Spontaneous conus medullaris infarction in a 79 year-old female with cardiovascular risk factors: a case report

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Objective: To detail the case of a 79 year-old female who presented with sudden bilateral neurological deficits of the lower extremities and was later diagnosed with non-traumatic conus medullaris infarction. The purpose of this case report is to inform primary contact practitioners of the presentation, diagnosis and the associated risk factors of this condition in order to facilitate prompt management.

Clinical Features: Spinal cord infarction presenting as low back pain with a high degree of bilateral loss of motor strength, sensation and reflexes in the lower extremities and bowel/bladder dysfunction, in a patient with previous coronary artery bypass graft surgery and renal insufficiency.

Intervention and Outcome: Referral to emergency within hours of symptom onset allowed for immediate assessment, management and relatively favourable partial recovery.

Summary: Although rare, conus medullaris infarction is potentially devastating and requires an appropriate clinical index of suspicion for timely diagnosis, treatment and optimal neurological recovery. (JCCA 2012; 56(1):58–65)

Key words: spinal cord diseases, spinal cord ischemia, spinal cord infarction, conus medullaris

But : Il s’agit d’un rapport détaillé du cas d’une femme de 79 ans souffrant de déficits neurologiques bilatéraux d’apparition soudaine aux membres inférieurs, qui a ultérieurement reçu le diagnostic d’infarcissement non traumatique du cône médullaire. Ce rapport de cas a pour objectif d’informer les praticiens de premier contact du tableau clinique, du diagnostic et des facteurs de risques associés à ce trouble afin de faciliter une prise en charge rapide.

Caractéristiques cliniques : Infarctissement de la moelle épinière se manifestant sous forme de douleur lombaire accompagnée d’un degré élevé de perte de motricité bilatérale, de sensation et de réflexes dans les membres inférieurs ainsi que d’une dysfonction intestinale/vésicale chez une patiente ayant déjà subi un pontage coronarien et une insuffisance rénale.

Intervention et résultat : La patiente a été transférée au service d’urgence dans les quelques heures suivant l’apparition des symptômes où elle a pu être immédiatement évaluée et prise en charge, et bénéficier d’un rétablissement partiel relativement favorable.

Résumé : L’infarctissement du cône médullaire, bien que rare, a le potentiel d’être dévastateur et nécessite un indice de suspicion clinique approprié pour que le diagnostic, le traitement et le rétablissement neurologique optimal puissent se faire en temps opportun. (JCCA 2012; 56(1):58–65)

Mots clés : Maladies de la moelle épinière, ischémie médullaire, infarctissement médullaire, cône médullaire

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Consent: Patient gave consent verbally to use her file and images for the purpose of this case report.

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Introduction

Spinal cord infarction occurs much less frequently than cerebral infarction, accounting for 1% of all strokes. This can occur anywhere along the length of the spinal cord, with infarctions at the conus medullaris thought to be particularly rare. Although the condition is rare, it is potentially devastating and may result in significant and residual impairments in sphincter control and ambulation. Thus, prompt assessment and diagnosis is necessary to ensure appropriate management in order to preserve neurological function.

The purpose of this case report is to heighten awareness of spinal cord infarction since the main predictors of long term outcome are the neurological state and duration of symptoms at time of diagnosis. The report details the clinical presentation of a non-traumatic conus medullaris infarction in a 79 year-old female with associated cardiovascular risk factors of previous coronary artery bypass graft surgery (CABG) and renal insufficiency. The prompt assessment and management resulted in relatively favourable partial recovery of neurological function.

The report underscores the varied presentation of conus medullaris infarctions and emphasizes its consideration to facilitate immediate referral to the emergency department for surgical consultation and appropriate management. Further, the demographics and outcomes of conus medullaris infarction are not well studied. This case report sheds some light on the potential risk factors and treatment outcomes of spontaneous conus medullaris infarctions.

Case Report

A 79 year-old female awoke in the morning with spontaneous low back and right leg pain that developed into bilateral total lower extremity paresthesias and progressive bilateral motor weakness. She was transferred to the emergency department by ambulance shortly after the onset of symptoms. On initial examination, hip flexion, knee extension, and ankle dorsi- and plantar flexion strength were graded as 0/5 bilaterally. Lower limb deep tendon reflexes were absent, while upper limb reflexes were normal. Sensation to pin prick was decreased below the T12 level. The plantar reflex was flexor bilaterally (i.e. negative Babinski sign). There was a decrease in anal tone and moderate urinary retention. Vital signs were within normal limits. Cardiovascular, respiratory, and abdominal examination were unremarkable. Peripheral pulses within the lower limbs were also normal. Past medical history was remarkable for coronary artery disease with previous coronary artery bypass graft (CABG) surgery, right mastectomy with lymph node dissection, bipolar disorder, and renal insufficiency.

An MRI of the thoracic and lumbar spine and CT angiogram were conducted. MRI revealed diffuse abnormal high T2 signal centrally within the expanded conus medullaris with a peripheral rim of sparing; this extended over approximately the distal 9 cm of the spinal cord (Figures 1 and 2). Associated restricted diffusion was additionally noted in this region (Figure 3). CT angiography images demonstrated marked diffuse atheromatous disease of the aorta and its major branches including a posterior penetrating ulcer at approximately the T12 to L1 vertebral levels with evidence of plaque rupture, as
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demonstrated by an adherent intraluminal filling defect / thrombus within the aorta (Figures 4A and 4B).

Following the diagnosis of conus medullaris infarction, the patient was admitted under the neurology service for management. A vascular surgeon was consulted but medical management was favoured. The patient was also seen by psychiatry for her history of bipolar disorder and a labile mood during her stay in the hospital. No psychiatric treatment was deemed necessary as mood changes (depression) were thought to be related to her current physical condition. She experienced minor complications in renal function, as well as occasional low blood pressure that required a modification to her medications.

In-hospital treatment included anti-platelet (clopidogrel) and anti-coagulation medications (initially with intravenous and subsequently subcutaneous heparin), as well as physical therapy and occupational therapy directed at her activities of daily living, in preparation for transfer to a rehabilitation hospital for more extensive care. The symptoms of bilateral total lower limb paresthesias and motor weakness gradually improved through the length of her hospital stay. The patient regained partial strength in her lower extremities, with greater improvement on the right. At the time of transfer to another hospital for rehabilitation, her strength on the right was 3/5 for hip flexors, 2/5 for knee extensors, and 1/5 for right ankle dorsi- and plantar flexors. On her left extremity, the hip flexors were rated 2/5, while knee and ankle strength was 0/5. The neurological symptoms were stable at the time of transfer, indicating partial but relatively favourable recovery of the conus medullaris infarction.

Discussion

The incidence of conus medullaris infarction is not well understood. Some of the first reports on this condition were published in the 1980’s and studies to date have largely explored this entity retrospectively and in concert with other spinal cord infarctions. In a 2007 retrospective review of 175 patients diagnosed with a spinal cord injury clinical syndrome in an acute rehabilitation centre, 14 patients were classified as having conus medullaris syndrome. Only three of these patients were associated with non-traumatic etiologies, namely hypoperfusion, infection or tumour.

The most recognized etiologies of ischemia of the spinal cord are atherosclerosis and cardio-embolic infarc-
tion, leading to obstruction of the tissue's blood supply. Rare causes of conus medullaris injury include traumatic fractures, acute disc herniations, intradural tumours and aortic surgery. To appreciate how such causes may result in the subsequent neurological deficits of conus medullaris infarctions, the arterial and neurological components of the spinal cord are briefly reviewed.

Anatomy of the cord
It has been suggested that spinal cord infarctions may result from 1) interruption of the blood supply anywhere between the aorta and intramedullary vasculature; 2) deficient systemic perfusion pressure; 3) hypercoagulation; or 4) a combination of these. Since the anterior and posterior spinal arteries closely approximate the ver-

Figures 4A and 4B  Sagittal and axial CT images, acquired with intravenous contrast in the arterial phase, reveal diffuse calcified and noncalcified atheromatous disease of the aorta and its major branches. At approximately the L1 level, there is a posterior penetrating atheromatous ulcer of the descending aorta (arrowhead) with evidence of plaque rupture (intraluminal thrombus/plaque material) (arrow).
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Tebral bodies, discs and ligamentum flavum, prolonged compression of these arteries from pathologies in these structures may also result in arterial compromise and formation of an infarct. In consideration of the spontaneous nature of this patient’s injury and in the absence of any direct trauma or compressive lesions at the spinal cord, the presumed cause of the infarction was vascular disease.

In general, the major extrinsic arteries of the spinal cord are the anterior and posterior spinal arteries, anterior and posterior radiculomedullary arteries and pial arteries. Originating from the vertebral arteries, the single anterior spinal artery supplies the anterior third of the spinal cord, including the central grey matter and anterolateral white matter, while the paired posterior spinal arteries supply the posterior third, including the posterior columns (Figure 5). The radiculomedullary arteries, originating from the segmental spinal arteries, meet the anterior and posterior spinal arteries at various levels, and pial arteries run circumferentially between the anterior and posterior spinal arteries.

However, the spinal cord vasculature at the level of the conus medullaris has some added characteristics. First, it has been reported that anastomosis invariably occurs between the anterior and two posterior spinal arteries, known as the anastomotic ansa of the conus. The most caudal region of the spinal cord, extending from T8 to the conus, is additionally supplied by the artery of Adamkiewicz, branching from the intercostal artery, and occasionally the Desproges-Gotteron artery, branching from the internal iliac artery. This regional vasculature supports the notion that infarctions at the conus medullaris are more infrequent than those at other parts of the spinal cord. Further, Monteiro et al. proposed that the anastomotic ansa of the conus allows eventual reperfusion of the necrotic tissue after obstructed blood flow from the artery of Adamkiewicz, thereby providing insight into its potential natural history.

Clinical presentations

Neural ischemia at the conus medullaris can result in variable clinical presentations, depending upon the degree of compromise along the length of the spinal cord. Injury to the conus medullaris commonly results in sudden non-specific low back pain that progresses to bilateral leg pain with bladder and bowel dysfunction and saddle anesthesia. Specifically, conus medullaris injuries lead to a lower motor neuron (LMN) syndrome, where sacral and limb reflexes are diminished and muscles become flaccid and atrophic. There are also varying degrees of LMN bladder-bowel dysfunction (atonic bladder and flaccid anal sphincter) and sexual dysfunction (loss of reflexogenic but preserved psychogenic erection in men). Although the LMN syndrome is similar to cauda equina lesions, injuries to the cauda equina are typically characterized by asymmetrical lower extremity weakness and variable sensory and reflex deficits. However, due to its close proximity to the epiconus, conus medullaris lesions can present with mixed neurological deficits of combined upper and lower motor neuron syndromes. For instance, upper motor neuron syndrome is often reported with conus medullaris lesions, likely due to ischemia that overlaps the epiconus region, which characteristically presents as a pure upper motor neuron syndrome.
To further add to this diagnostic challenge, the conus medullaris has a variable anatomical location. Although the conus medullaris represents a transition from the central to the peripheral nervous system, there is no fixed anatomical landmark. A 2007 review by Kesler et al. found the most common location for termination of the conus medullaris was the L1-2 disc space, but had a large anatomical range extending from the T11-T12 disc space to L4 vertebra. Others have suggested that distinguishing between the conus and the cauda equina is of little value and difficult to conduct. Thus, these findings suggest that both the conus medullaris and cauda equina regions in these patients. For example, the infarction in the case presented herein was found to be located outside of the commonly cited region and situated more superiorly at the T12-L1 level. This emphasizes the importance of evaluating the epiconus, conus medullaris and cauda equina regions in these patients.

In light of this varied clinical appearance, clinicians should also evaluate and be aware of potential risk factors. Cheng et al. (2008) found that in a small sample of 22 subjects with spinal cord infarctions, hypertension was the leading risk factor, followed by diabetes mellitus and heart disease. These vascular diseases are in line with the biological plausibility of arterial occlusion that leads to eventual tissue necrosis. However, other less obvious risk factors have been suggested. A recent case report described the onset of bladder and bowel incontinence from a partial conus medullaris lesion immediately following a CABG. In our case report, the infarction did not occur immediately after the patient had undergone a CABG procedure, suggesting that intraoperative micro-injury may have led to an occult decrease in vascular supply that, over time, resulted in conus medullaris ischemia, in the setting of this patient’s aortic atherosclerotic disease. Previous reported cases of conus medullaris ischemia following aortic surgery support such a mechanism of injury.

Clinical management and prognosis

There appears to be no clear guidelines for the treatment of spinal strokes. Initially, magnetic resonance imaging and surgical consultation are used to evaluate the need for immediate surgical decompression of the spinal cord, particularly in patients with underlying tumours. For spontaneous infarctions, the most appropriate pharmacological interventions have not been confirmed. In animal studies, some benefit has been found with certain agents, such as prostaglandins or corticosteroids, but these have not been studied prospectively in humans. In a retrospective study by De Seze et al., no differences in treatment outcomes were found in subjects who were treated with corticosteroids and anti-platelet therapy compared to those who received anticoagulation with anti-platelet therapy. Following this, similar to cerebral strokes, spinal cord infarctions require close monitoring, controlling for hypotension and managing co-morbidities that may impair patient recovery.

Patient recovery from a conus medullaris infarction is dependent on a number of prognostic factors. Severe neurological impairment at the time of assessment has been associated with poor recovery of motor functions. In addition, older age has generally been associated with poor outcomes, though not in all studies. As in our case report, the patient’s older age did not appear to be a negative factor for functional recovery, as motor strength stabilized prior to rehabilitation. In extreme cases, a mortality rate of approximately 20% has been reported in retrospective analyses of spinal cord infarctions, though the causes of death were not conclusive.

The degree of morbidity and mortality emphasizes the potential burden of spinal cord infarctions and importance of prompt diagnosis. Yet promptness in diagnosis depends upon the clinician’s awareness and maintenance of high index of suspicion, particularly in conditions that do not present or respond as expected. Caputo et al. (1997) suggested that a tendency for delayed diagnosis was more frequently found in rarely encountered conditions with central neurological presentations, which increased the risk for ineffective, harmful or lack of treatment. For example, Morandi et al. published a case where caudal spinal cord ischemia was diagnosed in a subject 36 hours after receiving lumbar vertebral manipulation for an acute exacerbation of chronic low back pain, with no evidence of a proper physical or neurological examination prior to treatment. Given its likelihood for sudden and severe onset, it is possible that such spinal cord ischemia was in evolution prior to the manipulative treatment and perhaps a thorough neurological examination would have identified the incriminating red flags. While the authors did discuss potential mechanisms for this occurrence, including arterial occlusion from fibrin-platelet or disk material...
embolism, it is important to note that case reports are unable to determine causal relationships. Thus, primary contact practitioners, including chiropractors, should conduct thorough examinations to facilitate prompt referral to the emergency department for suspected cases of spinal cord infarction.

Summary
This case report illustrates a relatively favourable partial recovery of a conus medullaris infarction in a 79 year-old female who received prompt assessment. The report underscores the importance of awareness of this condition among clinicians in order to facilitate immediate management.

While the incidence of conus medullaris infarction is rare, it has the potential of a variable clinical presentation, making it difficult to recognize. This may result in a delayed diagnosis, progression of neurological symptoms, and the increased likelihood of poor recovery. In extreme cases, spinal cord infarctions have led to permanent paraplegia and even mortality. This case report illustrates the importance of establishing an appropriate clinical index of suspicion that can lead to timely diagnosis and treatment for suspected cases of spinal cord infarction.

References
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