Acute Venous Congestive Myelopathy in a Patient with Neurosarcoidosis

Nicola Hazel Guy

1 Department of Neurosurgery, Capital and Coast District Health Board, Wellington Regional Hospital, Newtown, Wellington, New Zealand

Address for correspondence Nicola Hazel Guy, MMedSci, MBChB, Department of Neurosurgery, Capital and Coast District Health Board, Wellington Regional Hospital, Mein Street, Newtown, Wellington 6021, New Zealand (e-mail: nicola.guy@ccdhb.org.nz).

Introduction

The clinical red flags of cauda equina syndrome are bowel and bladder dysfunction, saddle anesthesia, and power or sensation deficits in the lower limbs. In clinical situations where there is suspicion of cauda equina, a magnetic resonance imaging (MRI) of the lumbar spine is generally expedited. Most commonly, a discogenic etiology is uncovered with investigative imaging; however, intramedullary lesions of the conus medullaris can also present with these symptoms if there is compression of the exiting nerve roots. Conus lesions often present with mixed upper and lower neuron compression signs as both the conus medullaris and the cauda equina can be affected.2

There is a wide range of differentials of intramedullary conus lesions, summarized in Table 1. These include neoplastic and non-neoplastic lesions. As not all of these conditions are best managed surgically, or even require an invasive biopsy for diagnosis, it is important that neurosurgeons maintain a high degree of suspicion for diagnoses further down the list of differentials.

We present a case that was referred to our unit as a conus tumor for an invasive biopsy; however, the multidisciplinary team of surgeons, neuroradiologists, and oncologists considered alternative diagnoses. The initial diagnosis of neoplasm gave way to a diagnosis of venous congestive myelopathy secondary to inferior vena cava agenesis and spinal neurosarcoidosis. This case demonstrates the importance of neurosurgeons retaining a high degree of suspicion for alternative diagnosis to avoid unnecessary surgical risk.

Case Report

A 45-year-old man presented to his local hospital after a week of lower back pain, 48 hours of ascending numbness in the lower limbs, difficulty mobilizing, and urinary retention. He was previously fit and well, had an active job, and was part of a social rugby team. An MRI of the lower spine revealed abnormal T2 hyperintensity and widening of the distal spinal

Abstract

There is a range of differential diagnoses for intramedullary lesions of the conus medullaris, both neoplastic and non-neoplastic. There is a limited role for surgery in a large proportion of these diagnoses, and operative risks can outweigh any benefits of surgery. Here a case is presented of a patient referred to a neurosurgical center for a biopsy of a presumed neoplastic conus tumor. However, through the collaboration of a multidisciplinary team, further diagnoses were considered. After thorough investigation, two conditions were diagnosed: venous congestive myelopathy secondary to inferior vena cava agenesis and spinal neurosarcoidosis. This case demonstrates the importance of neurosurgeons retaining a high degree of suspicion for alternative diagnosis to avoid unnecessary surgical risk.

Keywords

► neurosarcoidosis
► venous congestive myelopathy
► cauda equina
► intramedullary myelopathy

Case Report

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cord. The orthopaedic team in the local hospital made an acute referral to our neurosurgical service for the management of a spinal cord tumor, and on our advice the team administered dexamethasone prior to transfer.

On arrival to our hospital, a full spinal neurological exam was performed and noted normal power with decreased pain and light touch sensation below T12, with diminished but present reflexes. Proprioception was impaired distally below the ankle. On further interrogation of his symptoms, he reported years of exercise-induced transient numbness in the ankle. On further interrogation of his symptoms, he was discussed at a neurosurgical multidisciplinary meeting, inferior vena cava was noted. These images were then representative axial slice through L1 level showing enlarged cord and enhancing region located in dorsal part of cord. (B) T2 fat-saturated sagittal image demonstrating central cord myelopathy from T8 to L2. (C) Sagittal T2 image demonstrating flow voids of the dilated paraspinal varices (E) Axial through T11 demonstrating flow voids of the dilated paraspinal varices, and central cord myelopathy.

Table 1 Differential diagnosis for intramedullary lesions in the conus medullaris (adapted from Ebner et al19)

| Neoplastic          | Ependymoma | Astrocytoma | Glioblastoma | Ganglioma |
|---------------------|------------|-------------|--------------|-----------|
|                     | Hemangioblastoma | Lymphoma | Melanoma | Metastases |
| Non-neoplastic       | Granulomatous—Tuberculosis, sarcoidosis | Infection—Abscess, parasitic | Vascular—Cavernoma, malformations | Demyelinating—Multiple sclerosis | Dysembryogenic—Lipoma, epidermoid cyst |

where the focus shifted from a diagnosis of spinal cord tumor to venous congestive myelopathy.

Following this, a computed tomography (CT) venogram was performed to investigate possible thrombosis causing the acute presentation given the lifelong underlying vascular abnormality. No thrombosis was seen and thus we proceeded with a spinal angiogram that did not reveal a dural arteriovenous fistula or arteriovenous malformation. Vascular surgery and hematology were consulted, and there was some discussion about anticoagulation in this patient, who had venous congestion and varices without proven thrombosis. Ultimately, it was decided not to initiate anticoagulation.

During the admission to the neurosurgical service, the patient’s neurology improved rapidly after administration of steroids and he regained his mobility and bowel and bladder function. Given the significant positive response to steroids, a neurology opinion was sought and the workup started for an inflammatory myelopathy. The patient was transferred back to his local hospital for a lumbar puncture and further investigations. A CT of the chest demonstrated mediastinal lymphadenopathy and shortly following this the cerebrospinal fluid angiotensin-converting enzyme level came back significantly elevated. A diagnosis of neurosarcoidosis was made and the patient was started on immunosuppression therapy.

Two months after his initial presentation, he was fully mobile with a normal neurological exam and some mild urinary hesitancy.

Discussion

This case highlights the importance of the neurosurgeon maintaining a high index of suspicion for alternative diagnosis when referred patients with spinal pathology. Although common things happen commonly, what is first thought to be a spinal tumor is not always a spinal tumor. Non-neoplastic lesions, vascular malformations, and demyelinating disorders can also present as regions of spinal cord enlargement. In our case, there were some red flags in both the clinic history and the imaging that led us away from planning a spinal cord biopsy, toward a diagnosis of venous congestive myelopathy, and further to the discovery of sarcoidosis.

The discovery of inferior vena cava agenesis and significant retroperitoneal and paraspinal varices allowed us to consider vascular myelopathy as a potential diagnosis. The patient presented acutely with cord compression myelopathy symptoms on a background of years of exercise-induced transient lower limb numbness, which is a temporal pattern strongly associated with vascular myelopathy. There have been cases reported previously where patients have presented with cauda equina symptoms due to venous congestion from dilated varices. The underlying etiology of cases with venous congestion is more commonly lower limb deep vein thrombosis than from inferior vena cava agenesis as in this case. Although the history of exertion-induced lower limb sensory changes could be attributed
to the venous congestion, the acute presentation did not clinically behave like a vascular myelopathy case as there was clinical improvement after steroid administration. As 50% of patients with venous congestion have worsened neurology after steroid dosing, the team was suspicious of an alternative diagnosis and turned to our neuroradiologists and the MRI for diagnostic help.

On the first lumbar MRI scan, the region of T2 hyperintensity was in the shape of the conus, not displacing the cord that led us away from the diagnosis of spinal tumor. In our institution, we do not have the ability to perform spinal cord diffusion tensor imaging; however, tractography is used in other centers to aid in the differentiation between tumor (spinal cord tract displacing) and inflammatory, non-displacing, lesions.8

In this case, the features of cord myelopathy extended from T7 to L2 (Fig. 1B). However, the imaging findings were out of proportion to what was demonstrated on clinical examination, and given the extent of the myelopathy on imaging we would have expected a greater neurological deficit. This discrepancy between the imaging and the clinical presentation raised the suspicion of neurosarcoidosis with our radiologists. And this suspicion was heightened in the context of the postcontrast MR spine demonstrating a contrast-enhancing region within the area of myelopathy (Fig. 1A).9 The pattern of enhancement was not the typical patchy nodule pattern observed in most reported cases of neurosarcoidosis and there was no pachymeningeal enhancement.10,11 Although the radiological findings are not specific to sarcoidosis, there was enough suspicion to justify continued investigation into inflammatory etiologies by our neurology colleagues which eventually yielded the diagnosis of sarcoidosis.

This case was interesting to us both as a diagnostic challenge and as a novelty of seeing dual presentations of rare conditions. Spinal sarcoidosis only occurs in 0.43 to 1% of neurosarcoïdosis cases, in addition to congestive myelopathy from agenesis of the inferior vena cava, of which there are seven cases in the reported literature.6,13–18 However, the probability of having two uncommon diagnoses presenting simultaneously should not detract from the learning point that a surgical intervention is not always required in all referrals of spinal cord lesions. The neurosurgeon should be alert to mimics of spinal neoplasia such as neurosarcoidosis and venous congestive myelopathy.19 There is no role for surgery in the treatment of venous congestive myelopathy caused by inferior vena cava abnormalities and the majority of cases are medically or conservatively managed.13 Diagnosis of neurosarcoïdosis is often a diagnosis of exclusion and the role for surgery is limited to diagnostic tissue biopsy.20 In some clinical series, spinal cord biopsy carries a 21% complication rate with significant risk of neurological damage21 not to mention the bleeding risk would be significant in this patient whose venous drainage of his abdomen and lower limbs passes through the venous plexes surrounding his spine.22 The collaboration of a multidisciplinary team in critically reviewing the imaging and considering alternative diagnoses in this patient avoided the likely significant harm of a surgery.

Conflict of Interest
None declared.

References
1 LoRusso S. Disorders of the cauda equina. Continuum (Minneap Minn) 2021;27(01):205–224
2 Hida K, Iwasaki Y, Cho K, Imamura H, Abe H. Gliomas of the conus medullaris. Paraplegia 1994;32(01):52–58
3 Zalewski NL. Vascular myelopathies. Continuum (Minneap Minn) 2021;27(01):30–61
4 Muralidharan R, Saladino A, Lanzino G, Atkinson JL, Rabinstein AA. The clinical and radiological presentation of spinal dural arteriovenous fistula. Spine 2011;36(25):E1641–E1647
5 Hallan DR, McNutt S, Reiter GT, Thamburaj K, Specht CS, Knaub M. Dilated epidural venous plexus causing radiculopathy: a report of 2 cases and review of the literature. World Neurosurg 2020;144:231–237
6 Ghiasi M, Ghiasi M, Kahn E, Tomycz L, Ayad M, Aaronson O. Cauda equina syndrome secondary to an absent inferior vena cava managed with surgical decompression. J Neurosurg Spine 2012;16(02):201–205
7 Nasr DM, Brinjikji W, Rabinstein AA, Lanzino G. Clinical outcomes following corticosteroid administration in patients with delayed diagnosis of spinal arteriovenous fistulas. J Neurointerv Surg 2017;9(06):607–610
8 Egger K, Holenhuis M, Van Velthoven V, Heil S, Urbach H. Spinal diffusion tensor tractography for differentiation of intramedullary tumor-suspected lesions. Eur J Radiol 2016;85(12):2275–2280
9 Soni N, Bathla G, Pillenahalli Maheshvarappa R. Imaging findings in spinal sarcoidosis: a report of 18 cases and review of the current literature. Neuroradiol J 2019;32(01):17–28
10 Kaiboriboon K, Olsen TJ, Hayat GR. Cauda equina and conus medullaris syndrome in sarcoidosis. Neurologist 2005;11(03):179–183
11 Carvalho GBS, Sandim GB, Tibana LAT, Tertuliano FF, Idagawa MH, Abdala N. Magnetic resonance imaging in the differential diagnosis of infectious and inflammatory conus medullaris lesions. Radiol Bras 2013;46(01):51–55
12 Stern BJ, Royal W, Gelfand JM, Clifford DB, Tavee J, Pawate S, Baughman RP. Definition and consensus diagnostic criteria for neurosarcoïdosis: from the neurosarcoïdosis consortium consensus group. JAMA Neurology 2018;75(12):1546–1553
13 Yigit H, Yagmurlu B, Yigit N, Fitzs S, Kosar P. Low back pain as the initial symptom of inferior vena cava agenesis. AJNR Am J Neuroradiol 2006;27(03):593–595
14 Oterdoom DLM, de Jong BM, Hoogland PV, Groen RJM. Transient cauda equina compression syndrome and headache caused by internal vertebral venous plexus engorgement in a teenage female with vena cava inferior agenesis and iliac vein thrombosis. J Neurol Neurosurg Psychiatry 2007;78(11):1283–1284
15 Kamerath J, Morgan WE. Absent inferior vena cava resulting in exercise-induced epidural venous plexus congestion and lower extremity numbness: a case report and review of the literature. Spine 2010;35(18):E921–E924
16 Dudeck O, Zeile M, Poellinger A, Klubs L, Ludwig WD, Hamm B. Epidural venous enlargements presenting with intractable lower back pain and sciatica in a patient with absence of the infrarenal inferior vena cava and bilateral deep venous thrombosis. Spine 2007;32(23):E688–E691
17 Yugueros X, Alvarez B, Fernández E, Boquè M, Matas M. Compensative symptoms due to thrombosed or hypertrophic collateral circulation in infrarenal inferior vena cava agenesis. Ann Vasc Surg 2013;27(02):238.e9–238.e13
18 Wruuk E, Maj E, Dziedzic T, Podlecka Piętowska A. Spinal epidural venous plexus enlargement as a cause of neurologic symptoms: vascular anatomy and MRI findings. Neurol India 2020;68(05):1238–1241

Journal of Neurological Surgery Reports Vol. 83 No. R3/2022 © 2022. The Author(s).
19 Rodriguez FJ, Crum BA, Krauss WE, Scheithauer BW, Giannini C. Venous congestive myelopathy: a mimic of neoplasia. Mod Pathol 2005;18(05):710–718

20 Ebner FH, Roser F, Acioly MA, Schoeber W, Tatagiba M. Intramedullary lesions of the conus medullaris: differential diagnosis and surgical management. Neurosurg Rev 2009;32(03):287–300, discussion 300–301

21 Cohen-Gadol AA, Zikel OM, Miller GM, Aksamit AJ, Scheithauer BW, Krauss WE. Spinal cord biopsy: a review of 38 cases. Neurosurgery 2003;52(04):806–815, discussion 815–816

22 Yoon JW, Ganaha S, Watridge C. Vertebral venous collaterals with underlying agenesis of the inferior vena cava: implications for spinal surgery. World Neurosurg 2018;114:63–67