Spinal cord infarction

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ABSTRACT

Spinal cord infarction is a relatively rare but potentially devastating condition, where part of the spinal cord gets infarcted secondary to an interruption to its blood supply. The literature on this topic does not correlate with the increasing prevalence and importance of this condition. Using a case example to illustrate the clinical problem, this article aims to provide an overview of the anatomic basis, clinical presentation, pathophysiology and diagnostic approach to this condition. A brief discussion of the management principles is also included.

CASE VIGNETTE

A 74-year-old lady, with no previous medical history, was admitted to the hospital following a sudden onset of central chest pain radiating posteriorly, associated with nausea. Her systolic blood pressure (BP) was noted to be persistently >250mmHg. A serial troponin level was stable. An urgent CT angiography showed an extensive aortic dissection beginning just distal to the left subclavian artery and extending into the right iliac artery.

She was admitted to the Intensive care unit for intravenous control of BP. Progressively over days, she started to report severe pain and feeling of 'pins and needles' in her back, legs and buttocks, which then developed into almost a complete paralysis of her lower limbs. A repeat CT angiogram showed no extension of her type B dissection. On examination by a neurologist, she had bilateral profound flaccid leg weakness from hip flexion (i.e. L2-3) down, with sensory loss on levels below L3 on the right and L2 on the left. She also lost control of bowel and bladder function.

The clinical diagnosis was an anterior spinal cord infarction. An imaging of the spine was not done because the diagnosis was clear from the context and clinical findings. She spent over a month and a half in the acute hospital and then in the rehabilitation ward. Unfortunately, not much recovery was gained. Upon discharge, she was immobile, wheelchair bound and was subsequently discharged to a nursing unit.

INTRODUCTION

Spinal cord infarction (SCI) is a relatively rare but devastating condition, where part of the spinal cord gets infarcted secondary to an interruption to its blood supply. SCI or spinal stroke comprises only around 1% of all strokes.[1] Therefore, the literature written about SCI is much less than that about strokes, and hence the true prevalence in the general population is not known. However, just like strokes affecting the brain, SCI can leave the patient with variable neurological deficit, from minor weakness to paraplegia or quadriplegia, depending on the level that is involved and the severity of the infarction. Other deficits include bladder and/or bowel dysfunction and chronic back pain at the level of the infarction.[2] The recovery of function is variable, depending on the duration, severity and location of the infarction, as well as the presence of collateral blood supply. Furthermore, there is an increasing awareness about the condition partly due to the increasing prevalence of vascular disease and its operations as well as the increasing availability of advanced diagnostic imaging modalities. This review will briefly outline the anatomic basis for SCI, with a correlation to its clinical presentations, aetiologies, diagnostic approach and management principles.

SPINAL CORD FUNCTIONAL AND VASCULAR ANATOMY

In order to understand the causes of SCI, a good understanding of the vascular anatomy of the spinal cord is necessary because the resultant deficit in SCI usually matches a vascular territory in the cord. However, it is also possible that a diffuse hypoperfusion affects multiple watershed territories. The cord has highly complex and variable vascular anatomy among different individuals, however general principles still apply to the majority of patients.[3]

The spinal cord has segmental ventral (motor) and dorsal (sensory) roots that unite close to the cord to form 31 paired spinal nerves carrying afferent and efferent nerve fibres. Motor axons have their cell bodies in ventral horns of the central grey matter. Central processes of sensory neurons enter the dorsal cord. Light touch, vibration and proprioception afferents ascend rostrally in the dorsal column on the same side. Fibres carrying coarse touch, temperature and pressure sensations cross the central grey matter to the anterolateral spinothalamic tract, on the opposite side of the cord. The descending tracts that are most relevant to symptoms and clinical diagnosis of spinal cord lesions are the lateral corticospinal tract, which carries motor fibres to the ventral horns for voluntary movement, and the subjacent autonomic pathway for bladder control[4] (figure 1).

The spinal cord is supplied mainly by three arteries; one anterior spinal artery (ASA) and two (less well-defined) posterior spinal arteries (PSA). All three arteries branch off from the vertebral arteries, just before they merge to form the basilar artery, at the level of brainstem, right where the spinal cord starts. The 3 arteries run alongside the spinal cord: ASA anteriorly and the 2 PSAs posteriorly. At each level, the ASA supplies the anterior two thirds of cord, and the PSA pairs supply the rest. The three arteries finally anastomose at the conus medullaris, where the spinal cord

finishes, usually at the level of L1 or L2 vertebra.[5] Importantly, the ASA supplies the spinothalamic and lateral corticospinal tracts, whereas the PSAs supply the dorsal horn and column.

Along its course, the spinal cord also receives radicular arteries. These arteries supply the nerve roots exiting the spinal cord. These arteries also feed the ASA and hence the spinal cord. At the cervical level, the radiculomedullary arteries originate from the vertebral arteries to supply the top part of the spinal cord. From the level of aorta down, they originate from the intercostal arteries or the aorta itself.[6] These arteries enter the spinal canal through the neuroforamina (figure 2).

Since the intercostal arteries are mainly in the thoracic region, the thoracic spinal cord is particularly dependent on these arteries, making it the most vulnerable in acute aortic pathologies.[7] The largest and most clinically significant radiculomedullary artery is the artery of Adamkiewicz, which is the main supplier of the lumbo-sacral spinal cord. The level of this artery is variable, but it most commonly arises at the level of 8th to 12th intercostal arteries in 62-75% of patients.[8]

CLINICAL FEATURES

There are two SCI vascular syndromes described based on which spinal artery is affected; anterior and posterior spinal artery syndromes. Anterior spinal artery syndrome (ASA) is by far the most common syndrome, partly because the ASA gets direct feeding from the aorta, whose pathologies are the most common cause of SCI. It tends to be severe because it supplies the anterior two thirds of the cord and is a single artery, which explains why the symptoms of the ASA syndromes are bilateral. Based on the neuroanatomy described above, the ASA syndrome would cause an abrupt onset of bilateral weakness, areflexia for reflexes whose connections pass through the infarcted region, and pain and temperature sensory deficits while sparing proprioception and vibration.[9, 10] Other symptoms include sexual dysfunction, bladder and/or bowel dysfunction and autonomic dysfunction, manifesting as retention of urine followed by overflow incontinence, unstable blood pressure, and paralytic ileus.[9, 11]

Because of the nature of the usual triggers that interrupt the blood supply, SCI onset is usually abrupt, but can be subacute, evolving over hours to a few days, as in the presented case. Just like traumatic spinal cord injuries, it initially manifests as a flaccid weakness, usually with extensor plantars that evolves over days to weeks to more chronic upper motor neuron signs: spasticity, exaggerated reflexes and clonus.

The deficits depend on the level that is affected. It can involve only lower limbs if the thoraco-lumar region is affected, or all limbs if cervical region is affected. There is also typically a sensory level, which is best demonstrated by a pin prick test. However, it usually occurs a few segments below the actual spinal level, because of decussation of the spinothalamic tract over several segments. Table (1).

Lesions above C3 segment can compromise spontaneous respiration due to a loss of diaphragmatic innervation. Orthostatic control of blood pressure can be lost if the lesion is above T5, the origin of the greater splanchnic nerve,[12] which supplies the sympathetic innervation to the adrenal medulla to stimulate catecholamine release. This sympathetic neurohumoral diminution makes patients prone to postural hypotension in the acute phase, accompanied by symptoms related to diminished cerebral perfusion, including lightheadedness, syncope, tinnitus, facial pallor and blurred vision.[13] Over time, the spinal cord tissue undergoes neuroplastic changes, which result from interruption in the autonomic relaying of the afferent signals to the brain and loss of cortical inhibitory signals to the spinal reflex. This results in an exaggerated sympathetic spinal reflex to the afferent signals, called autonomic dysreflexia. This is particularly relevant in cervical and high thoracic lesions. It is characterised by very severe hypertension, with systolic BP reaching up to 300mmHg, and can be accompanied by headache, sweating, chest tightness, bradycardia and blurred vision.[14, 15] Dysreflexia symptoms can be triggered by a range of stimuli, particularly from visceral distention (usually from a full bladder or bowel), pressure sores, urinary catheterisation, urinary tract infection, cytoscopy, ejaculation and surgical procedures.[15]

This usually causes a great deal of discomfort and compromise to the patient's quality of life. if untreated, autonomic dysreflexia can lead to serious consequences, including intracranial haemorrhage, retinal detachment, seizures, cardiac arrhythmias, and death. Having said that, some paralympic athletes with spinal cord lesions intentionally induce it before their competitions to boost their performance, necessitating medical examinations before their competitions.[15]

Other less common syndromes include posterior spinal artery syndrome and Brown-Séquard syndrome. Posterior spinal artery syndrome is rarely encountered in clinical practice. Because there are two PSAs, this syndrome tends to be unilateral and less severe. Based on the functional neuroanatomy, it makes sense that this syndrome leads to ipsilateral loss of light touch, vibration and proprioception, while mostly sparing the motor function.[10] Brown-Séquard syndrome is caused by the lateral hemisection of spinal cord. It causes ipsilateral loss of motor function and tactile discrimination, vibratory, and position sensation, and contralateral loss of pain and temperature sensation. It is usually caused by a trauma, such as a gunshot or puncture wound, but it has been reported to occur following aortic surgeries.[16]

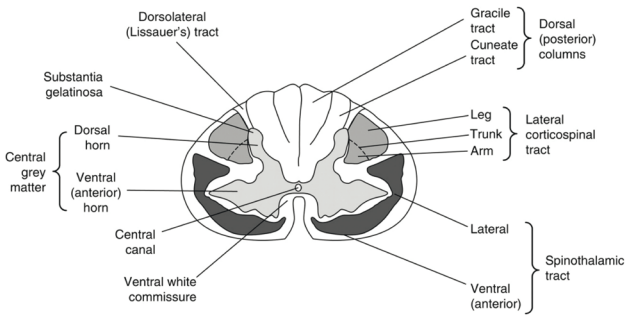

Figure 1: Mid-cervical spinal cord sectional anatomy. [25]

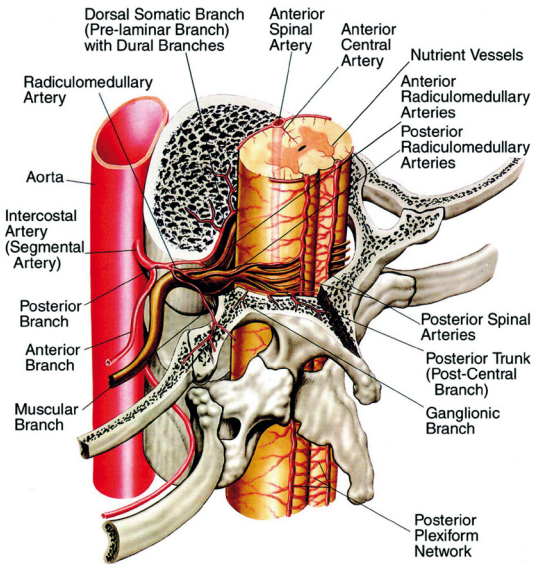

Figure 2: Spinal anatomy and associated structures. [27]

Level of Infarction	Clinical Features
Cervical	Bilateral lower and possibly some upper (depending on level) limb weakness Bilateral loss of pain and temperature sensation in upper and lower limbs If lesion above C3, compromise of spontaneous respiration Autonomic dysreflexia, autonomic dysregulation and sphincter dysfunction
Thoracic	Bilateral lower limb weakness Bilateral loss of pain and temperature sensation in lower limbs If lesion above T5, autonomic Dysreflexia with autonomic dysfunction Sphincter dysfunction
Lumbar	Bilateral lower limb weakness Bilateral loss of pain and temperature sensation in lower limbs Sphincter dysfunction

Table 1: Summary of Clinical Features Associated with Levels of Anterior Spinal Cord Infarction.

CAUSES OF SCI

There are so many reported causes for SCI, and so it is easier to think about most of them according to the pathophysiology of the infarction. When it comes to causes of SCI, it is not much different from stroke in the cardiovascular differential diagnosis: cardiogenic thromboembolism (e.g. atrial fibrillation, infective endocarditis), spinal artery atherosclerosis, global (e.g. circulatory failure) or regional ischaemia (e.g. aortic injury, arteriovenous malformation, epidural haematoma), vasculitis (e.g. polyarteritis nodosa), compressive lesions (e.g. disc prolapse) and hypercoagulability.

Aortic pathology is the most common cause of SCI. Acute aortic events, such as aortic dissection, trauma, thrombosis and aneurysm rupture can compromise the blood flow to the spinal arteries, by either reduced circulating blood volume or direct event extension involving the spinal arteries. There have been cases where asymptomatic abdominal aortic aneurysm presents as a SCI by an embolic mechanism.[17] Aortic surgery and repair, whether open or endovascular, are known to be highly associated with SCI (11% of aortic surgeries, according to some estimates 18). This is partly explained by the presence of a period of generalised diminished perfusion during aortic cross-clamping and/or aortic surgical instrumentation. The aortic cross-clamping causes hypertension proximal to the clamping, but this is followed by sudden hypotension upon releasing the clamp, which leads to low spinal perfusion.[19] Emboli dislodged by the procedure are also a likely cause. Non-aortic surgery, especially spine surgery,[11,20] is also reported to cause SCI.

Direct occlusion of the spinal arteries is a less common aetiology for SCI. This is usually due to atherosclerotic lesions, embolic events, vasculitis or thrombophilia. Systemic hypoperfusion has been reported to cause SCI, however SCI can be less concerning compared to their underlying illness. This association has been illustrated in a post-mortem retrospective case series, where 46% of patients who died after a global ischaemic event (i.e. cardiac arrest or severe hypotension) were found to have ischaemic myelopathy.[21]

DIAGNOSIS AND IMAGING

Diagnosis is usually suspected based on the abrupt onset and pattern of symptoms and findings. The aetiology is usually inferred from the clinical context. However, other similar conditions, such as cord compression, multiple sclerosis, and myelitis have to be excluded. History of malignancy and back pain should raise suspicion of compressive myelopathy from spinal metastases, while additional neurological deficits not related to spinal pathology or inflammatory cerebrospinal fluid analysis raise suspicion of multi-focal disease (i.e. multiple sclerosis) and myelitis, respectively. Other conditions that can mimic anterior cord syndrome are anterior radiation myelitis and anterior intervertebral disc herniation, both of which can be suspected from the history and the imaging.

Imaging is used to confirm diagnosis and infer the aetiology if not clear. The imaging modality of choice for spinal cord ischaemia is MRI. However, as for brain imaging, MRI can be normal in the acute phase and usually evolves to show spinal cord swelling and high T2 signal after several days.[22] When interpreting spinal MRI and relating the lesion to its level, it is important to remember that the spinal cord is shorter than the vertebral column, as it finishes at L1 – L2. Below this level, the vertebral column contains only lumbosacral spinal nerves contained in a thick fibrous strand (the cauda equine), which runs down to the coccyx. In general, there is little difference between the spinal and vertebral levels in the upper levels, whereas in the lower thoracic and lumbar region, the spinal nerves have to travel down multiple levels to reach their corresponding neuroforaminae.

CT or MR angiography can be helpful in identifying the underlying vascular problem (e.g. atherosclerosis, arterio-venous malformation, aneurysms, occlusions). This can be particularly useful if a surgical repair can be offered. It also aids identification of artery of Adamkiewicz prior to aortic surgery to avoid intraoperative spinal ischaemia.[23]

MANAGEMENT

Management largely depends on the underlying cause for the infarction. Quick corrective surgery is needed if acute aortic event, vascular compression, malformation or bleed (haematomyelia) start to cause spinal ischaemia. In the event of global hypoperfusion, maintaining adequate blood pressure to maintain spinal perfusion is crucial, especially if there are vascular atherosclerotic lesions. Antiplatelet therapy may be needed to prevent vascular occlusions or embolism. Corticosteroids may be used in cases of vasculitis or aortitis.

The management strategy should be tailored to the patient's specific neurologic deficits and should begin right after the infarction. After controlling the underlying cause (e.g. aortic dissection, hypoperfusion), measures are targeted at alleviating the symptoms and preventing further complications while rehabilitating the patient to regain as much function as possible. In the acute phase, patients are prone to complications including pneumonia, urinary tract infections, deep vein thrombosis, pulmonary embolism, pressure sores and neurogenic shock. Effective clinical and nursing cares should focus on monitoring for and treating these complications.

These patients should be placed in specialised spinal rehabilitation units equipped with resources to deal with common issues such as bladder and bowel dysfunction, limb weakness or paralysis, limb spasticity and contractures, infections, skin care and psychological sequelae.[24] Autonomic dysreflexia can be prevented by avoidance of the triggers and using α-adrenergic blocking antihypertensive agents. Acute treatment of dysreflexia is by sitting the patient upright, removing the trigger and controlling the hypertension using fast-acting antihypertensive agent.[25] Spasticity is a big concern in these patients because it leads to contracture and severe complications, such as permanent loss of limb function and joint mobility, skin pressure areas, pain and uncomfortable postures. It can be minimized by passive stretch exercises and oral medications, such as GABA agonists (e.g. Baclofen, Benzodiazepines) and calcium release blockers (e.g. Dantrolene).[26]

Multi-disciplinary involvement with these patients is crucial to their recovery and to achieve the rehabilitation goals. These include physical and occupational therapists to improve their power, balance, function and adaptive skills. The psychological decline leads to demotivation and further loss of function. Hence, clinical psychologists are essential part of the multi-disciplinary team. The social worker's role is vital in facilitating social support, housing alterations, and finding suitable nursing homes for these patients in the community when needed. The long-term goal should be toward increasing the potential of the patient's residual function as well as facilitating a supportive environment. This might mean, in some cases, a residential level of care, although some patients regain some degree of independence and function after an intensive rehabilitation programme. The patient described

in the case did not experience significant improvement in mobility and remained wheelchair bound. Most of the recovery occurs in the first days to weeks following the event, and hence her prospects of further long-term recovery are limited. This highlights the variability of severity of deficits, patient's progression and prognosis after SCI.

CONCLUSION

Spinal cord infarction is a potentially devastating condition caused by an interruption to the spinal blood supply, usually through aortic events or surgeries. The usual presentation is of an anterior spinal artery syndrome, where there is bilateral loss of motor function and pain and temperature sensation below the affected spinal level. If the diagnosis is not evident clinically, spinal MRI is the diagnostic modality of choice to confirm diagnosis. Management involves preventive and acutely controlling the underlying vascular event. Multi-disciplinary team in a rehabilitation ward are sometimes to maximize the recovery of the patient.

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