Thrombus in coarctation of the aorta masquerading as an interrupted aortic arch

P Bonhoeffer, D Bonnet, D Sidi, J Kachaner

Coarctation of the aorta can be congenital or develop shortly after birth at the time of duct closure. The interrupted aortic arch is a different congenital heart disease both in its development during embryogenesis and in the anatomy of the aortic arch. Complete disconnection between the ascending and descending aorta is characteristic of interruption of the aortic arch. Atresia of the aortic arch is an extremely rare form of aortic coarctation in which the continuity of the aortic arch persists only through an imperforate fibrous cord.

Here we report an unusual case of intra-aortic thrombosis masquerading as an interrupted aortic arch in a newborn infant who was 12 hours old. Aortic coarctation was clinically suspected because of multiorgan failure in the absence of femoral pulses. The echocardiogram showed a horizontal and initial descending aorta of normal size but a mass in the aortic arch caused complete obstruction to flow at this level. The left ventricle was hypokinetic and the duct was closed at the time of examination. The interventricular septum was intact. A left ventricular angiogram was obtained by the transvenous approach with the catheter passed through the patent foramen ovale. The angiogram confirmed occlusion of the aortic arch after the origin of the brachiocephalic trunk and a large thrombus was identified in the aortic arch (figure). The lower body was supplied with blood only through thoracic collateral vessels, and the contralateral upper part of the body was perfused by retrograde flow from the left common carotid artery through the circle of Willis.

At the age of three days (3000 g) the child underwent surgery. Above a severe typical coarctation of the aorta, the surgeon found a large old thrombus extending from the isthmus back to the brachiocephalic trunk which obstructed the left subclavian and common carotid arteries. The thrombus was removed and the coarctation was successfully repaired. Despite heparinisation and thrombolysis, multiple venous thrombi developed, particularly in the inferior vena cava, leading to death 13 days postoperatively. Together with the aortic thrombus, these diffuse clotting complications were suggestive of an underlying constitutive haemostatic disease that could not be identified.

(A) Left ventricle angiocardiography in left anterior oblique projection. A Berman catheter was introduced into the left ventricle by using a transvenous approach. Note that the interventricular septum is intact. The ascending aorta is slightly dilated. A large thrombus (arrows) obstructed the horizontal aorta after the origin of the brachiocephalic trunk. (B) A zoom view of the area containing the thrombus shown in A.
Interruption to flow in the aortic arch caused by a thrombus is an extremely uncommon finding. An abnormal mass in the horizontal aorta can be identified by echocardiography. An underlying coarctation should be suspected. However, this can not be diagnosed with certainty either by ultrasound studies or angiography. Haematological investigations for the presence of clotting abnormalities should be undertaken.

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