Fatal Bleeding Disorder as the First Manifestation of a Giant Aortic Aneurysm

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ABSTRACT

Aortic aneurysms are a rare cause of consumption coagulopathy. We describe an 82-year old woman with a previously unrecognized giant aneurysm of the thoracic aorta. The first manifestation of this condition was hemoptysis that was due to an exacerbation of chronic disseminated intravascular coagulation. The patient eventually died despite intensive conservative treatment, as she declined late surgical intervention. This case underscores the need to exclude aortic aneurysms by performing a chest X-ray in elderly patients with bleeding disorders.

CASE REPORT

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An 82-year-old woman with a two-year history of ecchymoses was admitted for acute hemoptysis. On physical examination, she had multiple ecchymoses in the trunck and the extremities and decreased breath sounds over the left lung. A PA chest radiograph revealed a giant thoracic aortic aneurysm (Fig. 1). Spiral computed tomography confirmed the presence of an aneurysm, 5.5 cm in diameter in the ascending aorta, 6 cm in the aortic arch and 8 cm in the descending aorta (Fig. 2). Among patients who present with hemoptysis in the United States, bronchitis, bronchiectasis and lung cancer each account for about one fifth of the cases, although the proportions vary according to age. Other causes that should be routinely considered include community-acquired pneumonia, congestive heart failure, pulmonary embolism, bleeding diatheses, tuberculosis, aspergillosis, vascular abnormalities and drugs, including cocaine (1).

The chest computed tomography that was obtained excluded most of the above-mentioned causes of acute

hemoptysis. The patient's platelet count was 83,000/mm3, the international normalized ratio 1.8, the fibrinogen level only 20 mg/dL and D-Dimer 6.14 μ g/mL, findings consistent with the presence of DIC. Following extensive laboratory investigations other causes of coagulopathy were excluded (no schistocytes were found in the blood smear) and her condition was attributed to an exacerbation of chronic DIC due to the giant aneurysm. She received fresh frozen plasma but declined further surgical treatment of the aneurysm. She died suddenly, two months later, due to a massive hemoptysis.

DISCUSSION

Disseminated intravascular coagulation (DIC) is a severe clinical condition characterized by the widespread activation of coagulation process which results in the intravascular deposition of fibrin in small and medium sized-vessels. Fibrin deposition may result in thrombotic occlusion of the vessels that may contribute to the decreased blood supply to vital organs and therefore to hemodynamic derangements. Except from the thrombotic and ischemic injury of multiple organs, the consumption and subsequent depletion of platelets and coagulation factors may result in the emergence of severe bleeding (2). In some cases, usually in malignancies, DIC develops gradually and is known as chronic DIC, which is presented predominantly with thrombotic events (3).

The most common causes associated with DIC are infections, especially septicaemia, severe trauma, neoplasms, obstetrical complications, vascular disorders such as giant hemangiomas or aortic aneurysms and severe immunologic disorders such as hemolytic transfusion reactions or severe allergic reactions (2). Aortic aneurysms are a rare cause of DIC, accounting for only 0.5%-6% of such cases (3-5). Conversely, clinical overt DIC is an even rarer complication of large aortic aneurysms, occurring in approximately 0.5%-1% of these patients (6, 7).

The cornerstone of DIC management is the treatment of the underlying disorder. Treating DIC without treating its underlying cause is predestined to fail (2). Surgical repair usually provides cure to the aneurysm-associated consumptive coagulopathy (6, 8). Late recurrence of DIC after surgical repair appears to be very rare (9). Supportive treatment might be useful in ameliorating symptoms of DIC preoperatively. Unfortunately, in cases of inoperable aneurysms supportive measures are the only choice (10, 11).

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Aortic aneurysm is a circumscribed axial or lateral area of localized widening in the aortic wall (12). Aneurysm is the most common condition of the thoracic aorta requiring surgical treatment (13). Findings on the chest radiograph may be diagnostic of a thoracic aortic aneurysm. However, approximately 17% of the patients with documented aortic aneurysms or dissections have no abnormalities on chest radiography (14). Substantial enlargement of the ascending aorta may be confined to the retrosternal area, so that the aortic silhouette appears normal. Aneurysms that involve the ascending aorta and the aortic arch cannot always be differentiated from tumors or other masses of the mediastinum (15). Computed tomography is the most widely used noninvasive technique for the diagnosis of thoracic aortic disease, as it provides information about the location, size and extent of the disease. It is of particular value in diagnosing thoracic aortic dissection, in determining the timing of operation for asymptomatic patients and for postoperative evaluation. Its main disadvantage is that it requires the use of contrast medium for precise delineation of the aortic disease, which is contraindicated in patients with renal failure and in patients with allergies to the contrast medium. Magnetic resonance imaging is emerging as the imaging method of choice for the diagnosis of thoracic aortic disease in patients with allergies to contrast agents used in computed tomography. Its use in patients with renal impairment is not advocated lately due to the risk of nephrogenic systemic fibrosis caused by gadolinium contrast (16). Transesophageal echocardiography with Doppler imaging is considered the most accurate and useful technique in diagnosing acute aortic dissection (13). Aortography is still widely used for the detection of aortic dissection and is usually acquired for most patients undergoing elective surgery, but has the same disadvantages with computed tomography (14).

The key to a successful diagnosis in this case of aortic aneurysm related DIC was careful attention to the patient's history and expansion of the differential diagnosis according to the findings (chest X-ray, CT, laboratory values). Consultation with a surgeon and a hematologist is warranted in cases of large aneurysms complicated by DIC, as prompt clinical decision-making may prove very difficult.

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FIGURES

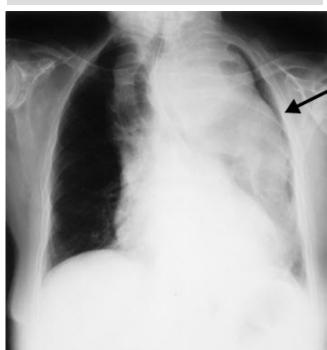


Figure 1: Anteroposterior chest X-ray of a 82-year-old female patient who presented with an aortic aneurysm related DIC. A soft-tissue mass continuous with the aortic silhouette and associated with the aneurysm of the ascending aorta is seen, that projects to the right displacing the trachea and producing widening of the superior mediastinum, while the aneurysm in the descending aorta projects to the left covering almost the whole left hemithorax.

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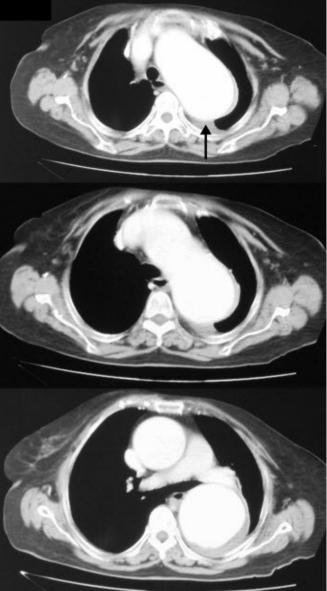
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Figure 2: 82 year old woman with fatal bleeding disorder as the first manifestation of a giant aortic aneurysm. Contrastenhanced axial chest CT depicting the large aortic aneurysm, 5.5 cm in diameter in the ascending aorta, 6 cm in the aortic arch and 8 cm in the descending aorta.

ABBREVIATIONS

CT= Computed tomography DIC = disseminated intravascular coagulation

KEYWORDS

Aortic aneurysm, disseminated intravascular coagulation, DIC, chest X-ray, Computed tomography, CT