Aortocaval fistula: a rare cause of paradoxical pulmonary embolism

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Summary: An 83 year old woman died suddenly from a paradoxical pulmonary embolus which had originated in an abdominal aortic aneurysm and embolised via an aortocaval fistula. This lesion should be considered in the differential diagnosis of embolic disease.

Introduction

Paradoxical emboli are uncommon and usually associated with cardiac septal defect. Origin of the embolus from an aortic aneurysm sac with passage via an aortocaval fistula is rare with only two cases found in the literature. One case occurred in a 60 year old man who presented with intractable cardiac failure;¹ pulmonary angiography demonstrated emboli and aortography showed an aortocaval fistula which was successfully repaired. Massive embolism has also occurred during surgery on an aortic aneurysm with a preoperative angiographic diagnosis of fistula.²

There appears to be no similar case to that presented now in which paradoxical embolism caused sudden death in a previously asymptomatic individual.

Case history

An 83 year old woman was found at home having died in the night. She was known to have an asymptomatic abdominal aortic aneurysm. She had been living an active life and was otherwise fit and well. There was no other significant medical history.

Autopsy findings

Within the left main pulmonary artery was a large, firm 2.5 by 2.5 by 1 cm embolus composed of laminated thrombus. The lungs showed no evidence of previous embolic disease or pulmonary

hypertension. There was a saccular abdominal aortic aneurysm, 8 cm diameter, which arose below the origin of the renal arteries. An aortocaval fistula was present, measuring 2.5 by 1 cm, and a thrombus could be seen protruding through into the inferior vena cava (Figure 1). There was no evidence of right ventricular hypertrophy; the coronary arteries showed moderate atherosclerosis.

Discussion

Most paradoxical emboli pass from the venous side through an atrial septal defect to the arterial side and cause systemic infarcts, particularly in the brain. They rarely pass through ventricular septal defects because the pressure and flow relationships are usually against it. This case demonstrates an apparently rare association with aortocaval fistula. This type is unusual in that the embolus passes from the arterial to the venous side and manages to do it without a massive arteriovenous shunt. This complication may be expected to become more common with the increasing age of the population and thus increase in incidence of abdominal aortic aneurysm.

Aortocaval fistula itself is an uncommon complication of abdominal aortic aneurysm. The aetiology may be traumatic or spontaneous. The commonest cause is secondary to rupture of an atherosclerotic aortic aneurysm into the inferior vena cava;³ this complication was found in three of 211 operated abdominal aortic aneurysms. Trauma accounts for up to 20% of the cases⁴ and includes gunshot wounds and iatrogenic fistulae

following intervertebral disc surgery.⁵ Less common causes are syphilis, bacterial infections, Marfan's or Ehlers-Danlos syndromes.⁶

The cardiovascular complications of abdominal aortic aneurysm have recently been reviewed.⁷ High output cardiac failure with features suggesting tricuspid insufficiency may develop. Most patients have a pulsatile mass associated with a bruit that is usually continuous. Arterial insufficiency can lead to angina. Raised venous pressure may cause lower limb oedema, haematuria or priapism.

The diagnosis in this case was made at autopsy and emphasizes the importance of this investigation in medical education. Aortocaval fistula should be added to cardiac septal defect as the source of paradoxical emboli and considered in the clinical differential diagnosis of embolic disease.

Figure 1 Aortocaval fistula with protruding thrombus (arrow), viewed from the medial aspect of the opened inferior cava. Part of the aortic aneurysm is also seen (arrowhead).

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