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## CASE REPORT

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# Acute Tumefactive Demyelinating Lesion Confined to Spinal Cord and Brainstem Mimicking Abscess: A Case Report

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## BACKGROUND

Primary demyelinating diseases of the central nervous system include several entities such as multiple sclerosis, neuromyelitis optica spectrum disorder, and acute demyelinating encephalomyelitis. Demyelinating disease may manifest as a solitary enhancing lesion that can resemble a neoplasm or abscess and poses a diagnostic challenge to both clinicians and radiologists. In the literature, these lesions are termed tumefactive demyelinating lesions (TDLs).<sup>1</sup> Supratentorial TDLs have been frequently described but there are very few cases reported with involvement confined to the spinal cord.<sup>2</sup>

## CASE REPORT

In March 2017, a 64-year old man was admitted to Tuen Mun Hospital, Hong Kong, with acute neck pain, right upper limb weakness with sparing of other limbs and sphincter disturbance with acute retention of urine.

Initial blood tests revealed a mildly elevated white cell count and erythrocyte sedimentation rate. Cerebrospinal fluid (CSF) analysis demonstrated elevated white cell count, high protein level and low glucose level. Magnetic resonance imaging (MRI) of the whole spine with contrast (Figure 1) showed a long-segment intramedullary

lesion with diffuse cord swelling extending from the craniocervical junction down to T6/7 level. MRI brain the next day (Figure 2) excluded a supratentorial lesion but revealed new involvement of the central medulla. The lesion demonstrated open ring enhancement with T2-weighted hypointense rim. Peripheral restricted diffusion with sparing of the central portion and dark apparent diffusion coefficient arc were also observed. Biochemical profile suggested infective myelitis with possible abscess formation. The patient was prescribed intravenous antibiotics and dexamethasone.

Despite antimicrobial treatment, the patient deteriorated rapidly with tetraplegia, and subsequently required intubation. New CSF analysis revealed a markedly elevated white blood cell count, further raised protein level and extremely low glucose. The antibiotic regimen was stepped up to include “big gun” antibiotics and antituberculous drugs. Dexamethasone was withheld.

Follow-up MRI of the whole spine on the following day showed rapid change in the pattern of the restricted diffusion of the medullary lesion that now also affected its central part (Figure 3). There was also further progression in the extent from brainstem down to T10 level.

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**Ethics Approval:** The patient was treated in accordance with the Declaration of Helsinki. Informed consent was obtained from the patient for all procedures.



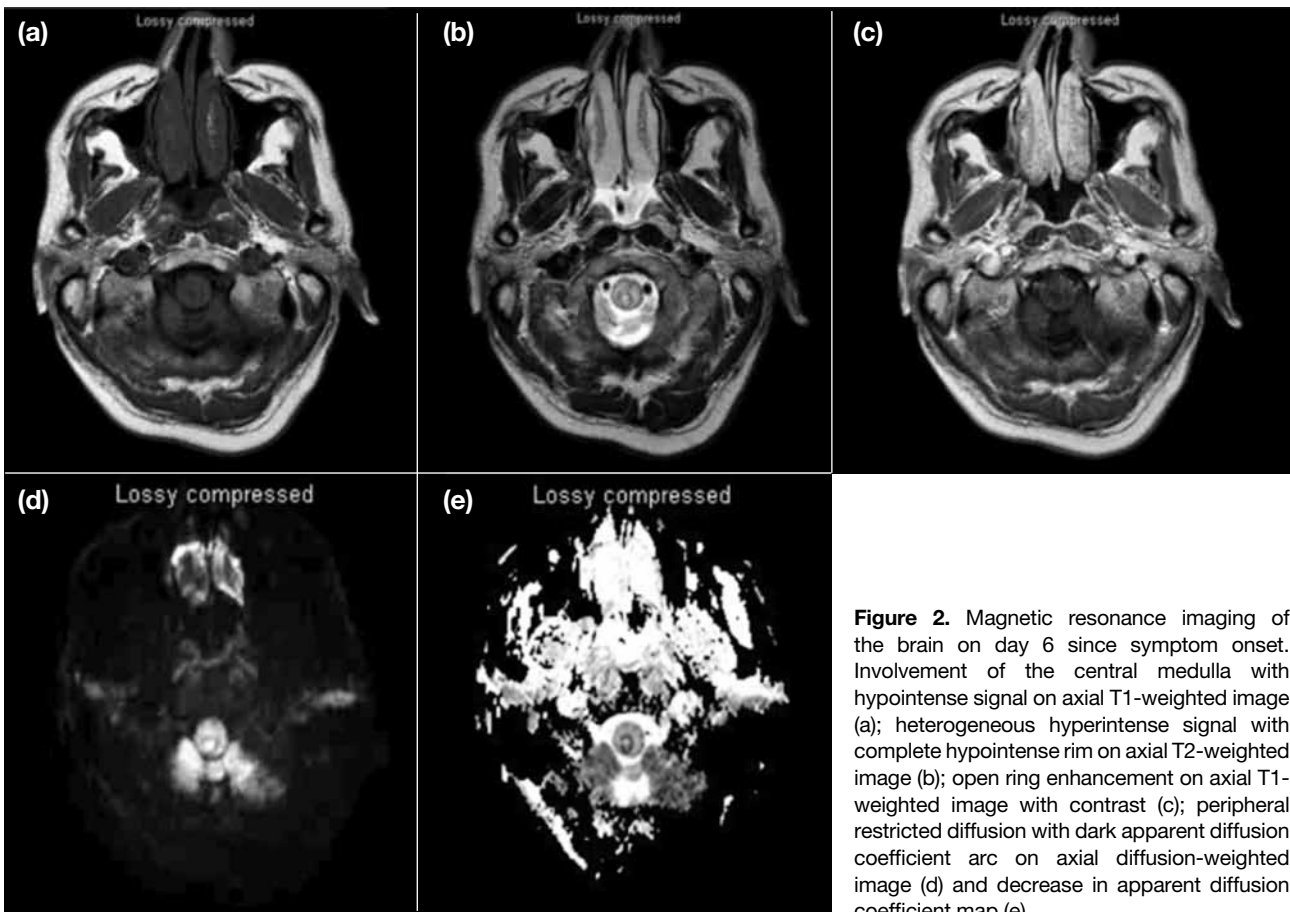
**Figure 1.** Magnetic resonance imaging of spine on day 5 since symptom onset. Long-segment intramedullary lesion with T2-weighted hyperintense signal and diffuse cord swelling from craniocervical junction down to T6/7 level on sagittal T2-weighted image (a); rim enhancement on sagittal T1-weighted image with contrast (b).

Despite treatment, the patient showed no clinical improvement and CSF viral polymerase chain reaction assays, cultures and autoimmune markers were all negative. The rapid change in radiological findings and absence of a response to antibiotic treatment raised the suspicion of acute inflammatory TDL. Antibiotics were withheld and pulse steroid therapy commenced. The patient's condition improved dramatically. He was extubated and regained almost full bilateral upper limb power although diplegia of the lower limbs persisted. CSF study also gradually normalised.

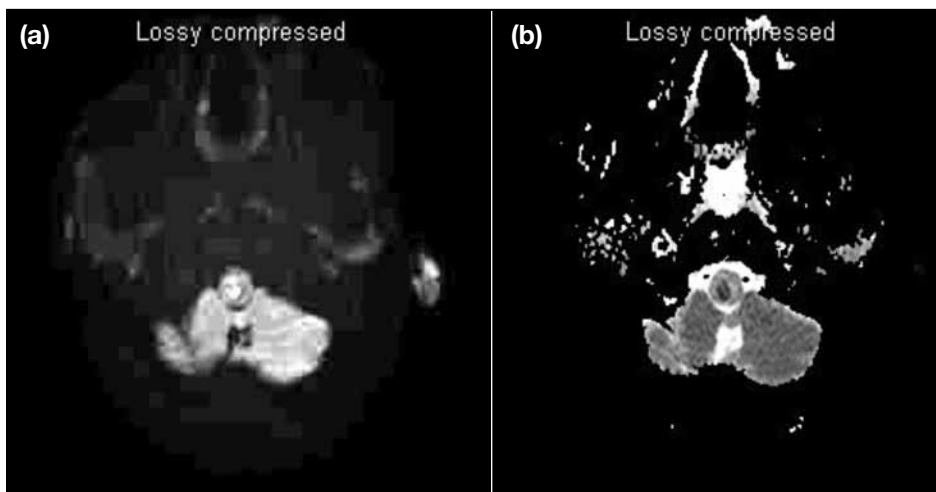
Subsequent follow-up MRI studies (Figure 4) revealed regression of the intramedullary lesion with residual signals at the posterior aspect of the cervical cord and thoracic cord atrophy.

## DISCUSSION

The definition of a TDL includes size >2 cm with mass effect and contrast enhancement. The current literature on TDLs limited to the spinal cord and brainstem is scarce: most focuses on supratentorial lesions. Owing to



**Figure 2.** Magnetic resonance imaging of the brain on day 6 since symptom onset. Involvement of the central medulla with hypointense signal on axial T1-weighted image (a); heterogeneous hyperintense signal with complete hypointense rim on axial T2-weighted image (b); open ring enhancement on axial T1-weighted image with contrast (c); peripheral restricted diffusion with dark apparent diffusion coefficient arc on axial diffusion-weighted image (d) and decrease in apparent diffusion coefficient map (e).



**Figure 3.** Magnetic resonance imaging of spine on day 7 since symptom onset. Rapid change in the pattern of the restricted diffusion of the lesion involving the medulla, now affecting its central part as well on axial diffusion-weighted image (a) and decrease in apparent diffusion coefficient map (b).



**Figure 4.** Magnetic resonance imaging (MRI) of the spine at 3 weeks after symptom onset. Interval reduction of the cord swelling and oedema of the upper segment of lesion from cervical to upper thoracic level (a) but increased caudal extension to involve the conus on sagittal T2-weighted images (b). Magnetic resonance imaging of the spine at 3 months after symptom onset. Significant regression of the lesion with residual signals at the posterior aspect of the cervical cord (c) and thoracic cord atrophy (d) on sagittal T2-weighted images.

significant overlap in clinical presentation and imaging findings, such a lesion frequently poses a diagnostic challenge to distinguish it from tumour or abscess.

Our patient presented with an acute and rapidly progressive solitary lesion that affected the spinal cord and brainstem with no supratentorial involvement.

The clinical and radiological differential diagnoses included infective, autoimmune and paraneoplastic causes. The initial CSF analysis raised the suspicion of infective myelitis although subsequent clinical progress and negative extensive microbiological investigations made this unlikely. Without an antecedent history of malignancy or rheumatological disease, paraneoplastic

or secondary autoimmune causes were less likely. Considering primary autoimmune causes, the extensive and aggressive nature of the lesion was inconsistent with multiple sclerosis. Neuromyelitis optica spectrum disorder can have extensive myelitis but should show supratentorial or optic nerve involvement with positive anti-neuromyelitis optica. Meticulous review of the clinical, microbiological and radiological features enabled the diagnosis of acute TDL of the spinal cord and brainstem to be established.

Imaging findings of TDLs typically include T1-weighted hypointense and T2-weighted hyperintense signals with mass effect and oedema proportional to the lesion size. T2-weighted hypointense rim may be observed.<sup>3-5</sup> The pattern of ring enhancement is not differentiating as both open and closed ring enhancement have also been reported in TDLs, abscesses and neoplasms. Peripheral restricted diffusion with a dark apparent diffusion coefficient arc or ring at the lesion edge reminiscent of that seen in our patient is significantly more common in TDLs,<sup>6</sup> as an abscess tends to have central restricted diffusion due to the highly viscous abscess cavity.<sup>7</sup> Such finding also echoes our previous report of a case of supratentorial TDLs in acute demyelinating encephalomyelitis complicating human swine influenza (H1N1).<sup>8</sup> Recognition of the rapid temporal changes on serial imaging to apparent diffusion coefficient patterns of TDLs is helpful as such changes would not be expected in a neoplastic process.<sup>6</sup>

TDL is a rare but important entity when an acute spinal cord lesion is encountered; early recognition may avoid the need for invasive biopsy that is often not feasible in a critically ill patient and may result in significant neurological deficit. Although the radiological characteristics of brain and spinal cord TDLs are similar, radiological diagnosis in the spinal cord is much more

difficult due to the limited usage of diffusion-weighted MRI, perfusion-weighted MRI and spectroscopy as a result of small tissue volume and imaging artefacts compared with that of the brain. Meticulous analysis of the radiological and clinical findings is thus of utmost importance to establish the diagnosis.

In conclusion, TDL confined to the spinal cord and brainstem without supratentorial involvement is rare and often proves a diagnostic challenge to both clinicians and radiologists. Familiarity with the clinical and radiological findings is crucial for accurate and early diagnosis, thus guiding appropriate treatment and optimising clinical outcomes.

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