CASE REPORT

Aortic Aneurysm Involving a Right-sided Arch Complicating
Aortobronchopulmonary and Aortoesophageal Fistula

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Abstract

A 66-year-old man with hemoptysis, chest pain, fever, and hoarseness was admitted to our department. A right-sided aortic arch and three aneurysms in the proximal arch, distal arch, and descending aorta were confirmed by aortography and surgery. Fistula formations were discovered between the proximal arch aneurysm and the right upper lobe (aortobronchopulmonary fistula: ABF), and between the descending aorta and the esophagus (aortoesophageal fistula: AEF). Concomitant ABF and AEF are very rare. Aortopulmonary and/or aortoesophageal fistula complicated by a right-sided aortic arch have not been previously reported.

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Key words: oral bleeding, mediastinal tumor, congenital anomalies

Introduction

Thoracic aneurysms have a variety of manifestations. Aortopulmonary fistula (ABF) and aortoesophageal fistula (AEF) are exceedingly rare complications of thoracic aortic aneurysm, and are generally fatal if not treated surgically. Concomitant ABF and AEF have rarely been reported in the literature (1). We describe the first reported case of aortic aneurysm complicated by ABF and AEF in association with a right-sided aortic arch.

Case Report

A 66-year-old man was transferred to our department, because of intermittent hemoptysis for 3 days. He presented with a history of hoarseness, subsequent to fever, and chest pain that began 2 months earlier. He was initially diagnosed with mediastinal tumor and treated in another hospital. He was a heavy cigarette smoker for 40 years and had diabetes mellitus (DM), diagnosed 26 years earlier, which was not managed well. Upon admission, the patient was febrile (38.5°C) with a regular pulse (96 beats/min), and a blood pressure of 130/70 mmHg. Coarse inspiratory crackles were audible at the bilateral base on auscultation. No heart murmurs were auscultated. A complete blood count revealed a hemoglobin concentration of 10.3 g/ml, 46.6×10^4 platelets, and 18,000 white blood cells with 90.6% neutrophils. C-reactive protein was 19.57 mg/dl and the erythrocyte sedimentation rate was 90 mm/h. Arterial blood samples with the patient breathing room air indicated a PaO2 of 64.5 mmHg and a PCO2 of 28.2 mmHg.

The initial bronchoscopy revealed that the right bronchi of B2 and B6 were filled with blood clots and necrotic material, but no bleeding point was detected. Gastrointestinal fibroscopy revealed stenosis of the middle portion of the esophagus without pulsation, but also did not detect the bleeding point. A chest radiograph showed a tortuous aorta that caused right hilary enlargement, right upper mediastinum widening, and esophageal nevi (Fig. 1). Echography showed bilateral pleural effusion, the analysis of which was as follows: specific gravity 1.027, total protein 3.73 g/dl, lactic dehydrogenase 361 IU/l, pH 7.484, glucose 188 mg/dl. Culture of the effusion yielded no organisms, fungus, or acid-fast bacilli. Broad-spectrum antibiotics (cefotiam) were administered for 7 days. During this time, the patient complained of nausea, slight fever, and chest pain, which was relieved by pentazocine. Intermittent hemoptysis continued for several days, and then stopped due to hypotension. Contrast-enhanced computed tomography (CT) of the chest (Fig. 2) revealed three aortic aneurysms, a retroesophageal right-sided aortic arch, pleural effusion, and dilatation of the esophagus due to compression by the aneurysm. The greatest diameters of the proximal arch, distal arch, and descending aneurysm were 2 cm, 2.5 cm, and 4.0 cm, respectively. Aortography was performed. Digital subtraction aortography demonstrated three thoracic aneurysms of the proximal and distal aortic arch, and a saccular aneurysm arising from the descending aorta (Fig. 3). Neither CT nor aortography indicated fistulas.

An emergency operation was performed. The aneurysm of the proximal arch was adherent to the right upper lobe of the

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Right-sided Arch with Aortic Aneurysm

Figure 1. Chest radiograph showed a tortuous aorta that caused right hilar enlargement, upper mediastinum widening, and esophageal nevi.

Figure 2. Contrast-enhanced computed tomography of the chest revealed a right-sided aortic arch, aortic aneurysms of the proximal and distal arch (A), dilatation of the esophagus (B), aortic aneurysm of the descending aorta, thrombus (C) and pleural effusion (A, B, C).

lung. The surgeons confirmed the presence of a fistula between the right upper lobe and the proximal arch. The aneurysm of the proximal arch was treated with a gelseal patch closure, and the saccular aneurysm of the descending aorta was treated using a prosthetic gelseal graft anastomosed end to end to the descending aorta proximally and distally to the aneurysm. The aneurysm of the distal arch was adherent to the esophagus, but was left untreated. Atherosclerotic changes were detected in the aortic wall. Exploration revealed a right-sided aortic arch with four arch vessel branches, in the following order; the left common carotid (LCC), the right common carotid (RCC), the right subclavian artery (RSA), and the aberrant left subclavian artery (LSA). The LSA was anastomosed with the left common carotid artery retroesophageally. No signs of infection were detected during the operation, and no organisms grew on blood culture.

Five days after the operation, the patient developed a fever. A chest radiograph and chest CT did not reveal a focus of infection. Antibiotics were changed from cefazolin and cefotetan to imipenem/cilastatin, and miconazole was added a few days later. Eleven days after the operation, a chest radiograph revealed an infiltrative shadow in the left lung field and a ho-
mogenous shadow in the right lung field. *Pseudomonas aeruginosa* grew on blood and sputum cultures. Tobramycin was added to the antibiotic regimen, but the fever continued despite the disappearance of the organism. Thoracic CT indicated the formation of an abscess in the right lung, which was drained by puncture-track. One month after admission, massive oral bleeding began, which could not be controlled. The patient became hypotensive and died despite attempts at cardiac resuscitation.

Postmortem examination revealed that the aneurysm of the distal aortic arch ruptured into the esophagus and there was a perigraft abscess around the graft replacement of the descending aorta. An acute esophageal ulcer adhered to the aneurysm of the descending aorta. The distal aortic arch aneurysm was a pseudo-aneurysm. Histologic examination suggested mycotic aortic aneurysm, although no organisms were detected upon examination. Pneumonia and pulmonary abscesses were also detected. Ductus arteriosus and coarctation of the thoracic aorta were not detected. Anomalies of the arch vessel branches were confirmed by examination.

Discussion

This case is very unusual, because the thoracic aortic aneurysm with a right-sided aortic arch was complicated with both ABF and AEF. ABF and AEF often cause oral bleeding, and are uniformly fatal in the absence of surgical treatment (2). Recent reports indicate that postsurgical, mycotic, and ath-erosclerotic aneurysms are the most common causes of ABF. Hemoptysis, which can appear either intermittently or massively, is the most common presenting symptom in 95% of cases (3, 4). The patient's hemoptysis appeared intermittently and stopped in association with a reduction in blood pressure. The cause of intermittent hemoptysis could be due to sealing of the fistulous communication by a thrombus (4). On the other hand, the fistulous communication in the present case might have been sealed by reduced intraluminal pressure on the fistula due to hypotension. The left lung is the most frequently affected site of ABF (4). In the present case, however, the right side was affected by the ABF, because the right-sided aortic arch was closed to the right lung.

AEF is most commonly caused by aortic aneurysm, which accounts for 75% of all cases (5). Hooper (6) estimated that 10% of aneurysms rupture into the esophagus and 6% of fatal upper gastrointestinal bleeding is caused by an aneurysm. Aortography is useful for detecting aneurysms, although it does not serve to detect fistulas (7). Esophagrams often reveal fistulas, and are more useful than aortography. Thoracic CT rarely reveals fistulas. In this case, an esophagram was not performed, and neither aortography nor thoracic CT with contrast revealed the fistula. Gastrointestinal fiberscopy also did not reveal the bleeding point, or the pulsating and obvious fistula.

A right-sided aortic arch is found in approximately 0.1% of the general population. The most common type of right aortic arch is associated with an aberrant left subclavian artery and aortic diverticulum, in contrast to the mirror-image type of right-sided aortic arch, which is frequently associated with congenital heart disease (8). A vascular ring, which typically consists of a connection between the left subclavian artery and the left ductus, is usually present and is often the cause of any clinical symptoms. The patient did not have aortic diverticulum, or a typical vascular ring, and was not complicated by congenital heart disease. He did not have a past history of clinical symptoms caused by the ring, although anastomosis between LCC and LSA was detected retroesophageally.

There are two reports of dissecting aortic aneurysms involving a right-sided aortic arch (9, 10), but to our knowledge there are no previous reports of associating non-dissecting aortic aneurysms in the English literature. In aortic dissection cases, it is thought that a short radius and an acute aortic curve lead to dissection in a right-sided aortic arch. We speculated that these anatomic features and atherosclerotic changes of the aortic arch induced the aneurysm. To our knowledge, an aortic aneurysm with a right-sided aortic arch that penetrates to the bronchopulmonary parenchyma or esophagus has never been reported.

Histology of the aortic wall confirmed the pseudoaneurysm,
which was presumably of mycotic origin. It can thus be speculated that the fever on admission was due to the aneurysm. Mycotic aneurysm has a poor prognosis because of rapid perforation and infection (11). In 24% of patients with mycotic aneurysm, there is an association with depressed immunocompetence, such as DM, alcoholism, collagen vascular disease, corticosteroid administration, and malignant neoplasms, while primary mycotic aneurysm is very rare (12). Especially in older patients, it is common to find infection superimposed on atherosclerotic plaques (12). The present patient had a past history of DM, therefore, it could have had a causative role in the development of the mycotic aneurysm; DM could cause both atherosclerotic changes and an immunosuppressive state. There was an esophageal ulcer with an AEF. The descending aorta was replaced with artificial blood vessels, but the esophagus was not treated in the operation. Leakage of the untreated esophagus might have lead to perigraft abscess, which could have caused the fever in the early postoperative period. Mycotic aneurysms might be caused by an infectious disease, bacterial endocarditis, and concurrent sepsis (12), so sepsis due to the postoperative infections worsened the untreated aneurysm, leading to rupture into esophagus. Timely management of the esophageal fistula can reduce the possibility of recurrent fistula and mortality due to mediastinal contamination.

In conclusion, ABF and/or AEF should be considered when oral bleeding presents with a mediastinal mass. Anatomic features, as well as atherosclerotic and inflammatory changes of the aortic arch probably induced the aneurysm. A precise anatomic definition is important for the diagnosis and treatment of an aortic aneurysm with congenital anomalies, such as a right-sided aortic arch. In present case, good management of the DM is important for the prevention of aneurysms and ruptures. Successful treatment result is based on an immediate operation and an aggressive approach to gain aortic control and remove the penetrated esophagus.

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Reference