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## INFLAMMATORY DEMYELINATING DISEASE MIMICKING AN INTRA-MEDULLARY CERVICAL CORD TUMOR: A CASE REPORT

Yu-Ray Shen MD, Hwa-Jeng Yan MD, Chien –Min Chen, MD

Department of Surgery, Division of Neurosurgery, Changhua Christian Hospital, Changhua, Taiwan

### ABSTRACT

We report a case of tumefactive demyelinating lesion in the cervical cord without intracranial involvement, mimicking an intramedullary cervical spinal cord tumor. This young girl had a progressive right sided hemiparesis (1/5 strength) and paresthesia. The Horner's syndrome was also noted. The cervical MRI revealed an ill-defined high signal lesion on T<sub>2</sub>WI images and partial enhancement with gadolinium located at C1 to C3 levels. Significant perifocal white matter edema was displayed from C1 to T2 levels and an intramedullary spinal tumor was highly suspected. Since preoperative examinations cannot differentiate spinal cord tumor from the other intramedullary cord lesions such as demyelinating disease, surgical intervention would be approved in such atypical primary spinal cord multiple sclerosis. Physicians should be alerted that demyelinating disease can mimic a spinal cord tumor, even on MRI, and must be considered in the differential diagnosis of a symptomatic spinal cord mass.

**Key words:** inflammatory demyelinating disease ♠ multiple sclerosis ♠ cervical cord tumor

### Objective and Importance

Since the invention of magnetic resonance imaging, it has become possible and convenient to better image lesions of multiple sclerosis. However, in some cases with primary spinal cord multiple sclerosis, they are difficult to diagnose on imaging features alone. Regional spinal cord enlargement, mimicking an intramedullary tumor is less common <sup>[1]</sup>. Multiple sclerosis involving the spinal cord without intracranial lesions has been reported in up to ten percentage <sup>[2]</sup> of patients with multiple sclerosis. This case emphasizes the difficulty in diagnosing rapidly progressing regional intramedullary lesion.

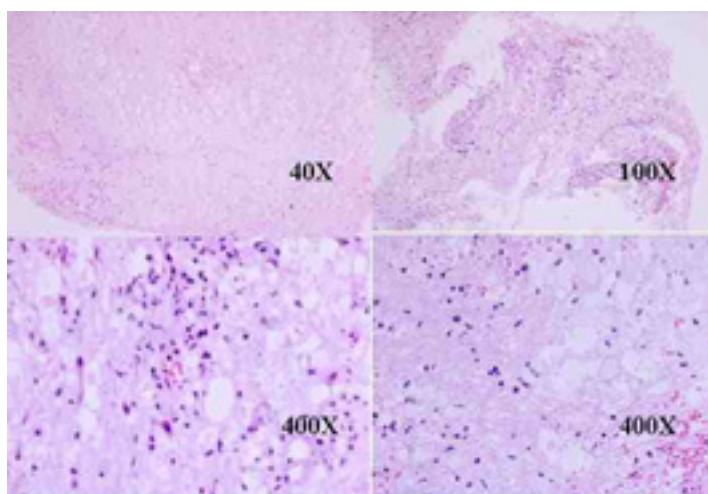
### Clinical Presentation

A 9-year-old girl had presented with a history of neck pain and tenderness for one week. Rapidly There was progressive right side hemiparesis (1/5 strength), paresthesia and right ptosis for four days,

presenting as a suspected cervical spine lesion with Horner's syndrome. There were no visual deficits during the admission. Physical examination revealed right ptosis, right miosis, right facial anhidrosis, right neck muscle hypertrophy and positive Babinski sign (right side upward). Lumbar puncture had a normal opening pressure. The CSF study showed colorless, with a white cell count of 0/mm<sup>3</sup> and the protein of 26 mg/dl, and glucose 59 mg/dl. The cervical MRI (Fig. 1) revealed an ill-defined lesion from C1 to C3 with partial gadolinium enhancement. Significant perifocal white matter edema was displayed from C1 to T2 level; this was felt to be an intramedullary spinal tumor. Surgical intervention was performed because of exacerbation of right side weakness. Laminectomy from C1 to C7 was performed for decompression. Then, the dura mater was incised in the midline from C1 to C7, and the severe cord swelling was noted. The myelotomy was performed gently in the midline via central sulcus. Under the microscopic field, poorly demarcated lesion was



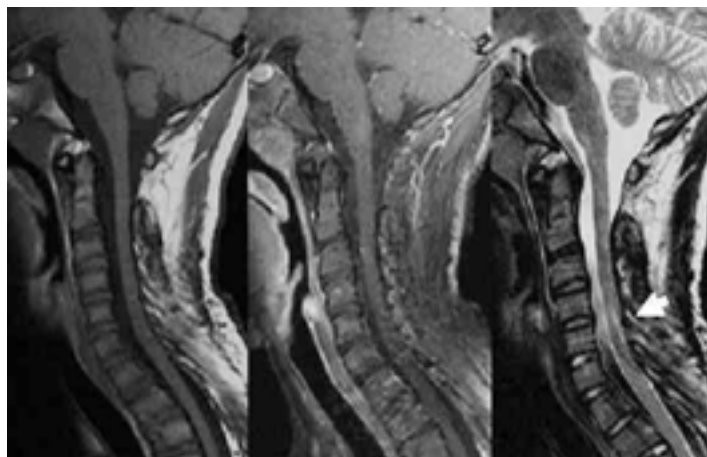
**Figure 1:** A) Sagittal T1-weighted MRI of the cervical spine showed an ill-defined lesion (arrow) with swelling of spinal cord. B) T1-weighted image after intravenous gadolinium showed partial enhancement (arrowhead) in the C1 to C3 level. C) T2-weighted image showed significant perifocal white matter edema (arrow) from C1 to T2 level.



**Figure 2:** Areas of marked myelin vacuolation confirmed with histological examination suggestive of intramyelinic edema. No evidence of neoplasm or infection.

found and biopsied. The pathological specimens showed focal aggregation of macrophages, scattered lymphoplasmacytic infiltrates and gliosis. Special stain revealed focal marked myelin vacuolation, suggestive of intramyelinic edema (Fig. 2). The histological examination showed no evidence of neoplasm or infection. Based on the clinical findings and pathologic report an inflammatory demyelinating disease was diagnosed. High-dose intravenous methylprednisolone was administered.

After the operation, the power in her right hand gradually improved to 2/5. Then, she was transferred



**Figure 3:** Slight increased linear high T2 signal, located in right side of the spinal cord from the level of the lower border of C2 to upper border of C6 (arrow).

to rehabilitation unit and was followed-up in Pediatric out-patient clinic. Her power recovered to 4/5 strength in right upper extremity two years later. The follow-up cervical MRI showed slight increased linear high T2 signal, located in right side of the spinal cord from the level of the lower border of C2 to upper border of C6 (Fig. 3). The finding is consistent with focal malacia of the spinal cord from the previous spinal lesion.

## Conclusion

According to the radiological and pathological findings, acute inflammatory demyelinating disease is the most likely diagnosis. This is interesting because that the lesion is solitary and confined to medulla of cervical spinal cord on MRI imaging. The possible range of diagnoses of intrinsic spinal cord lesions are legion [3]. In previous clinical series, correlation between the level of the lesion and symptoms is well identified in the spinal cord [1-5]. The mimicking cervical spinal tumor is probably due to regional cord edema, which cause high signal in T<sub>2</sub>WI MRI [2]. This reflects blood-brain-barrier disruption, which is an early, transient phenomenon, due to perilesional infiltration, usually persisting less than five weeks [6]. Regional histological change, which with an intense macrophage infiltration, causes marked enhancement. Enlargement of the spinal cord is seen only during acute stage, due to spinal cord swelling. So, tumefactive demyelinating lesions may enhance, and show perilesional edema and thus be mistaken for neoplasms. MRI findings are nonspecific and may simulate ischemic, neoplastic or inflammatory lesion. MRS may not be able to differentiate from neoplasm [7-10]. Biopsy may be considered in some case if it is still difficult to distinguish from neoplasm. Maybe the biopsy is not an appropriate procedure, because it may cause permanent spinal cord damage. In this case, we would stress the importance of surgical decompression.

Even, the inflammatory disease has a self-limiting course and is not suited to surgical intervention. But, the decompression with long segments laminectomy and duroplasty can improve spinal cord perfusion and decrease the insult of ischemia. In the future, we may need more image studies or intraoperative equipments to assess the possibility of the inflammatory demyelinating disease to prevent the inadequate surgical procedure.

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**Correspondence:** Yu-Ray Shen, M.D, Division of Neurosurgery, Department of Surgery, Changhua Christian Hospital, 135 Nanhsiao Street, Changhua 500, Taiwan. FAX: + 886-4-7228289; TEL: + 886-4-7238595 ext. 1376; E-mail: 108936@cch.org.tw

## Comments

This is an interesting manuscript of an inflammatory process, multiple sclerosis, that has radiographical features suggestive of an intramedullary spinal cord tumor. The authors should be commended on this report, as they investigated the brain prior to surgery. There was no features on the brain MRI to suggest an inflammatory process, and thus a biopsy was performed. The features which suggest an inflammatory process rather than an intramedullary tumor, is the short history of progressive neurological deficits. Typically spinal cord tumors are benign and have an indolent or slowly progressive course, unlike the inflammatory processes. In terms of imaging, the reader should be aware that these inflammatory lesions are typically subpial and have patchy enhancement unlike a spinal cord tumor. Regardless this is an interesting case report of an inflammatory process in a young child which was treated in a prompt fashion.

**George Jallo MD**  
Baltimore  
gjallo1@jhmi.edu

The authors of this interesting case report describe a patient in whom magnetic resonance imaging (MRI) revealed a lesion that distended the cervical spinal cord and showed patchy enhancement after the injection of gadolinium contrast. The diagnosis of an intramedullary spinal cord tumor was suspected, and the patient underwent a laminectomy. Biopsy of the spinal cord showed that the patient suffered from an acute necrotizing demyelinating condition. As presented, the case raises two interesting and important points for discussion.

The patient developed a severe neurologic deficit with only 1/5 strength in the right upper extremity. The deficit evolved over a brief period. Intramedullary spinal cord tumors, intramedullary



ependymomas, and astrocytomas present in an insidious manner. Often, subtle symptoms are present for years before the tumor is diagnosed definitively. Malignant intramedullary astrocytomas can become symptomatic more abruptly. With the current state of knowledge and treatment, these tumors are uniformly fatal (3).

If the only information available to the neurologist or neurosurgeon is the MRI study, the diagnosis of intramedullary spinal cord tumor would be reasonable and surgery would be the appropriate intervention. In this case, however, the clinical picture was inconsistent with the diagnosis of intramedullary spinal cord tumor. The diagnosis of transverse myelitis or, as seen in this case, inflammatory demyelination would be the more likely diagnosis. Surgical biopsy is a reasonable approach, but presumptive treatment as if it were an aggressive form of multiple sclerosis would also be reasonable for a few weeks to see if the patient's course stabilizes. Based on the clinical information, doing so would not place the patient at risk and surgery could be avoided.

The second issue relates to the presentation of a patient with a right Horner's syndrome. Upper motor neuron lesions causing Horner's syndrome are recognized in infarction of the lateral medulla. The upper motor neurons of the sympathetic nervous system travel from the hypothalamus to the lateral medulla. They then exit from the intermediolateral cell column from C8 to the lower thoracic spinal cord. A lower motor neuron presentation of Horner's syndrome arises from the paraspinal ganglia. Horner's syndrome is much more frequently related to trauma or thoracic tumors. I personally have never seen a patient with pathology limited to the cervical spinal cord. In the literature related to Horner's syndrome, there are few reports of a patient with pathology limited to the cervical spinal cord presenting with these findings. Cross

referencing Horner's syndrome with intramedullary spinal cord tumors yielded two reports, one of which was an intramedullary lymphoma (4) and one of which was an ependymoma (1). Syringomyelia can become symptomatic with a Horner's syndrome (2, 5).

These two issues point to the importance of understanding the clinical setting in which the patient is seen. At this time neurosurgeons benefit from the elegant delineation of anatomy on MRI. A good history and attention to the neurologic examination remain an essential part of surgical decision making.

#### Harold Rekate MD

Phoenix  
neuropub@chw.edu

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