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Case Report

Unexpected death caused by rupture of a dilated aorta in an adult male with aortic coarctation

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ABSTRACT

Aortic coarctation (AC) is a congenital aortic narrowing. We describe for the first time the findings obtained by unenhanced post mortem computed tomography (PMCT) in a case where the death was caused by cardiac tamponade from a ruptured aneurysmal dilatation of the ascending aorta and the aortic arch without dissection combined with aortic coarctation. The patient, a 46-year-old man, was found dead at home. PMCT showed haemopericardium and dilatation of the ascending aorta and the aortic arch. This appearance led to the mistaken interpretation that the images represented a dissecting aneurysm. The autopsy showed instead a thin-walled and floppy dilatation of the ascending aorta and aortic arch with a coarctation just proximal to the ligamentum arteriosum. A longitudinal tear was found in the posterior aortic wall just above the valves. Blood in the surrounding soft tissue intersected with a large haematoma (1000 ml) in the pericardial sac. Cardiac hypertrophy (556 g) was observed in the patient, though no other cardiovascular abnormalities were found. Histological analysis showed cystic medial necrosis of the ascending aortic wall.

A ruptured aneurysmal dilatation of the ascending aorta and the aortic arch without aortic dissection associated with AC is an uncommon cause of haemopericardium that has only been described a few times before. The case is discussed in relation to other reported cases and in the context of the present understanding of this condition.

1. Introduction

Aortic coarctation (AC) is a congenital condition characterized by aortic narrowing. We describe here a case of AC in an adult male that exhibited dilatation of the ascending aorta and the aortic arch, rupture and cardiac tamponade. The condition was undiagnosed during life but was diagnosed after death by autopsy.

2. Case study

A 46-year old man was found dead with fully developed rigour and lividity lying supine on the living room floor of his home. There was no sign of a struggle or a robbery. The deceased was an unmarried man who lived alone in a village house. According to his general practitioner, who had not seen him during the prior 6 months, the man was healthy, received no prescription medication and was not abusing alcohol or drugs.

Unenhanced post mortem computed tomography (PMCT) showed haemopericardium with a double-concentric stratification of the intrapericardial blood with the densest ring on the epicardial surface (Fig. 1). The ascending aorta and the aortic arch seemed to be dilated, but the vessel wall could not be discerned, and no inwardly displaced intimal calcifications or intimal flaps could be observed (Fig. 2). According to these findings, a dissecting aneurysmal rupture was suspected. PMCT measurements of the aorta performed after the autopsy showed that the diameter just distal to the left subclavian artery was 1.3 cm and that the diameter in the thoracic aorta was 2.3 cm. The distal arch (the isthmus) may be narrower than the proximal descending aorta, but not to this degree [1].

The autopsy showed a dilatation of the ascending aorta and the aortic arch (maximal diameter 4.5 cm), which was thin-walled and floppy, but without dilatation of the aortic root. A sharply demarcated, short coarctation with a diameter of 1.3 cm was found just distal to the left subclavian artery and proximal to the ligamentum arteriosum (Fig. 3). A 2.6-cm longitudinal tear was found in the posterior wall of the ascending aorta located approximately 1 cm from the valves (Fig. 4). There was blood in the surrounding soft tissues in connection with a large haematoma

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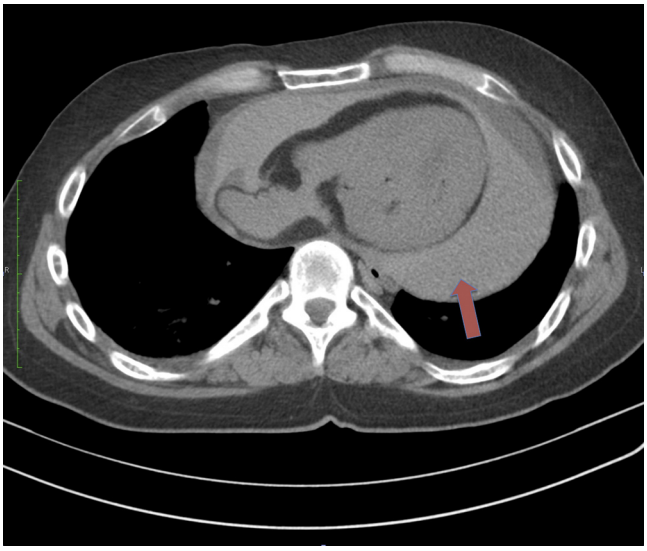

Fig. 1. Transverse PMCT image of the thorax demonstrating a large haemopericardium (arrow).

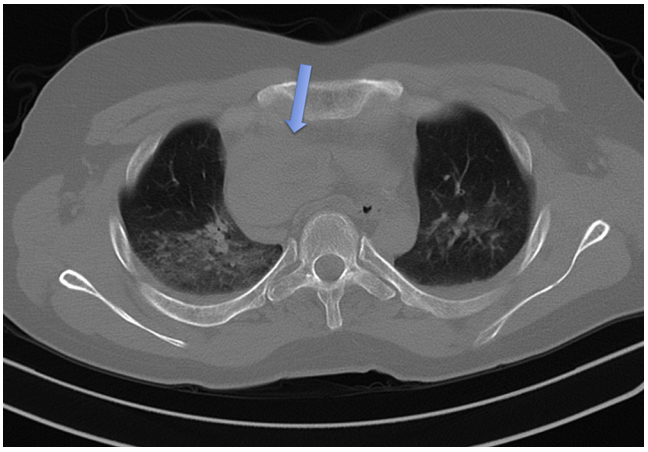

Fig. 2. Transverse PMCT image of the dilated ascending aorta (arrow).

(1000 ml), which was comprised of blood and clot, in the pericardial sac. There was no visible aortic dissection. The aorta distal to the site of coarctation appeared macroscopically normal with no post-stenotic dilatation or visible dilatation of the posterior intercostal artery ostia. Cardiac hypertrophy was observed (556 g) along with symmetrical concentric left ventricular hypertrophy (15 mm). No additional cardiovascular abnormalities were noted. The cardiac valves appeared normal; in particular, the aortic valves, which were tricuspid, showed no evidence of stenosis, and the aortic root was not dilated. There was no indication of coronary atherosclerosis, nor were there any signs of collateral circulation during the autopsy or inferior posterior rib notching observed by PMCT.

Microscopic examination was performed with four different staining protocols, including Haematoxylin and Eosin, Sirius, modified Verhoeff-elastin-Masson-trichrome, and periodic acid Schiff staining. The staining protocols showed marked abnormality of the thoracic aorta proximal to the site of coarctation with pronounced fragmentation of the elastic fibres, prominent pools of acidic mucosubstance and loss of smooth muscle cells in the media (Fig. 5). There was only slight dissection of blood within the media in the immediate vicinity of the rupture site. The myocardium was hypertrophic but did not show fibrosis. The kidneys showed no evidence of hypertensive changes. A fibroblast culture established from the Achilles tendon was not analyzed because the deceased had no close relations.

3. Discussion

This case study represents the first descriptive report of the use of unenhanced PMCT for investigation of a death caused by cardiac tamponade in an adult afflicted with AC. This complication of AC has rarely been reported in the forensic literature. De-Giorgio et al. [2] described the death of a 35-year-old woman during sexual intercourse from a cardiac tamponade caused by the rupture of an AC-related aortic aneurysm. Lynch et al. [3] described the death of a 17-year-old boy with an unremarkable medical history who had a dilated ascending aorta with a 3.5-cm ragged linear defect in the intimal surface; intramural and proximal tracking of blood to the region of the aortic valve resulted in rupture into the pericardial sac. The boy's heart was enlarged, weighing 522 g. There was an hourglass-shaped AC in his descending thoracic aorta near the ligamentum arteriosum. Histological analysis showed myxoid degeneration of the pre-stenotic aortic wall, and dissection of blood was observed within the outer third of the media.

AC has an estimated incidence of 1 in 2500 births [4]. Aortic coarctation is typically a discrete narrowing of the thoracic aorta just distal to the left subclavian artery. However, the constriction may be proximal to the left subclavian artery or rarely in the abdominal aorta. In some cases, coarctation presents as a long segment or a tubular hypoplasia [5]. Common symptoms of AC are dyspnoea, fatigue and cold feet. Important clinical signs are weakened or absent femoral pulse and systolic murmur [6]. The

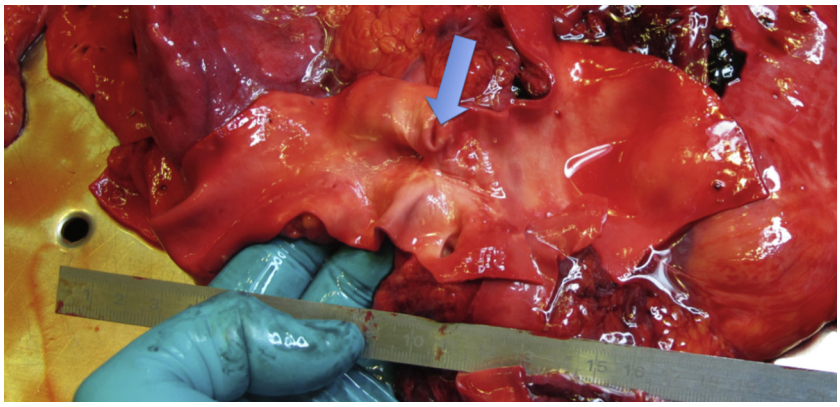

Fig. 3. The aorta after removal from the heart. The aortic coarctation (arrow) and the dilated aortic arch (to the right of the arrow) are visible.