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ANTERIOR SPINAL ARTERY SYNDROME COMPLICATING AORTIC DISSECTING ANEURYSM: CASE REPORT

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SUMMARY

Spinal Cord ischaemia is rare in the absence of trauma. We report a case of a 45 year old known hypertensive for six years, who presented with features of anterior spinal artery syndrome (ASAS) complicating acute dissection of the descending aorta. He developed sudden onset non-traumatic paraparesis, sphincteric dysfunction and dissociated anaesthesia with a sensory level at T6. This was preceded by a two weeks' history of severe, sharp, lancinating, tearing left parasternal chest pain radiating to the back. He was managed conservatively on pentazocine lactate (fortwin), calcium- and beta-blockers, steroids, anti-platelet and free-radical scavengers. On the 8th day of hospitalisation, he had a sudden abdominal distension, bled from the nose and mouth, went into hypovolaemic shock and died within a time frame of two minutes. He was presumed to have had a progression of the aortic dissection with subsequent rupture. Dissecting aortic aneurysm could run a benign asymptomatic or a lethal course and a high index of suspicion is necessary. The lack of exhaustive diagnostic investigative tools as well as surgical intervention in the management of this patient in a developing country was highlighted as was possible that the patient could have been mismanaged.

INTRODUCTION

Spinal cord ischaemia is rare in the absence of trauma. It is even more uncommon that an infarction of the spinal cord occurs in healthy patients with no obvious predisposition to vascular disease(1). In the United States and world wide, only fragmentary or indirect data are available on its incidence or prevalence. Spinal stroke constitutes approximately 1.2% of strokes(2), and it is seen in 2 - 4% of patients with Aortic dissection(3).

It occurs commonly as a result of injury to the artery of Adamkiewicz (arteria magna radicularis) and continues to constitute a diagnostic challenge. The diagnosis is easy when patients present with classical features of non-traumatic acute tetra /para-paresis/ plegia, sensory level with dissociated anaesthesia as well as sphincteric dysfunction(4). As is the case for the more common cerebrovascular accident affecting the cerebral circulation, an acute onset is of paramount importance. However, unlike cerebral stroke, which is painless, 80% of patients with spinal stroke present with pain(2). There is also, no established relationship with age, sex and race(5).

CASE REPORT

A.O. - a 46 year old known hypertensive for six years with poor drug compliance presented with two weeks' history of severe, sharp, tearing, lancinating left parasternal chest pain radiating to the back, and a day history of sudden onset weakness and numbness of both lower limbs. There was accompanying constipation and urinary retention without loss of sensation of bladder fullness. He was not diabetic and denied history of smoking or alcohol consumption. No history of trauma to the back, preceding or associated fever, headache, vomiting, blurring of vision, seizures nor loss of consciousness and there was no family history of the presenting complaints. He was an international trader along the West African coast with positive history of prolonged sitting during road travels.

Examination revealed a middle aged male, acutely in distress - groaning in pain (chest). He was afebrile, anicteric and not pale. Chest was clinically clear. The cardiovascular system examination revealed a pulse rate of 144 beats per minute, regular, moderate volume and there was no radio-radial delay. The arterial wall was thickened with locomotor brachialis and the blood

pressure was 130/100 mmHg supine and standing. The jugular venous pulsation was not raised. The precordium was not active and only heart sounds I and II were heard. The A2 was loud but no murmur of aortic incompetence was appreciated. Gastro-intestinal examination revealed no epigastric tenderness. Central nervous system examination revealed normal mentation and cerebation with intact cranial nerves. He had flaccid paraparesis with power of 2/5 in the right lower limb and 3/5 in the left lower limb. The planter reflex was flexor bilaterally and he had hypoesthesia and hypoaesthesia with a sensory level at T6. The joint position and vibration sensations were intact. By the 3rd day of hospitalisation, sphincteric function had returned, power was 3/5 on the right and 4/5 on the left lower limb with spasticity and extensor plantar response on the left foot.

Table 1

Aetiology of anterior spinal artery syndrome (5)

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- Aortic surgery - 25%
 - Systemic arteriosclerosis- 19.4%
 - Acute deficit of perfusion - 11.1% - as in complication of coronary bypass grafting.
 - SLE, Primary antiphospholipid syndrome; Protein S deficiency; protein C deficiency
 - Epidural analgesia and left celiac plexus block
 - Transthoracic oesophagectomy
 - Paradoxical embolism in patent foramen ovale (PFO) and embolism from nucleus pulposus (fibro-cartilaginous) with retrograde flow via veno arterial anastomosis.
 - Severe stenosis of the vertebral artery
 - Post - anterior spinal fusion surgery
 - Cocaine abuse and association with oestrogens and the 20210A allele of the prothrombin gene.
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His cholesterol was 256 mg/dl. Haemogram revealed mild leucocytosis of 9,000/mm³ with neutrophilia of 80%. The haematocrit was 12g and the platelet count was normal. The electrolytes, creatinine and urea were within normal reference range. VDRL and HIV screening were negative. The prothrombin and activated partial thromboplastin time were not done. Chest X-ray revealed widened superior mediastinum, aortic unfolding with evidence of calcification and a paravertebral soft tissue shadow extending over the thoracic and lumbar spines. ECG revealed left ventricular hypertrophy without ischaemic changes. Transoesophageal echocardiography, aortography, Doppler ultrasound, chest CT scan, digital subtraction angiography, magnetic resonance angiography as well as CT scan myelography and magnetic resonance imaging of the spine were not done because of unavailability. An assessment of anterior spinal artery syndrome secondary to dissecting aortic aneurysm was entertained.

He was managed conservatively with propranolol titrated against the pulse rate, a calcium channel

blocker- amlodipine, free radical scavengers, low dose aspirin, pentazocine (fortwin) and prednisolone with initial improvement in his neurological status. On the 8th day of hospitalisation, he had a sudden abdominal distension, bled from the mouth and nose, went into hypovolaemic shock and died within a two minute time frame. The relatives did not grant approval for post-mortem examination. The aneurysm was presumed to have ruptured but paracentesis abdominus / paracentesis thoracis for haemoperitoneum / haemothorax respectively was not done.

DISCUSSION

Anterior spinal artery syndrome (ASAS) complicating aortic dissecting aneurysm is not uncommon(5) and it could run a benign asymptomatic course for many years(5,6). We reported a poor outcome of ASAS. in a middle aged hypertensive male with aortic dissecting aneurysm. Apart from hypertension, hypercholesterolaemia and prolonged sitting during international road travels, he had no other risk factors for arteriosclerosis such as diabetes mellitus or smoking. The possibility exists in this patient that he developed arteriosclerosis with subsequent fracture of an arteriosclerotic plaque which extended to the aortic wall and precipitated the onset of the dissecting aneurysm. Occlusion of the anterior spinal arteries by the false lumen created by the dissection of the aortic wall could thus predispose to ASAS. It is also possible that he had a pre-existing/congenital weakness or aneurysm of the wall of the aorta, which subsequently ruptured consequent on the poor blood pressure control in the face of systemic arteriosclerosis. The treatment in this patient was expectative and conservative rather than surgery. We took into consideration the high morbidity and mortality rate in surgically treated patients(6) as well as the report of a middle-aged woman with Stanford type A dissection who survived for 25 years without operation(7). Thus, it seemed justified for this conservative approach in the management of this patient. The death of the patient was thus, an extreme of the condition, although, if left untreated, progression to renal failure, bowel infarction and limb loss were other likely complications(8). However, because of the limited investigated tools, it was possible that the patient could have been mismanaged. Aetiology of anterior spinal artery syndrome(5).

Clinical neuroanatomy and physiopathology of the anterior spinal artery syndrome(ASAS):

The neuroanatomy and physiopathology of ASAS is well understood. Majority of spinal cord lesions can be categorised into three distinct patterns: diffuse (Transverse myelopathy), hemi-(Brown-sequard) and anterior (ASAS)(9). The main arterial supply of the spinal cord arises from the aorta while its cephalad and caudal ends receive from tributaries of the subclavian and iliac arteries respectively. Eight to ten unpaired

anterior medullary arteries are branches of the larger afferent aorta, vertebral and iliac arteries. The single anterior spinal artery forms long anastomotic channels that lie at the mouth of the anterior central sulcus and supplies circulation to the anterior 2/3 rd of the spinal cord. It gives origin to sulcal arteries that take an arching course on both sides of the anterior grey horns and supplies structures of the anterior and lateral columns of the cord. The anterior horn is more vulnerable to ischaemia, as infarction from occlusion of a single posterior spinal artery is unlikely and uncommon(10). The posterior spinal arteries are smaller paired arteries lying just medial to the dorsal roots. The basic mechanism in anterior spinal artery syndrome complicating aortic aneurysm is the occlusion of afferent arterial branches by the false lumen created as a result of dissection of the aortic wall(11).

The thoraco-lumbar segment is commonly involved and this is due to occlusion of the largest anterior medullary artery - (The great medullary artery of Adamkiewicz *arteria magna radicularis*) located between T8 and L2. Involvement of the cervical segment is occasional and with certain aetiologic features that differentiates it from involvement of other spinal segments(12). Compression of anterior radicular-medullary arteries or the cervical anterior spinal artery against the anterior bony wall of the spinal canal is a possible pathogenic mechanism(13).

Clinical features of anterior spinal artery syndrome:

The typical clinical presentation has an apopleptic onset evolving over minutes. Neurologic dysfunction stems from a non-traumatic lesion located in the anterior 2/3rd of the spinal cord, and sparing the posterior column. The most common thoracic syndrome varies from mild to moderate, sometimes reversible and sometimes waxes and wanes(4,9). In the acute stage and lasting up to three weeks, spinal shock with flaccid paresis, areflexia, and absent planter reflex is commonly observed. Distal to the lesion, superficial pain and temperature discrimination are lost bilaterally with preservation of light touch, vibration and joint position sense. Sometimes, painful burning dysaesthesia may develop below the lesion(14). Loss of sphincteric control with hesitancy and inability to void or defecate becomes evident with time, usually without the loss of sensation of bladder fullness(15). This function is preserved because this sensory reflex of bladder distention passes via the posterior column of the spinal cord. The descending pathways subserving detrusor function, coordination of bladder and urethral sphincter are located mainly in the lateral column while its ascending pathway is in the dorsal column (15). Thus, the vesicomotor dysfunction in this syndrome is similar to that of traumatic spinal cord injury except that bladder sensation is preserved in ASAS.

Acute weakness of either arms or legs without further neurologic deficits, should suggest incomplete ASAS with predominant involvement of the anterior horn cells(16). Although, ischaemic brochial/lumbosacral plexopathy has a similar clinical picture.

Diagnostic work-up: The diagnosis of ASAS, can be made with fair certainty by thorough clinical examination and knowledge of the underlying neuroanatomy. However, spinal MRI is the neuro-imaging modality of choice(17). In a T2-weighted imaging, a high signal intensity in the anterior region of the spinal cord is visualized, due to oedema of the grey and white matter subsequent to ischaemia(17,18). In the chronic stage, the lesions appear hypointense, with local atrophy of the cord. Cranial MRI may exclude the possibility of 'cerebral' paraparesis following ischaemia of bilateral anterior cerebral artery, an anomalous common stem or unpaired anterior cerebral artery or other para-sagittal lesions(19). CT myelography with metrizamide can be used if MRI is unavailable or unsatisfactory as in very obese patients(19). EMG and F-wave studies are useful in anterior horn cell dysfunction. Normal or large somato-sensory evoked potential (SSEP) amplitudes resulting from loss of anterolateral inhibitory influences on the dorsal column-medial lemniscal system confirms sparing of the dorsal sensory tracts in ASAS(20,21).

In the diagnosis of aortic dissection, four imaging techniques are useful. These include, angiography, transoesophageal echocardiography, chest CT scan and MRI. MR angiography is an excellent investigative tool with 100% sensitivity and specificity. It delineates the desired anatomy as well as a thrombosed false lumen. Transthoracic echocardiography as well as digital subtraction angiography has sensitivity of 100% for detecting both proximal and distal aneurysms(22-24). Other relevant investigations would include a chest-X-ray for any evidence of dissecting aneurysms in which abnormalities could be detected in 80 - 90% of cases, as well as radiological evidence of aortic calcification(25). Plain X-ray of the spine is unrewarding, but could give suggestive features of paraspinal soft tissue shadow in aortic dissection. ECG is rarely helpful in diagnosing aortic dissection. The most common finding is left ventricular hypertrophy and sometimes heart block(26). Absence of ECG evidence of acute myocardial infarction in the face of typical chest pain(19,26) suggests aortic dissection as in this patient. The haemogram, blood sugar, lipid profile, electrolytes, creatinine, urea, serological test for syphilis and HIV, creatinine kinase (CKMB), and lactate dehydrogenase (LDH) are additional relevant investigations.

Treatment: General risk factors such as diabetes mellitus, hypertension and hyperlipidaemia should be treated. Neuroprotective agents including anti-oxidants,

antiglutamatergic and protease inhibitors have been tried with inconclusive results in humans(11). Direct perfusion of dexamethasone sodium phosphate into the artery of Adamkiewicz has been tried with promising outcome(27). Anti-platelets are of questionable efficacy. The general management of paraplegics including physiotherapy is also indicated. The guiding principle in the management of dissecting aorta could be medical or surgical and this includes a) alleviation of pain, using narcotic analgesics. b) Reduction of pulse rate using beta blockers such as propranolol or esmolol with a target pulse rate of 60 - 80/ min. c) Reduction of a systolic blood pressure to a target of 90-110 mmHg, using Nitroprusside and beta-blocker(7,8). Surgery is indicated in patients with ischaemic compromise of vital organs, in aortic rupture, persistent pain, Marfans syndrome and extension of dissection despite adequate medical therapy. However, high intra- and post-operative morbidity and mortality rate have been reported in surgically treated patients(6).

In conclusion this study highlights a "not uncommon" complication of aortic dissecting aneurysm with a fatal outcome. This patient was managed conservatively because of the high surgical morbidity and mortality. The diagnosis of ASAS can be made with fair certainty by thorough clinical examination, knowledge of neuroanatomy as well as a high index of suspicion.

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