

## Aortoesophageal Fistula Associated with Tuberculous Mediastinitis, Mimicking Esophageal Dieulafoy's Disease

Aortoesophageal fistula is a rare and lethal disorder that may result from primary diseases of aorta or esophagus, aortic bypass graft, ingestion of foreign body, trauma, surgical procedure or instrumentation. Tuberculous fistula is extremely rare. We present a 27-yr-old female patient with aortoesophageal fistula associated with tuberculous mediastinitis. The patient experienced massive hematemesis and esophagoscopy revealed a small mucosal defect with exudate-coated blood vessel like Dieulafoy's lesion on about 25 cm from the incisor teeth. Despite two sessions of endoscopic hemostatic procedures, active massive hemorrhage recurred and was controlled effectively with a prompt insertion of Sengstaken-Blakemore tube. The patient underwent open thoracotomy, which revealed aortoesophageal fistula. Numerous white-yellowish, millet seed-like tubercles were scattered in pleural and abdominal cavity. Division of fistular tract and esophageal resection with Ivor-Lewis anastomosis were performed. Histopathologic study confirmed tuberculous pleuritis and peritonitis. The patient died of postoperative pulmonary complication.

Key Words : *Esophageal Fistula; Aortic Diseases; Tuberculosis; Dieulafoy's Disease*

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### INTRODUCTION

Aortoesophageal fistula is a rare and lethal disorder. Primary aortoesophageal fistula is usually caused by primary diseases of aorta or esophagus such as thoracic aortic aneurysm, esophageal carcinoma, or reflux esophagitis (1, 2). Secondary aortoesophageal fistula may result from aortic bypass graft, ingestion of foreign body, trauma, surgical procedure or instrumentation (3-5). Tuberculous aortoesophageal fistula is extremely rare and its main cause is tuberculous esophagitis (6, 7). Moreover, aortoesophageal fistula caused by tuberculous mediastinitis has not been found in English literature.

We describe a patient with aortoesophageal fistula associated with tuberculous mediastinitis, which presented as Dieulafoy-like lesion with massive hemorrhage.

### CASE REPORT

In February 1998, a 27-yr-old Korean woman was admitted to Gyeongsang National University Hospital because of hematemesis. Six days before admission, she experienced hematemesis and visited a private hospital. Bleeding focus could not be found. Massive hematemesis recurred a day before admission. She had no history of tuberculosis, diabetes, hypertension, liver disease, trauma, and drug abuse. She had

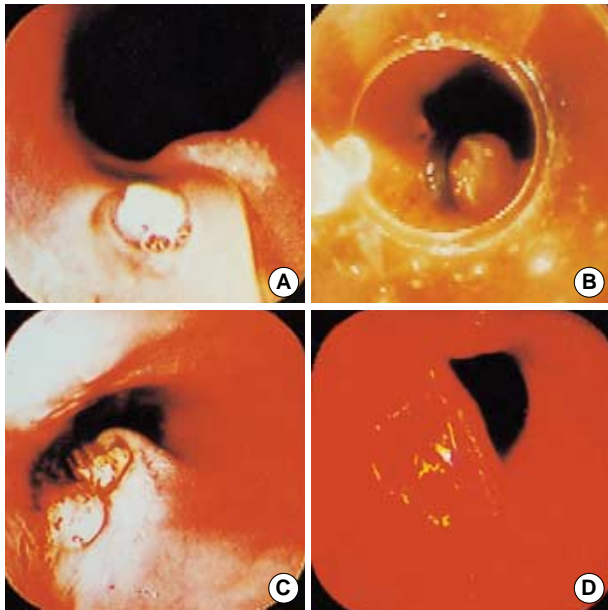
never smoked and seldom drank alcoholic beverage. Body temperature on admission was 36.2°C, and blood pressure was 118/80 mmHg. Her height was 164 cm and weight 64 kg. She was alert and her palpebral conjunctiva was pale. There was no abnormal finding in chest and abdomen. She had no lymphadenopathy.

Laboratory data were: hematocrit 24%, WBC 15,300/ $\mu$ L, and platelet 206,000/ $\mu$ L. Serum biochemistry, electrolyte, and urinalysis were normal. C-reactive protein was 28 mg/L. Hepatitis B surface antigen and antibody for hepatitis C virus by enzyme immunoassay were negative. Antibody for human immunodeficiency virus was negative. Chest radiography and electrocardiogram were normal.

Esophagoscopy revealed a tiny mucosal defect with visible blood vessel coated with exudates 25 cm from the incisor teeth (Fig. 1A). Esophageal Dieulafoy's disease was suspected and endoscopic ligation with elastic 'O' band was performed immediately (Fig. 1B). However, hematemesis developed again 4 days after ligation. Once multiple units of blood were transfused and the bleeding was controlled, pure ethanol was injected endoscopically around the same lesion (Fig. 1C). Five days after the second session of endoscopic hemostasis, another hematemesis developed, which necessitated further transfusion of multiple units of blood. Esophagoscopy showed active spurting bleeding from a large vessel (Fig. 1D). Sengstaken-Blakemore (S-B) tube was inserted immediately and

the bleeding was controlled. Once the patient was stabilized, she underwent emergent open right thoracotomy under the

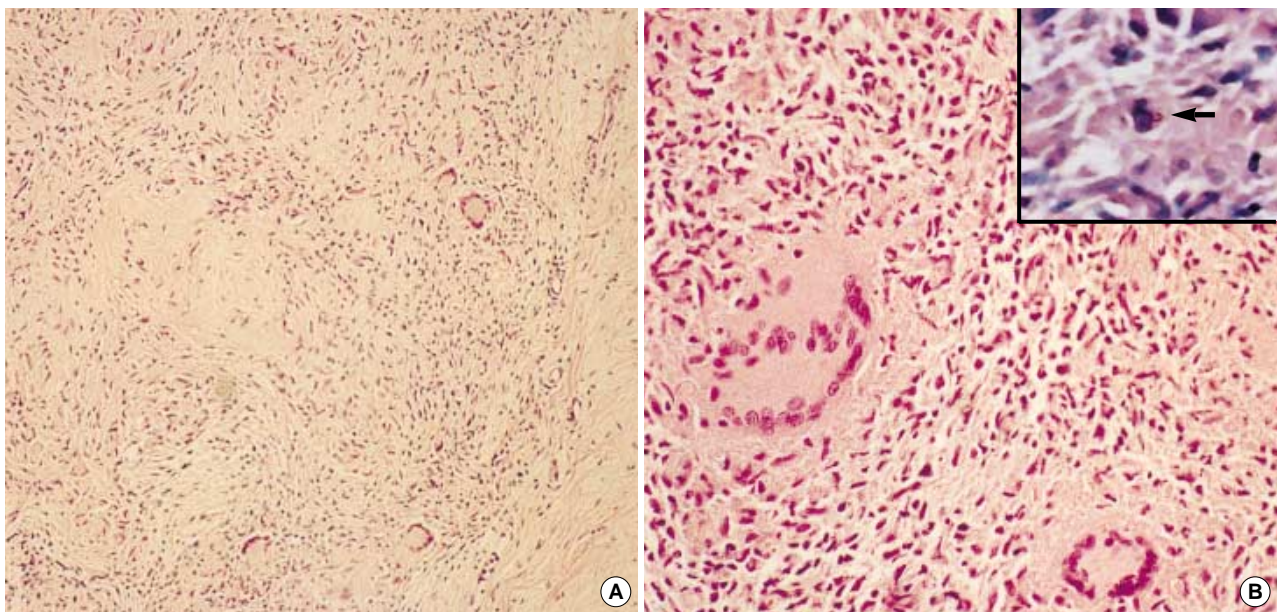
impression of large submucosal vascular malformation. On esophagotomy, a small mucosal defect coated with blood clots was found in the upper esophagus. It was obliterated with sutures in two mucosal layers. Eight days after the operation, massive hematemesis developed again and was controlled with S-B balloon tampon. Second open thoracotomy was



**Fig. 1.** Endoscopic photographs. (A) Esophagoscopy reveals a tiny mucosal defect with visible blood vessel coated with exudates 25 cm from the incisor teeth. (B) Esophageal Dieulafoy's disease is suspected and endoscopic ligation with elastic 'O' band is performed immediately. (C) She developed hematemesis 4 days after ligation and pure ethanol is injected endoscopically around the same lesion. (D) Esophagoscopy shows active spurting bleeding from a large vessel 5 days after injection of ethanol.



**Fig. 2.** Esophagus (E) is adhered to aorta (A) and fistular tract (arrow) can be identified on thoracotomy. Numerous, whitish, millet seed-like granules were scattered in the pleural surface and mediastinal tissues.



**Fig. 3.** Microscopic findings of biopsy specimens of omentum and mediastinal pleura. Epithelioid granulomas with multinucleated giant cells and caseous necrosis are observed in omentum (A: H&E stain,  $\times 100$ ) and mediastinal pleura (B: H&E stain,  $\times 200$ ). An acid-fast bacilli (arrow) is visible in mediastinal pleura (B inset: Ziehl-Neelsen stain,  $\times 400$ ).

performed and the esophagus adhered to adjacent aorta and aorto-esophageal fistula were identified (Fig. 2). Numerous whitish plaques were scattered in the mediastinal and abdominal cavities. The resection of fistular tract and partial resection of esophagus with Ivor-Lewis anastomosis were performed. Biopsy specimens were obtained from mediastinal pleura and omentum.

Microscopically, the esophageal wall tissues were damaged disorderly because of the endoscopic procedures and the first operation, but no granulomatous inflammation was detected. The fistular tract was lined by necrotic debris, acute and chronic inflammatory cells, and looked like just a esophageal ulcer. Hemosiderin pigments and calcified materials were observed at the fistular tract and esophageal adventitia. Well formed epithelioid granulomas with multinucleated Langhans' giant cells and caseous necrosis were observed in omentum and mediastinal pleura. And an acid-fast bacilli was identified in mediastinal pleura (Fig. 3).

The medication for tuberculosis was started postoperatively. She suffered from postoperative complications such as anastomotic leakage, wound evisceration, pulmonary infection, tracheo-esophageal fistula, and tracheal stricture. She died of pulmonary complication 75 days after the operation.

## DISCUSSION

Before the 1950s, tuberculosis was one of the most common causes of aortoenteric fistula. Since 1960, enteric erosion by prosthetic aortic grafts has become the most common cause and fistulas complicating gastrointestinal carcinomas have increased (8). However, tuberculosis should be considered in the countries with its high prevalence and also in the immunocompromised patients.

Local extension of tuberculous lesion from mediastinum may lead to formation of fistula between mediastinum and esophagus (esophagomediastinal fistula) or respiratory tract (tracheomediastinal fistula) (9, 10). Tuberculous aorto-esophageal fistula is uncommon. In most reported cases of tuberculous aorto-esophageal fistula, the major cause was tuberculous esophagitis (6, 7). We could not find any report of aorto-esophageal fistula caused by tuberculous mediastinitis in English literature.

We described here a case of primary aorto-esophageal fistula complicating mediastinal tuberculosis. The patient had miliary tuberculosis. Presumably the offending mycobacteria eroded through the adjacent aortic and esophageal walls and thereby resulted in a fistulous tract and massive hemorrhage, as in aortoenteric fistula associated with infective aortitis (8). When an endoscopist identifies a visible blood vessel in the gastrointestinal mucosa during endoscopic examination of patient with hemorrhage, Dieulafoy's disease is usually suspected. Bleeding from Dieulafoy's lesion is not an uncommon cause of upper gastrointestinal bleeding. How-

ever, Dieulafoy's disease is very rare in esophagus and most common in stomach (11-13). By contrast, the aortoenteric fistulas are common in duodenum and esophagus (8). The lesion in Dieulafoy's disease consists of a normal caliber artery that runs very close to epithelial layers and erodes into mucosal surface causing massive hemorrhage (12, 13). It is very difficult to differentiate aortoenteric fistula from Dieulafoy's disease in a patient with visible blood vessel who has no primary disorder of esophagus or aorta.

Clinically, affected individuals present with an acute onset of massive bleeding and they are otherwise asymptomatic (12, 13). Classical presentations often observed in patients with aorto-esophageal fistula are transient "sentinel" or "signal" hematemesis, asymptomatic "latency period", and exsanguination (4, 14).

The authors initially misdiagnosed the patient as esophageal Dieulafoy's disease because she had no history of aortic or esophageal disease, pulmonary tuberculosis, trauma, or operation, and she manifested solely as massive bleeding. So we tried two sessions of endoscopic hemostatic procedures as well as the first operation. An aorto-esophageal fistula was suspected for the first time only after massive hematemesis recurred in spite of the initial operation.

Successful management of aorto-esophageal fistula requires not only a prompt diagnosis and surgical treatment but also a prompt control of bleeding with stabilization of the patient. At the time of spurting bleeding from fistula, immediate S-B balloon tamponing was very effective for the control of active hemorrhage in this case. We could stabilize the patient promptly despite fatal hemorrhage.

We recommend that aorto-esophageal fistula should be included for the differential diagnosis despite its rarity, in cases of recurrent massive hemorrhage from esophageal visible blood vessel like Dieulafoy's disease.

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