัยว่าผู้ป่วยมีภาวะ double aortic arch ควรหลีกเลี่ยงการใส่สายยางให้อาหารผ่านทางจมูก