Inflammatory Aneurysm of Arcus Aorta with Massive Hemoptysis

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ABSTRACT

A 72-year-old woman was admitted to the intensive care unit with a diagnosis of acute coronary syndrome. An aneurysm in the aortic arch was detected in the radiologic investigations. The aortic arch was replaced with a Dacron graft with the patient under total circulatory arrest. A pathology evaluation revealed an inflammatory aneurysm. To date, a few cases of inflammatory aneurysms of the aortic arch have been reported. Unlike the previously reported cases, the aneurysm in our case unexpectedly presented with massive hemoptysis.

INTRODUCTION

Inflammatory aneurysms can occur in all parts of the aorta, but involvement of the thoracic aorta is seldom detected. This entity has been known since 1935, and the term *inflammatory aneurysm* was first used by Crawford et al [1985], who reported a 5-case series of inflammatory aneurysm of the descending aorta. We present the case of a patient who had an inflammatory aneurysm of the aortic arch that manifested with massive hemoptysis and who underwent urgent operation.

CASE REPORT

A 72-year-old woman with a history of hypertension and cerebrovascular disease presented with chest pain and was treated in the intensive care unit after receiving a diagnosis of acute coronary syndrome. In routine investigations, the posteroanterior chest radiograph revealed a broad upper mediastinum and a lesion with ill-defined borders in the left lung hilus that penetrated into the parenchyma (Figure 1). On the same day as the hospitalization, the patient experienced recurrent episodes of sudden massive hemoptysis. A computed tomography scan revealed an aneurysm in the aortic arch

between the left carotid and subclavian arteries. The aneurysm had ill-defined borders and an irregular contour and was adjacent to the neighboring lung parenchyma. The diameter was 4 cm (Figure 2A). A magnetic resonance imaging examination performed on the same day to identify the vascular involvement of the lesion showed an aortic wall thickness of 3.5 cm (Figure 2B). An aortography evaluation showed a saccular aneurysm of 44×45 mm in the aortic arch that reached the left subclavian artery inferiorly and pushed the left common carotid artery (Figure 2C). Cardiac enzyme levels and the results of a coronary angiographic evaluation were normal. The cranial computed tomography scan showed advanced chronic cerebral atrophy. The results of an evaluation of the neurologic system were normal. A psychiatry consultation detected mood and affect dissociation. Before we could perform a planned bronchoscopy examination to exclude pneumologic problems, the total volume of the hemoptysis reached 700 mL within 6 hours because of the recurring hemoptysis attacks. Therefore, the bronchoscopy evaluation was postponed and instead rescheduled for the operating room, where the patient was taken for an urgent operation. Because of the progressively declining hemodynamic status of the
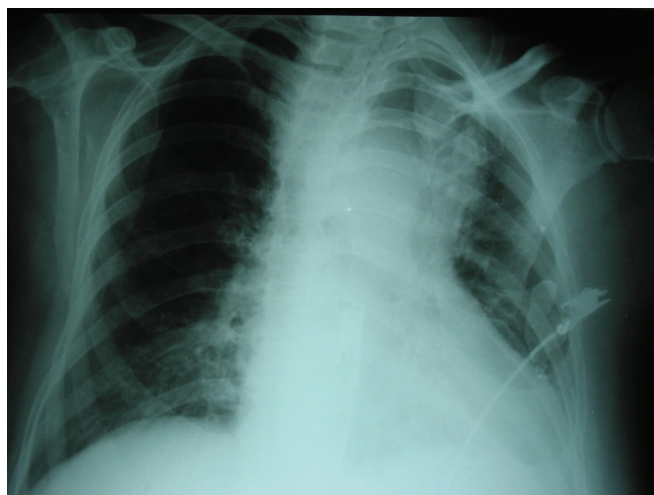

Figure 1. The posteroanterior chest radiograph showing the lesion in the left lung hilus.
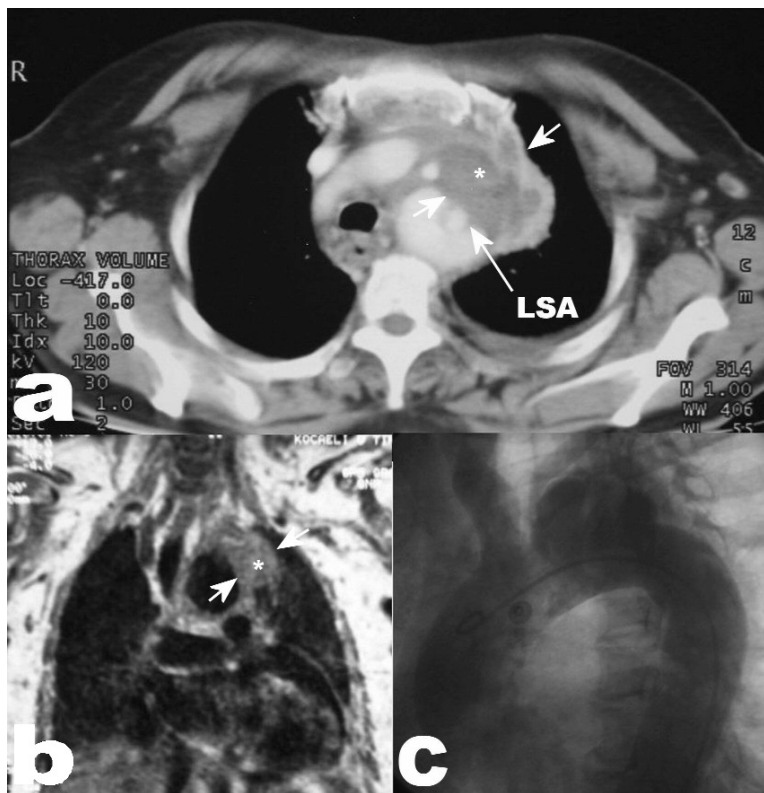
Figure 2. A, Computed tomography scan of the aneurysm in the aortic arch between left carotid and subclavian arteries. Asterisk indicates the diameter of the aneurysm. LSA indicates left subclavian artery. B, Magnetic resonance image showing the thickness of the aneurysm wall. Asterisk indicates the width of the aortic wall. C, Angiographic image of the aneurysm.

patient in the operating room, the bronchoscopy evaluation was cancelled and inotropic support was initiated. The patient then underwent operation emergently.

Cardiopulmonary bypass was established via cannulation of the right common femoral vein and artery. We then initiated total circulatory arrest and began retrograde cerebral perfusion through the superior vena cava. Exploration revealed the ascending aorta to be intact but also showed an aneurysm on the anterior aortic arch, which was localized to the proximal innominate artery. The aneurysm was dissected from the thoracic wall and the lung parenchyma. The aneurysm had damaged the apex of the lung, and there was a fistula between the aneurysm and the superior bronchus. The locations of the left common carotid artery and the left subclavian artery were shifted because of the aneurysm. The proximal part of the left subclavian artery was involved in the aneurysm sac. The aortic arch was incised longitudinally and divided from both ends. The aortic wall was extremely thick and hardly dissected. Although the innominate and left common carotid arteries were not completely intact, both were prepared as small cuffs of aortic tissue. The part of the subclavian artery that was entrapped in the aneurysm sac was separated from both the proximal and distal ends. There was wall thickening in the aortic arch and in the proximal 2 cm of the descending aorta. The aortic arch was replaced with a 28-mm polyethylene terephthalate fiber (Dacron) graft. Because of the excessive wall thickening in the descending aorta, 2/0 polypropylene (Prolene) suture had to be used with large needles in the distal anastomosis of the graft. The innominate and the left common carotid artery cuffs were sutured on the graft. Through meticulous dissection of the left subclavian artery, we were able to separate the artery but had to reduce it in length. Therefore, we interposed a 6-mm polytetrafluoroethylene graft between the left subclavian artery and the Dacron graft. The total time of circulatory arrest was 72 minutes. We capitonnaged the apex of the left lung and then terminated cardiopulmonary bypass without difficulty.

The patient was taken to the intensive care unit. There were no hemodynamic problems or hemorrhage during the early postoperative period. Early postoperative neurologic examination of the patient showed the patient to be awake and responding to verbal orders, but she was uncooperative. There were no pathologic reflexes. Because of the neurologic status of the patient, multiple attempts to wean her from the mechanical ventilator failed. The patient died from cardiac arrest in the intensive care unit on postoperative day 47.

A gross examination of the aortic arch specimen revealed the aortic wall to be excessively thickened and covered with connective tissue. There were no pathologic findings for the lung and the thoracic wall adjacent to the aneurysm.

Table 1. Demographic Characteristics of Cases of Thoracic Aortic Inflammatory Aneurysm Reported in the Literature*

Reference	Case			Aneurysm Characteristics		
	Age, y	Sex	Clinical Manifestation	Length	Width	Location
[Connery 1994]	69	F	Angina, COPD, HT, CHD	85 mm	10 mm	Ascending aorta
[Federmann 1996]	60	M	Angina	50 mm	≥8 mm	Ascending aorta
[Girardi 1997]	61	F	Angina, MI, HT	50 mm	30 mm	Ascending aorta and aortic arch
[Kunzli 1998]	60	M	Angina, gastritis	—	≥10 mm	Ascending aorta
[Roth 2003]	65	F	Depression/asystole	45 mm	10 mm	Ascending aorta

*COPD indicates chronic obstructive pulmonary disease; HT, hypertension; CHD, congestive heart disease; MI, myocardial infarction.

A microscopy investigation revealed highly thickened adventitia and media that were infiltrated by lymphocytes and plasma cells.

DISCUSSION

Inflammatory aneurysm of the aorta is a clinical entity of unknown etiology, even though it constitutes 2.5% to 10% of the abdominal aortic aneurysms that undergo operation [Connery 1994]. Only a few cases of inflammatory aneurysms confined to the arch and the ascending aorta have been reported to date. As far as we are aware, only 5 cases of inflammatory aneurysms confined to the ascending aorta and/or aortic arch have been reported in the English language literature (Table 1).

Chronic periarteritis, idiopathic mediastinal fibrosis, and perianeurysm retroperitoneal fibrosis have been suspected in the etiology of inflammatory aneurysm, but the exact reasons for the condition are unclear [Crawford 1985; Connery 1994]. An autoimmune reaction against an unidentified component of atherosclerotic plaques was believed to be the cause of inflammatory changes, so corticosteroids have been included in treatment regimens [Connery 1994].

The main pathology is extreme thickening of the aortic arch and its adherence to nearby organs because of the periaortic fibrosis. In our case, the aneurysm had advanced to the aortic arch and involved the proximal left subclavian artery. Connery et al [1994] complained of a challenging dissection in their case and stated that the advanced thickening of the aortic wall might have been the reason for the increased perioperative bleeding. On the other hand, Girardi and Coselli [1997] performed minimal dissection to avoid complications and did not perform an aneurysmectomy in their operation. In our case, we did not have the opportunity to choose a conservative treatment modality because the tumor was highly adherent to the adjacent tissues and had fistulized to the left lung apex.

Although rupture of the thickened aortic wall in inflammatory aneurysm may seem controversial, the rupture may eventually be from the infiltrated wall to a body space, as well as to an organ, as Girardi and Coselli [1997] stated in describing their case. The periaortic fibrosis in our case caused adherence to the lung and led to the formation of a bronchial fistula. The massive hemoptysis caused by the fistula was the leading clinical presentation. In the intraoperative exploration, we saw that the anterior part of the aortic arch was highly thickened, had hardened, and had become fibrotic; the adventitia had an irregular structure and had changed color to gray and white. We did not detect any dissection membrane or intramural hematoma in the thickened aortic wall. Macroscopically, the borders of the aortic wall layers could not be identified. We saw that excision of the aneurysm was possible only if the left lung apex and the posterior wall of the sternum were also excised. The macroscopic view in our case was unique for periaortic fibrosis in inflammatory aneurysms.

Because of the lack of cases in the literature, there is no consensus on the typical symptoms, complications, diagnosis, and operative techniques for inflammatory aneurysms of the aortic arch and the ascending aorta. The unique characteristic of inflammatory aneurysm is the periaortic fibrosis, which not only leads to dense adhesions that make dissection nearly impossible but also causes compression symptoms to nearby organs that lead to organ malfunction. Unlike previously reported cases, the aneurysm in our case unexpectedly fistulized to a bronchus to produce massive bleeding with hemodynamic deterioration. We believe that in cases of aneurysms of the thoracic aorta with highly thickened walls that adhere to neighboring tissues, a nonspecific inflammatory process must be kept in mind and the surgical planning must consider obstacles that may be unexpected.

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