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IMAGES in PAEDIATRIC CARDIOLOGY

Devara K V A,[1] Joseph S,[2] Uppu S C.[3] Spontaneous Subarachnoid Haemorrhage Due to Coarctation of Aorta and Intraspinal Collaterals: A Rare Presentation. Images Paediatr Cardiol 2012;14(4):1-5
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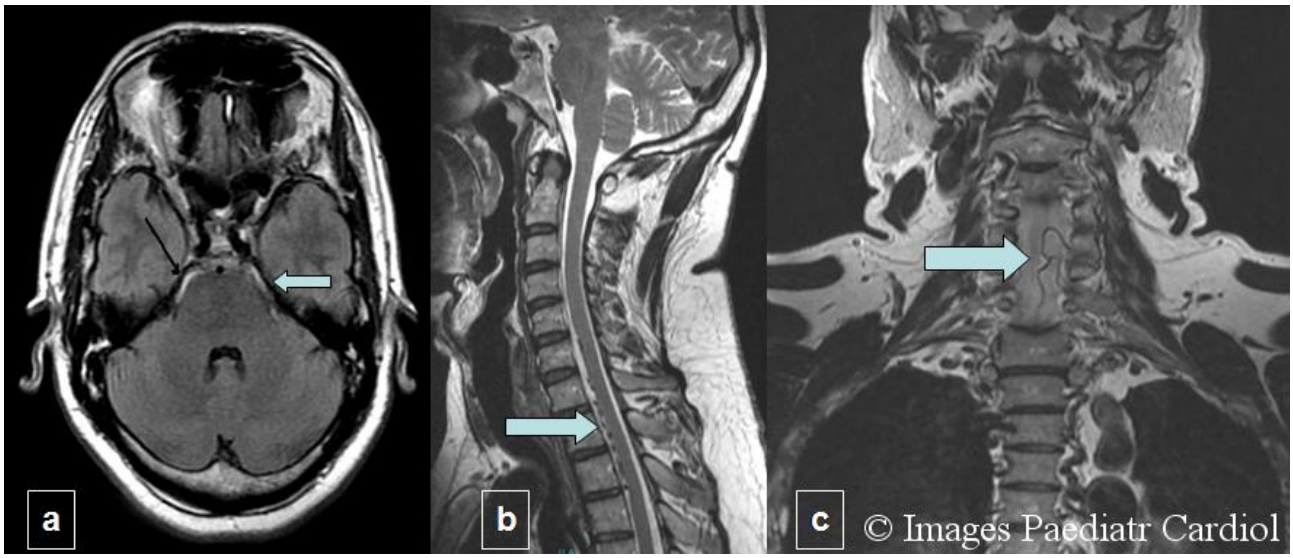
Key words: Subarachnoid haemorrhage, coarctation of aorta, intraspinal canal collaterals

Abstract
The occurrence of spontaneous subarachnoid haemorrhage (SAH) in association with coarctation of thoracic aorta and absence of intracranial aneurysm is a rare association. In spontaneous SAH, the predominant cause is intracranial aneurysmal rupture. This report describes a case of a 40 year-old male who presented with SAH and was incidentally diagnosed to have coarctation of aorta (CoA) with intraspinal collaterals on further work up. This case demonstrates the importance of detailed evaluation of patients with spontaneous SAH on whom common aetiologies have been ruled out.

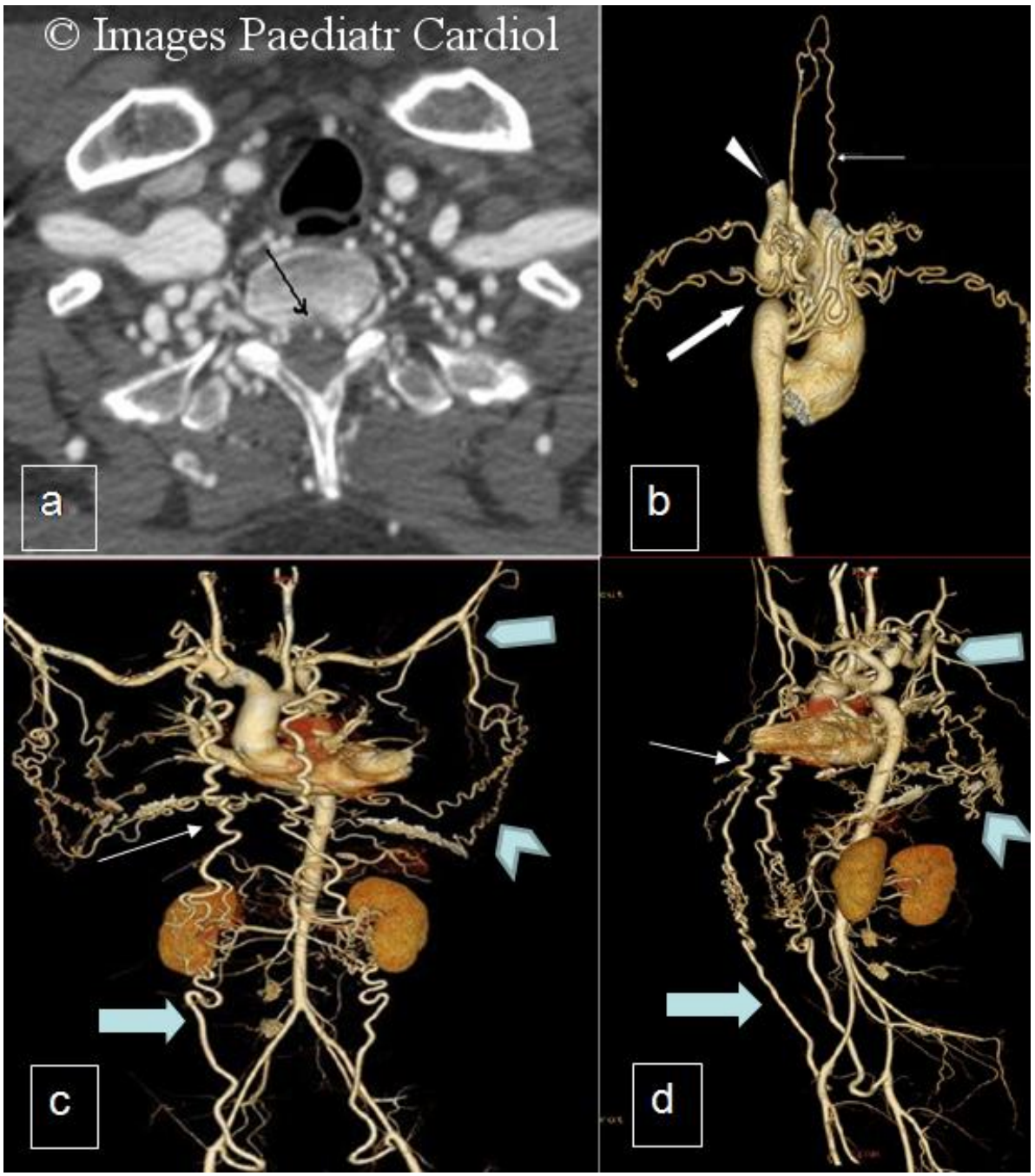
Case report
A 40 year old hypertensive male presented with sudden onset of severe neck pain, mild giddiness and numbness of bilateralupper extremities that improved spontaneously within two hours. There were no motor deficits, bladder or bowel disturbances. General examination revealed a differential blood pressure (BP) measurement with upper extremity BP of 150/90 mmHg and lower extremity BP of 130/80 mmHg. Cardiovascular examination revealed prominent brachiofemoral pulse delay, and a continuous systolic murmur in the interscapular area. Other systems were unremarkable. Routine laboratory investigations were normal. Chest radiograph revealed inferior rib notching of bilateral lower ribs. Electrocardiogram revealed left ventricular hypertrophy. Echocardiogram showed normal chambers, bicuspid aortic valve, trivial aortic regurgitation without aortic stenosis, and normal ejection fraction. The patient underwent a Magnetic Resonance Imaging (MRI) examination of brain and cervical spine, which revealed subarachnoid hemorrhage (SAH) in the prepontine cistern (Fig 1a) and prominent flow voids in the cervicothoracic subarachnoid space (Fig 1b & 1c). Narrowing of the thoracic aorta just distal to origin of left subclavian artery (aortic isthmus) was suspected in visualized aortic segment in the sagittal images of cervical spine.

Figure 1: A. Magnetic Resonance Imaging Axial flair image of brain shows subarachnoid hemorrhage (block arrow) in prepontine cistern. B: Magnetic Resonance Imaging Sagittal T2 weighted image of cervical spine showing intraspinal extramedullary collateral (block arrow). C: Magnetic Resonance Imaging Coronal T2 weighted image of cervical spine showing intraspinal extramedullary collateral (block arrow).



A Computerized Tomographic (CT) aortogram was performed to evaluate further, that revealed a prominent intraspinal extramedullary collateral arising from the left subclavian artery, coursing through the spinal canal ventral to the lower cervical and upper thoracic spinal cord from C5 to D5 vertebral level and terminates in the left 3rd intercostal artery (Fig 2a & b) anda short segment severe aortic stenosis with extensive collaterals between the pre-stenotic and post stenotic aortic segments along the anterior, posterior aspects of the chest and the abdominal walls (Fig 2 c&d). Left vertebral artery was seen separately. CT cerebral angiogram revealed no intracranial aneurysms. He was managed conservatively during which his pain subsided and was subsequently discharged with a planned coarctation of aorta (CoA) repair.

Figure 2: A. Contrast-enhanced computed axial tomographic image at the level of neck shows intraspinal collateral (black arrow) with multiple posterior wall collaterals. B. Volume rendered computed tomographic image showing the course of the intraspinal collateral (small arrow) arising from left subclavian artery (arrowhead) and terminating at the left third intercostal artery with coarctation of aorta (block arrow). C and D: Volume rendered computed tomographic image in coronal (2c) and oblique sagittal (2d) planes showing extensive thoracic collaterals supplying descending thoracic aorta via subscapular arteries (pentagon), intercostals arteries (chevron) and thoracoabdominal collaterals supplying the external illiac arteries via the internal mammary (small arrow) , inferior epigastric arteries (block arrow) with increased caliber of distal external iliac and common femoral arteries on both sides.



Discussion

CoA most commonly affects the aortic isthmus. These patients can present either in infancy or adulthood. Cerebrovascular complications of CoA include spontaneous SAH, intracerebral hemorrhage and cerebral abscess.

The incidence of spontaneous SAH is reported to be around 10.5 per 100,000 person years.[1] The leading cause of spontaneous SAH is the rupture of intracranial aneurysm, which accounts for about 80 percent of cases and has a high rate of death and complications.[1] In patients with CoA, intracerebral or subarachnoid hemorrhage is more common in the second and third decades. The risk of serious neurological complications associated with CoA and the resultant proximal hypertension mandates early diagnosis and treatment. Berry aneurysms are reported to occur in 10% of patients with CoA and this has been related both to high pressure of the vascular tree proximal to the coarctation and to congenital defects of the vascular tree.[1,2] Dilated collateral arteries within the spinal canal also occur in CoA, and these may compress the spinal cord resulting in paraparesis or may rupture and presents with clinical picture of SAH.[3-5]

Our patient was previously diagnosed as a primary hypertensive with no identifiable cause, presented with complaints of sudden onset spontaneous neck pain. His initial workup with MRI brain and cervical spine showed SAH and incidental CoA with intraspinal collaterals that was further characterized with CT Aortogram. The high incidence of berry aneurysms in patients with CoA is well known but none were seen in this case.[1,6] Banna et al.[5] reported a case of spinal SAH in a patient of CoA where post mortem examination revealed a ruptured pseudoaneurysm arising from the collaterals. However, we couldn't demonstrate any obvious aneurysm arising from the collaterals on the CT angiogram in our patient. Only one case of isolated SAH associated with this condition in the absence of cerebral aneurysm has been reported earlier.[7]Nonaneurysmal SAH differs from aneurysmal SAH in initial presentation, with fewer neurological consequences and less severe sequelae, which correlates with our patient.[8] We suspect spontaneous rupture of intraspinal collaterals might have resulted in SAH in our patient. A cerebral angiography would have been ideal to rule out the intracranial aneurysms, however CT angiogram is increasingly been used in the evaluation of SAH with high sensitivity and specificity.[9]This case demonstrates the importance of detailed systemic evaluation of patients with spontaneous SAH on whom common aetiologies have been ruled out.Evaluation of the intraspinal collateral circulation as a possible source of SAH must be kept in mind in patients with CoA and negative cerebral angiogram.

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