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Fatal haemoptysis due to ruptured aortic arch aneurysm with aorto-pulmonary fistula

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Summary

An 80-year-old woman presenting with a single episode of acute haemoptysis yielded a cup of fresh blood. She gave a history of acute onset breathlessness and retrosternal chest discomfort, breathlessness and chest discomfort (1 day). She had no history of pulmonary tuberculosis or bleeding diathesis. She was diagnosed with hypertension 20 years before, but she discontinued medications. On examination she was conscious, alert, pale and without cyanosis or icterus. Pulse rate was 100/mt, regular BP 200/140 mm of Hg right upper limb in the supine position. The radiograph of her chest showed an aortic arch aneurysm. A high-resolution computed scan of the chest showed a leaking aortic arch aneurysm with haemothorax and destruction of the bronchoalveolar architecture. Surgical correction was planned but she succumbed because of hypovolaemic shock. This case is reported as it is an important differential diagnosis of refractory haemoptysis.

BACKGROUND

Aneurysms can occur anywhere along the aorta but isolated aortic arch aneurysms are uncommon and are usually associated with ascending and descending artery aneurysms.¹ Rupture of a vessel is usually accompanied by severe pain. A rupture into the pleural cavity is more common on the left and can cause dyspnoea, pleuritic chest pain and hypotension. Aneurysms can rupture into the mediastinum, oesophagus, trachea or bronchial tree.¹ Aortic arch aneurysm with acute aorto-pulmonary fistula is usually a postmortem diagnosis. Few reports of successful surgical management have been described. Despite the many advances in cardiac surgery over the last 30 years, the observed death rate for surgical correction has been very high. Early diagnosis and prompt surgical intervention using profound hypothermia and total circulatory arrest are essential to successful outcome.² This uncommon complication of an aortic arch aneurysm rupturing, resulting in haemothorax with aortobronchial fistula culminating in refractory haemoptysis, is reported

CASE PRESENTATION

An 80-year-old woman presenting with a single episode of acute haemoptysis yielded a cup of fresh blood. She gave a history of acute onset breathlessness and retrosternal chest discomfort, breathlessness and chest discomfort (1 day). She had no history of pulmonary tuberculosis or bleeding diathesis. She was diagnosed with hypertension 20 years before, but she discontinued medications. On examination she was conscious, alert, pale and without cyanosis or icterus. Pulse rate was 100/mt, regular BP 200/140 mm of Hg right upper limb in the supine position, respiratory rate 28/min. There were no visible suprasternal pulsations and the peripheral pulses were equal and palpable. She had cardiomegaly with heaving apex. The heart sounds were normal except for a loud A2. Trachea was deviated to the

right with bilateral diminished movements and a reduction in breath sounds. Optic fundi showed bilateral papilloedema. Other systems were normal.

INVESTIGATIONS

All haematological parameters including a peripheral smear were normal. The coagulation parameters, antinuclear antibody, double-stranded-DNA, HBS Ag, Venereal Disease Research Laboratory, HIV I and HIV II were negative. Renal function tests were normal. Electrocardiogram revealed left ventricular hypertrophy with strain. The x-ray of the patient's chest PA view showed a haziness in the left upper zone with well-defined, rounded margins continuous with the aortic arch pushing the trachea to the right. No lung tissue was tracable between the upperlobe opacity and the mediastinum, suggesting an aortic aneurysm (figure 1A). CT scan of the thorax revealed aneurysm of the aortic arch (figure 1B), with contrast-filled lumen (figure 1B-A), massive thrombus (figure 1B-B), intimal calcification (figure 1B-C,D), peri-lesional inflammatory changes indicating leak into the pleura and lung parenchyma (figure 1B-E). The adjacent lung parenchyma on the left side shows alveolar opacities and collapse consolidation (figure 2A, upper arrow 1), left haemothorax confirmed by pleural aspiration (figure 2A, bottom arrow 2) and diffuse inflammation involving the wall of the aorta (figure 2B, white arrows).

DIFFERENTIAL DIAGNOSIS

She was diagnosed as having a ruptured aortic arch aneurysm with haemothorax and rupturing into the lung resulting in refractory haemoptysis.

TREATMENT

She was infused with sodium nitroprusside 0.3 µg/kg/mt over 10 min, which brought down her blood pressure to 160/120 mm Hg, and then maintained with labetalol.

Figure 1 (A) X-ray of the chest PA view showing haziness in the left upper zone with well-defined, rounded margins continuous with the aortic arch pushing the trachea to the right. No lung tissue was tracable between the upperlobe opacity and the mediastinum, suggesting an aortic aneurysm (white arrows). (B) CT scan of the thorax revealed aneurysm of the aortic arch: A, contrast-filled lumen; B, massive thrombus; C, intimal calcification; D, E, perilesional inflammatory changes indicating leak into the pleura and lung parenchyma.

Although her blood pressure was controlled, she developed massive haemoptysis and succumbed the following day due to hypovolaemic shock.

OUTCOME AND FOLLOW-UP

The patient succumbed the following day due to hypovolaemic shock.

DISCUSSION

Ascending aortic aneurysms mainly occur more frequently in men than in women, are typical fusiform and extend into the aortic arch. Consequently, aneurysms of the aortic arch are often contiguous with aneurysms of the aortic arch.³ The risk increases by 40% every 5 years after the age of 65. Smoking is an independent risk factor for AAA, with level of exposure being more significant than duration.⁴ Causes of aneurysm are syphilis, atherosclerosis, cystic medial necrosis, Marfans syndrome, Ehler-Danlos syndrome, syphilitic periaortitis and mesoaortitis and damaged elastic fibres, resulting in thickening and weakening of aortic wall. Approximately 90% of syphilitic aneurysms are located in the ascending aorta or arch of aorta.⁵ Thoracic aneurysms may involve one or more aortic segments (aortic root, ascending aorta, arch or descending

aorta) and are classified accordingly (60% of thoracic aortic aneurysms involve the aortic root and/or ascending aorta, 40% involve the descending aorta, only 10% involve the arch, and 10% involve the thoracoabdominal aorta).⁶ Aortic arch aneurysms are most often asymptomatic but can present with symptoms caused by compression or acute aneurysm expansion, such as upper chest or shoulder pain and hoarseness of voice.⁷ However, there are few reports of patients with an aneurysm rupturing into the lung and presenting with haemoptysis.⁷ Surgical replacement of the aortic arch aneurysms is particularly challenging and carries a significant risk of neurological damage from embolisation of atherosclerotic debris or from global ischaemic injury during circulatory arrest.⁸

CONCLUSIONS

A high index of suspicion is urged for an untreated hypertensive found to have an aneurysm presenting with haemoptysis. Early diagnosis and emergent surgery are the two prognostic factors determining survival. Surgical treatment of a leaking aortic arch aneurysm is impossible. Rupture is mentioned as one of the differentials of massive haemoptysis in an untreated severe hypertensive who is established to have an aneurysm.

Figure 2 (A) Upper arrow. 1, The adjacent lung parenchyma on the left side shows alveolar opacities and collapse consolidation. Bottom arrow. 2, Showing left haemothorax (confirmed by pleural aspiration). (B) White arrows showing diffuse inflammation involving the wall of the aorta.

Learning points

- ▶ Severe and refractory haemoptysis in elderly women with untreated hypertension having aortic aneurysm should raise the possibility of its rupture.
- ▶ In an 80-year-old one has to be aggressive in excluding a ruptured aneurysm. Such leaking aneurysms usually present with 'warning haemoptysis'.
- ▶ Ruptured aneurysms will carry a very high mortality with or without surgical intervention.
- ▶ A high index of suspicion will decrease delayed diagnosis. Early diagnosis and emergent surgery are two prognostic factors for survival.⁶
- ▶ Surgical replacement of aortic arch aneurysms is particularly challenging and carries a significant risk of neurological damage from embolisation of atherosclerotic debris or from global ischaemic injury during circulatory arrest.⁸

Competing interests None.

Patient consent Obtained.

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