

Aortic dissection presenting as posterior circulation stroke

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ABSTRACT

Aortic dissection is a rare and life-threatening event. While pain is the most common symptom of aortic dissection, it may be absent in ten percent of patients and present with a myriad of symptoms suggestive of a diverse range of other conditions. A high index of clinical suspicion is mandatory for the accurate and rapid diagnosis of aortic dissection. We report a 25-year-old woman with aortic dissection presenting as a posterior circulation stroke. This combination is very rare.

Keywords: aortic disease, aortic dissection, cerebellar infarct, cerebrovascular accident, chronic aortic dissection, posterior circulation stroke, stroke

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INTRODUCTION

Aortic dissection is a rare and potentially lethal disease, with an estimated incidence of 5–30 cases per million per year,⁽¹⁾ and a male-to-female ratio ranging from 2:1 to 5:1.⁽¹⁾ Classic aortic dissection is most often an acute event dominated by excruciating pain and other symptoms which suggest the diagnosis. Chronic aortic dissection, on the other hand, presents with atypical features which include painless dissections in conjunction with a variety of neurological or cardiac symptoms and signs. Neurological symptoms are commonly attributable to ischaemic cerebral infarcts, spinal cord ischaemia, ischaemic neuropathy and hypoxic encephalopathy.⁽²⁾ Infarcts of the vertebrobasilar territory in aortic dissection are very rare and have not been documented earlier. This article describes a case of painless chronic aortic dissection presenting as posterior circulation stroke in a young woman in her puerperal period.

CASE REPORT

A 25-year-old woman presented to our emergency service with complaints of headache and numbness of the left side of the face and extremities while going to bed. This was accompanied by dizziness, nausea, vomiting and slurring of speech. She arrived 12 hours after the onset of symptoms. She had no known history of diabetes mellitus, coronary artery disease, dyslipidaemia,

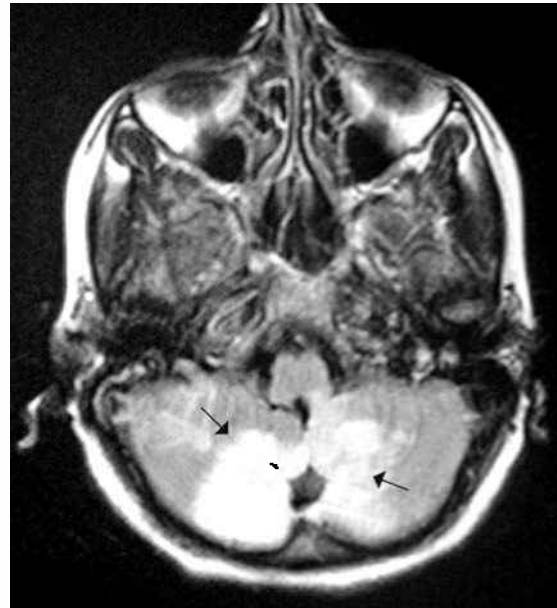


Fig. 1 Axial T2-W MR image of the brain shows areas of high intensity (arrows) involving both cerebellar hemispheres. This pattern of abnormal signal is consistent with an ischaemic lesion.

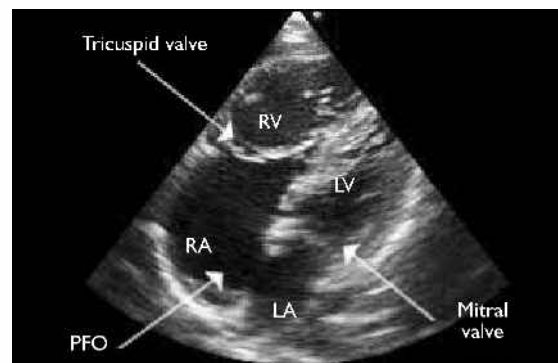


Fig. 2 Apical 4 chamber, 2-dimensional echocardiogram shows Ebstein's anomaly of the tricuspid valve. There is displacement of the tricuspid valve toward the apex of the right ventricle (RV) and a patent foramen ovale (PFO). LA: left atrium; LV: left ventricle; RA: right atrium.

connective tissue disease or any significant illness or hospitalisation, other than an uncomplicated full term normal delivery a month and a half prior. On admission, the patient was conscious and oriented, with a Glasgow coma score of 15. Her vital signs were: pulse 92/min and regular but non-palpable in the left radial and brachial arteries, blood pressure 140/110 mmHg in all limbs other than the left upper limb where it was not recordable, respiration rate 18/min with an oxygen saturation of 98% on room air and a body temperature of 36.7°C. Cardiac auscultation was significant for a wide and

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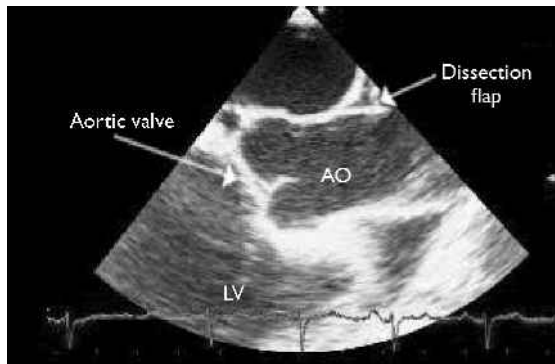


Fig. 3 Parasternal long axis echocardiogram shows a dissection flap in the ascending aorta (AO) posterolaterally and distal to the sinotubular ridge sparing the coronary arteries and aortic valve. LV: left ventricle.

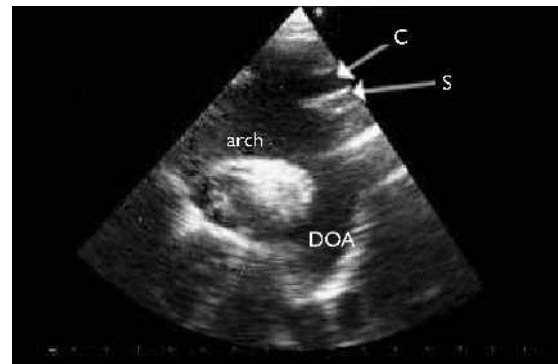
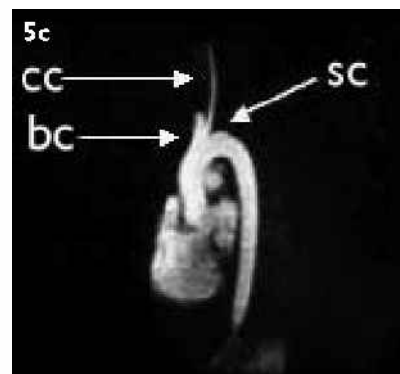
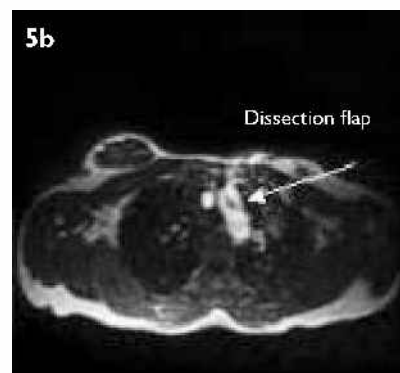


Fig. 4 Aorta as viewed from the echocardiogram at the suprasternal notch. The arch, left common carotid (C) and descending aorta are seen (DAO). Left subclavian artery (S) is not visualised. The expected location of the subclavian artery is shown by an arrow.



Fig. 5 (a) Transverse MR angiographic image taken at the level of the middle ascending aorta shows a dissection flap (arrow) in the ascending aorta with a normal descending aorta, consistent with a Stanford type A aortic dissection. (b) MR angiographic image taken through the aortic arch shows continuation of the dissection flap (arrow) into the arch of the aorta (c) MR angiographic image of the thoracic aorta and its branches. The emerging right brachiocephalic (bc) and left common carotid (cc) are made out. The left subclavian artery (sc) is not visualised due to obstruction of its origin by the dissection flap (arrow marked sc).



fixed splitting of the second heart sound, accompanied by an ejection systolic murmur in the pulmonary area, pan-systolic murmur in the tricuspid area and multiple clicks. Bilateral nystagmus, in the coordination of upper limb movements, gait apraxia-impaired tandem walking, dysarthria and normal muscle power were noted on neurological examination. Babinski's sign was absent bilaterally. The lungs were clear to auscultation. Abdomen was soft and non-tender with no organomegaly or distention. The remainder of the physical examination was unremarkable.

Cerebrovascular accident involving the posterior circulation of the brain was suspected. Magnetic resonance (MR) imaging of the brain revealed bilateral cerebellar

infarcts (Fig. 1). Colour Doppler ultrasonography of the left upper limb showed absent flow in the left subclavian and brachial arteries. Chest radiograph was insignificant except for cardiomegaly. Transthoracic echocardiography showed Ebstein's anomaly of the tricuspid valve (Fig. 2). A large sail anterior tricuspid leaflet along with apically displaced and septally plastered septal tricuspid leaflet, patent foramen ovale with right to left shunt and a low pressure tricuspid regurgitation were seen (Fig. 2). Both transoesophageal echocardiography and transthoracic echocardiography (Fig. 3) showed a dissection flap in the ascending aorta posterolaterally and distal to the sinotubular ridge, sparing the coronary arteries and aortic valve. Colour Doppler ultrasonography failed to

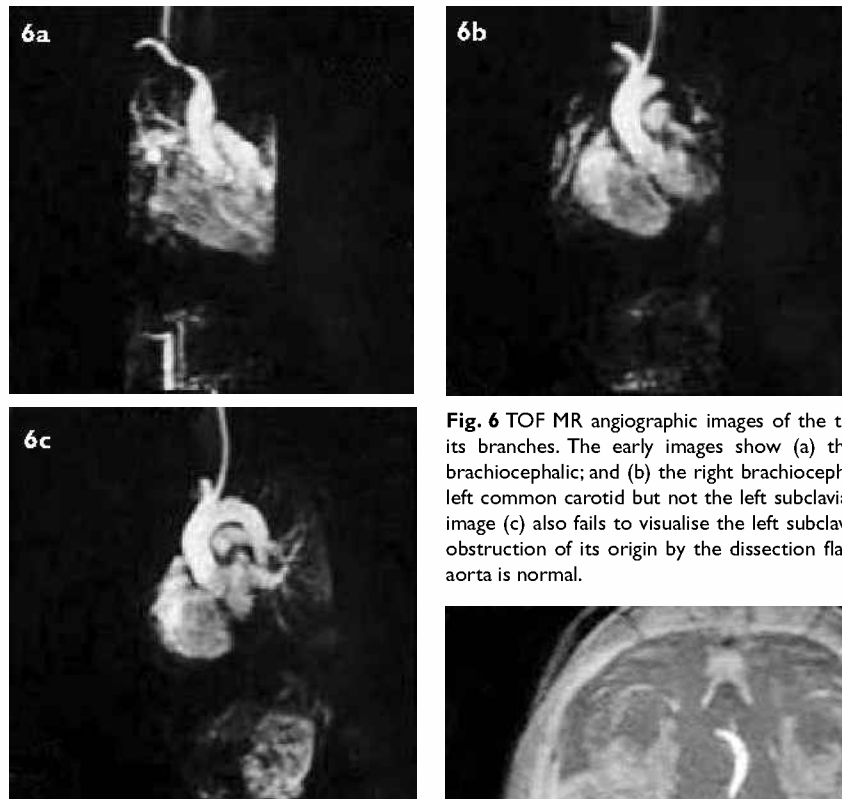


Fig. 6 TOF MR angiographic images of the thoracic aorta and its branches. The early images show (a) the emerging right brachiocephalic; and (b) the right brachiocephalic and emerging left common carotid but not the left subclavian artery. The late image (c) also fails to visualise the left subclavian artery due to obstruction of its origin by the dissection flap. The descending aorta is normal.

visualise the take-off of the left subclavian artery from the aortic arch (Fig. 4).

MR angiography (Fig. 5) supported the diagnosis of aortic dissection by showing a small linear flap in the right posterolateral aspect of the ascending aorta extending into the origin of the left subclavian artery and causing obstruction. The left subclavian artery (Fig. 6) and vertebral artery were not visualised (Fig. 7). Right subclavian and common carotid arteries were normal, as were the descending aorta and bilateral renal arteries. The findings were consistent with the diagnosis of a posterior circulation stroke possibly caused by obstruction of the origin of the left subclavian artery by extension of the aortic dissection flap (Stanford Type A dissection), thereby compromising the flow in the territory of the left vertebral artery. However, the possibility of an embolus originating from the dissecting flap and causing obstruction could not be ruled out definitely, as aortic angiography was refused by the patient.

Routine blood counts and chemistries were unremarkable except for a haemoglobin level of 8.2 g/dL. Electrocardiography showed sinus rhythm with right bundle branch block. The erythrocyte sedimentation rate was 50 mm/hr. The results of VDRL, antinuclear antibody, anti-phospholipid antibody, rheumatoid factor, antidouble-strand DNA, anti-SS-A and SS-B were all negative. The patient refused surgical management of



Fig. 7 TOF MR angiography of intracerebral blood vessels. Right vertebral artery (RVA) and basilar artery (BA) are shown. The left vertebral artery shows diminished flow and is not seen.

the aortic dissection, and hence was treated medically for blood pressure and heart rate control along with antithrombotic measures. The patient was discharged with a heart rate of 60/min and blood pressure of 100/60 mmHg. The patient was unfortunately lost to follow-up.

DISCUSSION

Posterior circulation stroke involves the vertebrobasilar system, which perfuses the upper cervical spinal cord, medulla oblongata, pons, midbrain, thalamus, cerebellum, occipital lobes and parts of the temporal and parietal lobes. They account for approximately 10-15% of all strokes, with ischaemia accounting for 80% of them.⁽³⁾ Large vessel atherothrombosis, lipohyalinosis of small arteries, embolic disease and dissections in the

extracranial vertebral arteries are presumed causes of stroke in the posterior territory.⁽³⁾ Migraine, fibromuscular dysplasia, coagulopathies, vasculitis and drug abuse are much less frequent causes.⁽³⁾ Chronic aortic dissection is not recognised in the literature as a cause of posterior circulation stroke.

Neurological ischaemic complications are associated with 18%–30% of aortic dissection cases and include anterior circulation stroke, lower extremity numbness, syncope, transverse myelopathy and hoarseness.^(2,4) Cerebral ischaemic stroke is the most common neurological manifestation associated with aortic dissection, affecting 5%–10% of patients, and arises either due to occlusion of the origin of the common carotid by the dissecting flap or artery to artery embolism from a thrombus developed on the intimal surface of the dissected artery.^(2,5)

To our knowledge, posterior circulation strokes in the form of cerebellar infarcts in aortic dissection have not been previously reported. Obstruction of the left subclavian artery at its origin, by either the dissecting flap seen on transoesophageal echocardiography and MR angiography or by a thrombus, most likely compromised the flow in the vertebral system in our patient leading to cerebellar infarcts. Insufficient contralateral flow in the opposite artery can account for the ischaemic syndrome produced by occlusion of a single vertebral artery. The possible explanation for the development of bilateral infarcts is however unclear, but may likely involve anomalies in the arterial supply.

Cystic medial degeneration is the main predisposing factor for aortic dissection.⁽⁶⁾ Chronic systemic hypertension is the main risk factor, followed by hereditary connective tissue disease (e.g. Marfan's syndrome and Ehlers–Danlos syndrome), coarctation of the aorta, bicuspid aortic valve, aortitis and arch hypoplasia.⁽⁶⁾ Hypertension and Marfan's syndrome are the most commonly occurring risk factors in cases of aortic dissection during pregnancy.⁽⁷⁾ Our patient, however, had no documented abnormal blood pressure throughout her pregnancy, no clinical evidence of hereditary connective tissue disease or other known predisposing congenital or acquired abnormalities for aortic dissection. Syphilis serology was normal. The role of pregnancy as an independent risk factor for developing aortic dissection in healthy women under the age of 40 years has been shown by autoptical studies and epidemiological data.⁽⁸⁾

Ebstein's anomaly in our patient was an incidental finding. To our knowledge, there are no reports of aortic dissection caused by it. In most cases, it is an isolated

defect and is rarely associated with extracardiac defects which most often involve the craniofacial region, central nervous system and limbs.⁽⁹⁾ Whether Ebstein's anomaly with extracardiac defects involves damage during a longer and earlier period or involves structural disorders of the arterial wall is something that requires further study.

The other interesting presentations seen in this patient were the complete absence of pain and absent peripheral pulses. Painless aortic dissection occurs in less than 10% of cases,⁽¹⁰⁾ and pulse deficits are seen in up to 30% of cases.⁽¹¹⁾ Multiple hypotheses have been advanced to explain the phenomena of a painless dissection, such as chronic dissection with less wall stretching and sparing of the adventitial layer which may not be associated with pain.⁽¹⁰⁾ However, the exact mechanism of painless dissection is still unknown. Patients presenting with a neurological disorder without any pain should thus also be suspected of having an aortic dissection, in addition to a cerebrovascular accident, as therapy with thrombolytics is harmful in the former.

This case highlighted the very rare but plausible causation of posterior circulation stroke by an ascending aortic dissection. We emphasise that aortic dissection should be carefully considered in all patients presenting with neurological symptoms, even if typical of posterior stroke, as the combination can present even if rare. Aortic dissection is a disease of many faces, thus requiring a high degree of clinical suspicion for an accurate diagnosis.

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