Ruptured Anterior Spinal Artery Aneurysm Associated with Coarctation of Aorta
Case Report and Literature Review

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Key words: anterior spinal artery aneurysm, coarctation of aorta and collateral circulation, spinal subarachnoid haemorrhage

Summary

A 39-year-old man presented with acute headache and neck pain, followed by quadriparesis and quadriplegia, accompanied by urinary and bowel incontinence. Lumbar puncture showed subarachnoid haemorrhage. Angiogram via a right axillary approach revealed severe coarctation of the aorta, between the left common carotid artery and left subclavian artery. Multiple collateral circulation including an enlarged anterior spinal arterial axis bridging the stenosed arch provided collateral circulation to the abdominal aorta. A small lobulated aneurysm was seen at the radiculomedullary-anterior spinal artery junction from the right ascending cervical artery. This patient underwent successful surgical clipping of the aneurysm. Pathogenesis of the spinal arterial aneurysm associated with coarctation of the aorta is likely to result from the haemodynamic stress from collateral circulation through the anterior spinal axis rather than segmental arterial disease or angiodysplastic disease. Aneurysms of the spinal artery are rare but can be unusually found in association with SCAVMs, coarctation of aorta, Klippel-Trenaunay syndrome or more rarely with aortic arch interruption.

Introduction

Subarachnoid haemorrhage (SAH) due to a lesion within the spinal canal is seen infrequently. It is estimated that less than 1% of all patients with SAH have lesions within the spinal canal; and the most common etiology is spinal cord arteriovenous malformations.

Arterial aneurysms of the spinal arteries are rare but can be rarely found in association with SCAVMs and coarctation of aorta or more seldom with aortic arch interruption. Klippel-Trenaunay syndrome is known to be associated with multiple dysplastic aneurysms of the radiculomedullary artery and SCAVMs.

We report a case of ruptured aneurysm of the anterior spinal artery associated with severe coarctation of the aorta.

Case Report

A 39-year-old, previously healthy male farm worker developed acute neck pain followed by dull headache after carrying a heavy burden for several hours. This pain radiated to both upper extremities and was followed by slight weakness of both hands. It subsequently radiated to both arms and both legs gradually before he fi-
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and respiratory rate 20/min. Cardiopulmonary auscultation appeared normal. Neurological examination showed paresis grade III/V of upper extremities and grade 0/V of lower extremities. There was lost sensation below 2nd thoracic level and lost rectal sphincter control. No cranial nerve palsy was detected.

Lumbar puncture showed bloody liquor from SAH.

Plain chest film (figure 1) showed evidence of rib notching with enlarged aortic knob and suspected inverted three sign of coarctation of aorta (retrospectively reviewed). MRI cervical spine, (figure 2), was suspicious for cervical spinal cord arteriovenous malformation (SCAVMs). He was then referred to our institute for further management.

The first angiogram by femoral arterial approach failed to pass the thoracic arch with blockage at a narrowed segment of the distal aortic arch (figure 3); angiogram by right axillary approach revealed severe coarctation of aorta, seated between the left common carotid artery and left subclavian artery (figure 4).

Numerous collateral pathways were demonstrated including right internal mammary artery (figure 4B), vertebrobasilar anastomosis with subclavian “steal”, muscular branches of the vertebral arteries and ascending cervical artery bilaterally (figure 5), across the midline and also dilated anterior spinal arterial axis (figure 6). The artery of the cervical enlargement arises from the right ascending cervical artery and fills retrogradely the radiculomedullary artery from the left 6th intercostal artery (figure 7). Enlargement of this cervicothoracic anterior spinal axis, represented one pathway of collateral circulation giving blood supply to the lower aorta below the coarctation through the thoracic radiculomedullary artery.

There was a small lobulated aneurysm at the radiculomedullary junction of the right ascending cervical artery and cervical ventral spinal axis (figure 6), measuring about 1.23 x 1.3 cm with the tip pointing superiorly, corresponding to the ruptured aneurysm and causing clinical SAH.

The patient underwent surgery with C4-6 laminectomy and successful clipping of the aneurysm. At follow-up, the neurological symptoms have not fully abated. Despite cardiosurgical consultation, the patient refused surgical treatment for his coarctation.
Discussion

Sudden excruciating pain is one of the hallmarks of intraspinal haemorrhage, so called “coup de poignard” (translated “stuck by a dagger”) and should alert the clinician to the possibility of intraspinal haemorrhage as one of differential diagnosis.

For SAH, location of the aneurysm is also an important factor determining symptoms and signs of ruptured spinal artery aneurysms. For patients with a lesion located in the thoracic spine, the chronological sequence of back pain, paraspinal muscle spasm, and a few hours later followed by cranial symptoms such as headache, photophobia and neck stiffness is a hallmark of spinal haemorrhage in the thoracic region, whereas the lesion in the cervical spine

Table 1  Reported Cases of Spinal Artery Aneurysm

<table>
<thead>
<tr>
<th>Associated condition</th>
<th>Series(ref. No.)</th>
<th>Location (N)</th>
<th>Sex/Age (years/mean)</th>
<th>Symptoms and signs</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Coarctation of aorta</td>
<td>This report</td>
<td>Cervical (1)</td>
<td>M/39</td>
<td>Quadriplegia, SAH</td>
<td>Surgical clipping</td>
</tr>
<tr>
<td></td>
<td>Rengachary SS^7</td>
<td>Cervical (1)</td>
<td>F/45</td>
<td>Quadriplegia, neck stiffness</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>Tsutsumi K^7</td>
<td>C4-5 (1)</td>
<td>F/71</td>
<td>Right leg paresis</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>Hino H^10</td>
<td>Cervical (1)</td>
<td>F/45</td>
<td>Quadriplegia</td>
<td>NA</td>
</tr>
<tr>
<td>2. Aortic interruption</td>
<td>Feng L^4</td>
<td>Cervical (1)</td>
<td>M/14</td>
<td>Paraplegia, SAH</td>
<td>Bypass surgery of aortic interruption</td>
</tr>
<tr>
<td>3. Bilateral vertebral artery occlusion</td>
<td>Kawamura S^5</td>
<td>High cervical (1)</td>
<td>M/42</td>
<td>Severe Headache, SAH</td>
<td>Surgical clipping</td>
</tr>
<tr>
<td>4. SCAVMs</td>
<td>Rengachary SS^7</td>
<td>Cervical (10)</td>
<td>M(5), F(4), NA(1)</td>
<td>Headache with tri-quadriparesis</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>Rengachary SS^7</td>
<td>Thoracic (3)</td>
<td>M (2)/F (1)</td>
<td>Abdominal and back pain</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>Thoracolumberal (6)</td>
<td></td>
<td>M(2),F(4)</td>
<td>Back pain and paraparesis</td>
<td>NA</td>
</tr>
<tr>
<td>5. Arteritis and inflammatory disease</td>
<td>Rengachary SS^7</td>
<td>Thoracic (2)</td>
<td>F (2) 48 and 50</td>
<td>Headache, neck pain, quadriparesis</td>
<td>NA</td>
</tr>
<tr>
<td>6. Pregnancy</td>
<td>Rengachary SS^7</td>
<td>Thoracic(1)</td>
<td>F/34</td>
<td>Headache, paraplegia</td>
<td>NA</td>
</tr>
<tr>
<td>7. Isolated spinal arterial aneurysm</td>
<td>Smith BS^4</td>
<td>NA</td>
<td>NA</td>
<td>SAH</td>
<td>Surgical treatment Surgery by posterior approach</td>
</tr>
<tr>
<td></td>
<td>Moore DW 15-16</td>
<td>C1(1)</td>
<td>M/29</td>
<td>SAH, R.hemiparesis and 6^5 nerve palsy</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>Vincent FM^6</td>
<td>NA</td>
<td>NA</td>
<td>SAH</td>
<td>NA</td>
</tr>
</tbody>
</table>

NA, information not available.

^with anomalous arrangement of the arteries without SCAVMs
may simulate intracranial aneurysm and lesions in the lower lumbar area may not exhibit any cranial symptoms.

Neurological signs and symptoms become an important criterion to specify intraspinal location of disease. Neurological deficits such as paraplegia or paresthesia, point to spinal cord pathology itself. SAH from an intracranial lesion frequently leads to computed tomogram (CT scan) and diagnostic cerebral angiogram. In contrast, CT or post-myelos CT are not sensitive for detecting the lesion in the spinal canal. A myelogram in this situation could demonstrate various findings such as nodular filling defects from blood clots in the subarachnoid space or aneurysm itself or enlargement of the spinal artery or blockage of the contrast column. MRI is now considered to be the most useful non-invasive investigation to detect and localize lesions in the spinal canal, such as

Figure 3  Aortogram from femoral arterial approach shows obstruction of the distal aortic arch.

Figure 4  Arch aortogram done from a transaxillary approach shows severe aortic arch coarctation between the left common carotid artery and left subclavian artery with complex collateral flow through an enlarged right internal mammary artery and anterior spinal axis with aneurysm.
haematoma, SCAVMs nidus, aneurysm, and abnormal enlargement of the spinal artery and medullary vein. The evidence of congestive myelopathy is very useful for follow-up study.

In a patient with proven SAH of spinal origin, even though the MRI and myelogram are negative, there is a possibility of an aneurysm or small malformation, one should consider spinal angiography.

There are few reports of aneurysms in the spinal arterial system. Rengachary\(^2\) reported a high proportion of patients with spinal artery aneurysm presenting with neurological symptoms (20/40 from all 57 patients) and most cases have more than one presenting symptom (table 1). Clinical manifestations of the spinal artery aneurysm are also interesting and unique despite their rarity, including the common severe neck or back pain, SAH\(^2\), neurological deficit and rarely compressive myelopathy from mass effect\(^6\). Mass effect as a presenting symptom appears to be rare. There is a report by Tsutsumi of a 71-year-old woman with aneurysm of a radiculomedullary artery at 4\(^{th}\) cervical level, associated with coarctation of aorta. She presented with Brown-Sequard syndrome from compressive myelopathy of the spinal cord\(^7\).

Cerebral angiogram can be negative when only a ruptured spinal artery aneurysm is present but it can be positive if there is a coexisting intracranial aneurysm (10%), which probably leads to misdiagnosis of the spinal aneurysm. Rengachary\(^2\) showed that most of the reported aneurysms are no larger than 3 mm in diameter, hence the fact that detailed and careful spinal angiography remains the gold standard of vascular imaging for specific diagnosis and for guidance of surgical intervention.

Djindjian\(^12\) found aneurysmal dilatation of the spinal artery in 18 patients with SCAVMs (20%) with pathological study in eight cases and angiographic study in ten. Isolated aneurysm of the spinal artery without other vascular malformations is exceptionally rare. Djindjian\(^11\) discovered only one isolated spinal aneurysm from 1000 spinal angiogram in his review series. There are few reported cases of isolated spinal aneurysm in the literature\(^14-16\).

Other etiologic factors of spinal artery aneurysm (table 1). Rengachary\(^2\) found a case of isolated spinal artery aneurysm and reviewed 57 cases of spinal artery aneurysms. The aneurysms were classified according to associated lesions like SCAVMs (25/57), coarctation of aorta (4/57), syphilis and arteritis (3/57), fibromuscular hyperplasia (1/57), Pseudoxanthoma Elasticum (1/57) and idiopathic (23/57). This suggests that spinal cord arterial aneurysms more commonly occur by haemodynamic stress from increased flow situations or collateral pathways than developmental arterial disease, arterial wall disease or inflammatory lesion. Another interesting fact is the mean age of patients with spinal arterial aneurysm at presentation when related to SCAVMs. In thoracolumbar and conus medullaris locations, the age of onset of symptoms is earlier than in other locations (table 1).
The incidence of all forms of aneurysm formation in aortic coarctation, markedly increases with age, up to 42% in patients over the age of forty. Feng reported a case of aortic arch interruption with myelopathy and multiple aneurysms of radicular arteries and distal vertebral artery that presented with acute SAH. Caroscio, in a review of the literature up to 1980, found 12 patients with arterial dilatation of the spinal arteries associated with coarctation of the aorta; only three were isolated aneurysm out of 30 aneurysm cases. Intercostal aneurysms have also been reported in up to 10% of patients with aortic coarctation; subclavian aneurysms are another rare associated finding. Ruptured aneurysms around the circle of Willis are seen in 10% of patients with coarctation of aorta whereas associated spinal artery aneurysms were sparsely reported.

Pathogenesis of the spinal arterial aneurysm associated with coarctation of the aorta appears to result from haemodynamic stress related to increased blood flow into the collateral circulation through the anterior spinal axis rather than segmental arterial disease or angiodysplastic disease.

Multiple routes of collateral circulations contribute to restore flow into the descending aorta below the coarctation from the contralateral subclavian artery, including the intercostal artery, internal mammary artery, bronchial artery, internal carotid artery and verterbrobasilar artery around circle of Willis and uncommonly the spinal arterial axis. In fact, there are also some reported cases of arterial aneurysms along the other collateral pathways which are not related to the same embryological origin such as the intercostal artery, subclavian artery and circle of Willis as mentioned above. Kawamura reported a case of spinal arterial aneurysm associated with bilateral vertebral arterial occlusion whereas the vertebrobasilar system was supplied by a dilated anterior spinal axis. These reports support the fact that spinal artery aneurysms along the collateral pathways for arterial occlusion should belong to the flow related group of aneurysms.

Angiogram in coarctation of the aorta usually fails if the puncture site is caudal to the coarctation such as the femoral artery. So retrograde angiogram through the axillary or brachial approach is mandatory.

Coarctation of aorta is a congenital condition known to shorten life expectancy. Untreated patients have an average life expectancy of thirty-two years, because of related complications such as heart failure, endocarditis, cerebrovascular haemorrhage and aortic rupture. About 25.5% of patients with untreated aortic coarctation die of cardiac failure. But some of the patients can survive normally to adulthood without any specific symptoms and the coarctation is discovered incidentally.

Hypertension of one or both upper extremities depending on type of coarctation is one of the key diagnostic symptoms. The Aortic arch is not the only location that can be involved by disease of coarctation. Iwata et al reported a case of abdominal coarctation presenting with spinal haemorrhage.

Even arterial aneurysm presenting with sub-
arachnoid haemorrhage is improved by endovascular treatment of SCAVMs with a decrease in size of the arterial aneurysm. However, in the treatment of the ruptured aneurysm associated with coarctation of the aorta, a surgical approach of clipping the aneurysm with immediate anatomical correction of the disease best prevents subsequent subarachnoid haemorrhage. Rupture of an arterial aneurysm from the anterior spinal axis can sometimes be documented as a pseudoaneurysm and a change in size of such aneurysm represents an unstable situation requiring immediate treatment.

Surgical treatment or correction of coarctation of aorta, was first performed in 1944. The younger the patient (especially below six months), the greater the possibility of residual or recurrent coarctation and the lower the probability of systemic hypertension (10% compared with patients treated in older age, 19.9-68%).

Figure 7 Supraselective angiogram of the right ascending cervical artery shows a dilated anterior spinal axis contributing to collateral circulation through lower radiculomedullary artery at left T6, supplying the lower descending aorta below the coarctation.
References


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