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Images in Congenital Cardiac Disease

Massive haemoptysis and pulmonary arterial pseudoaneurysm in a patient with unrepaired tetralogy with pulmonary atresia

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THIS COMPUTED TOMOGRAPHY SCAN OF THE CHEST was performed as an emergency in a 33-year-old gentleman, with unrepaired tetralogy of Fallot. He presented to the emergency department with massive haemoptysis. In addition to pulmonary atresia, a large perimembranous ventricular septal defect, and overriding aorta, he had severe Eisenmenger's syndrome from major aorto-pulmonary collateral vessels. The maximum intensity coronal projection (Fig 1) shows an enlarged right-sided aortic arch of 4.2 centimetres maximal diameter (Ao), mirror image branching of the great vessels, and extensive aorto-pulmonary collateral arteries (C). Arising from the collateral to the left upper lobe is a 5.2 centimetre pseudoaneurysm (PsA) with surrounding ground-glass and centrilobular nodularity, consistent with acute pulmonary haemorrhage. On the axial reconstruction (Fig 2), there is a large rind (R) of soft tissue along the outer rim of the pseudoaneurysm, consistent with developing clot. The trachea is mildly compressed by the right-sided aortic arch. A 10-millimetre Amplatzer closure device (Am), placed for a similar presentation 5 years before, is visualised within a thrombosed, aneurysmal aortopulmonary collateral to the right lung. Given the patient's initial haemodynamic stability and reluctance for further intervention, he was treated conservatively. Unfortunately, despite aggressive non-invasive therapy, he slowly deteriorated over the ensuing week, eventually dying of progressive respiratory failure 9 days after admission.

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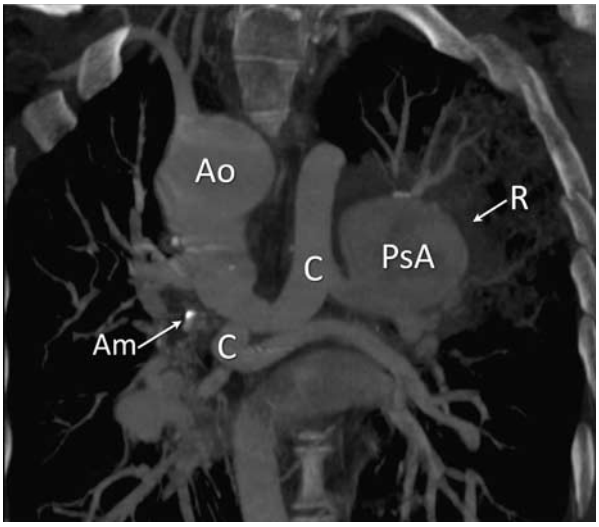

Figure 1.

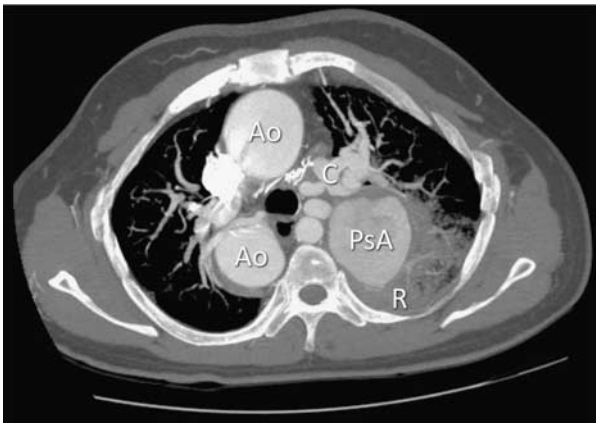

Figure 2.