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Case Report

Isolated large demyelinated plaque with clinical and radiologic appearance suggestive of cervical intramedullary tumor diagnosed after surgery ☆,☆☆

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ABSTRACT

Isolated spinal demyelinating lesions are rare and often associated with multiple sclerosis. While initial radiological findings may suggest a tumor, a definitive diagnosis requires a histological diagnosis. A 45-year-old woman presented with progressive spastic tetraparesis for 1 week. She had no prior history of neurological or systemic illness. Brain and thoracic magnetic resonance imaging (MRI) were normal, but cervical MRI revealed an intramedullary tumor extending from C3 to C4. Surgery was performed. Histopathological examination revealed an inflammatory demyelinating plaque, not a tumor. The patient experienced significant improvement in her clinical condition postsurgery and remains under neurological follow-up. We discuss this case alongside a review of similar cases reported in the literature, focusing on clinical presentation, laboratory findings, MRI features, and follow-up of patients with tumor-like inflammatory demyelinating diseases of the spinal cord initially diagnosed as intramedullary tumors.

Introduction

Inflammatory demyelinating pseudotumor, also known as a pseudotumor, typically affects the brain but rarely occurs in the spinal cord [1–3]. On imaging, inflammatory demyelinating pseudotumor appears strikingly similar to intramedullary tumors, like astrocytomas. This resemblance often leads to misdiagnosis and surgical resection. We reviewed the medical literature and analyzed a series of 36 patients diagnosed with inflammatory demyelinating pseudotumor of the spinal cord based on clinical presentation, magnetic resonance imaging (MRI) findings, and pathological confirmation [1,3,4]. Most patients experienced an acute or subacute onset with sensorimotor impairment. However, only 6 patients received a definitive diagnosis of inflammatory demyelinating pseudotumor

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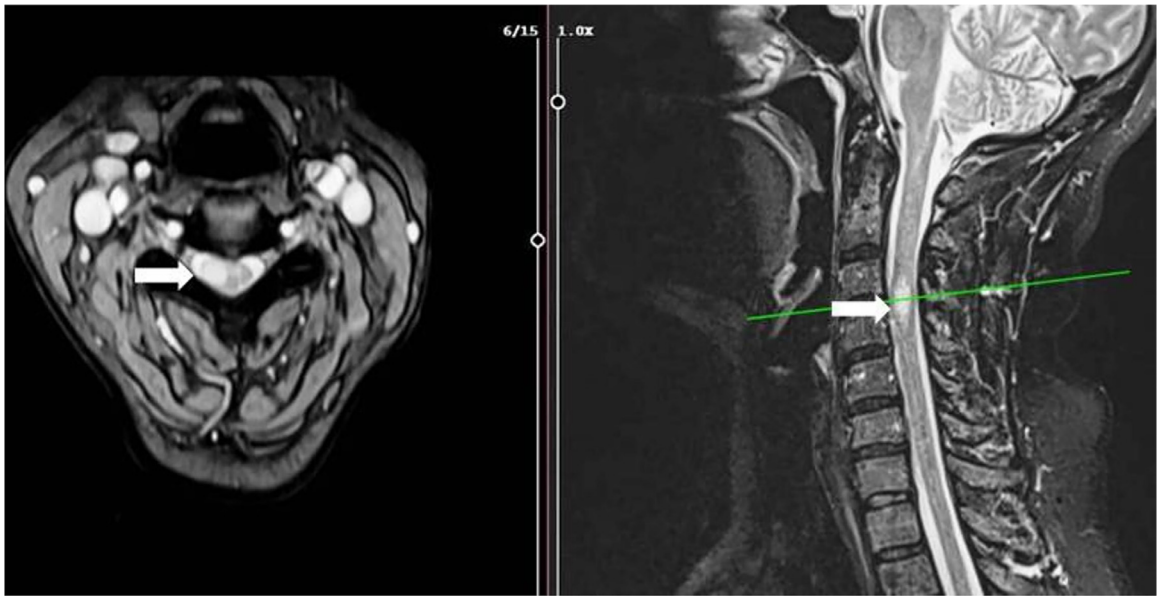

Fig. 1 – Preoperative axial and sagittal contrast-enhanced magnetic resonance imaging (MRI) scan of a 45-year-old woman with tumor-like inflammatory demyelinating disease. The image demonstrates a solitary intradural mass at the C3-C4 level of the cervical spinal cord (Indicated by the white arrow).

through pathological examination, including the case presented here. This limited number highlights the rarity of this condition [1–4].

Pathologic analysis revealed perivascular cuffing dominated by lymphocytes alongside evidence of demyelination [1,2,5,6]. These findings support the classification of tumor-like inflammatory demyelinating disease of the spinal cord as a specific type of demyelinating disease categorized as an inflammatory pseudotumor [1,2,6]. The solitary nature of these lesions makes them prone to misidentification as intramedullary neoplasms [1,4,6].

Case

Presentation and examination

A 45-year-old woman presented with a 1-week history of worsening weakness on her right side. Due to significant muscle weakness, she was started on prednisolone and underwent an MRI scan. The MRI revealed an intramedullary tumor extending from C3 to C4 (Fig. 1). She was then referred to our clinic for further evaluation. Her neurological examination revealed a Glasgow Coma Scale score of 15 and significant right-sided weakness, approaching complete right plegia.

Operation

The patient underwent surgery under total intravenous anesthesia with neuromonitoring guidance. She was positioned prone for the procedure. A midline incision was made from C2 to C5 to access the surgical site. Due to limited space and the tumor's location near the antero-lateral region of the spinal cord, a total laminectomy was performed. The dura mater was then opened and suspended. A myelotomy was performed to access the tumor. The tumor was found to be firmly attached to the spinal cord and had a grayish appearance. It was meticulously removed piece by piece. Ultrasound was used again to confirm complete removal (Fig. 2). After achieving hemostasis, the dura was primarily closed, and the surgical site was stabilized using a short-segment lateral mass screw technique.

Pathological findings

Microscopic examination of the removed tissue revealed reactive gliosis, infiltration of macrophages within the spinal cord tissue, and collections of lymphocytes around the blood vessels, with a predominance of CD3+ T-cells. The Ki-67 proliferation index was low in the inflammatory infiltrate. These findings suggest a chronic inflammatory process, possibly including demyelinating disease (Fig. 3). Importantly, there was no evidence of a neoplastic process. Cerebrospinal fluid analysis, which can help diagnose demyelinating diseases, did not reveal any oligoclonal bands.

Postoperative course

Postoperative MRI scans of the brain and spinal cord did not reveal any evidence of demyelinating disease (Fig. 4). The patient continued to receive dexamethasone after surgery and experienced significant improvement over the following months. Following inpatient rehabilitation, she regained most of her upper-extremity strength and showed moderate improvement in her lower-extremity strength and walking ability. Her pain significantly improved.

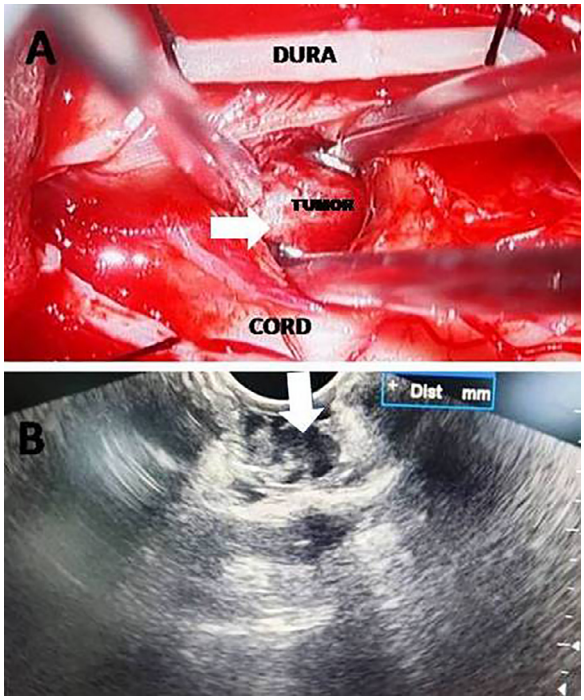

Fig. 2 – (A) The tumor appears to be tightly attached to the spinal cord. (B) Ultrasound was used again to confirm complete removal (indicated by the white arrow).

Discussion

Multiple sclerosis (MS) is the most common primary demyelinating disease of the central nervous system (CNS). It typically affects people between 10 and 50 years old, with a peak around 30. Women are diagnosed with MS about twice as often as men [1–4].

In the spinal cord, MS lesions are most commonly found in the cervical region. Isolated spinal cord lesions occur in 10%-20% of MS cases. Symptoms vary depending on the affected spinal cord segment and the degree of exposure. They include paresthesia, spastic paresis, paraplegia, hyperreflexia, neurogenic bowel and bladder, and sexual dysfunction. Spinal MS plaques tend to be located on the outer edge of the spinal cord and often cross boundaries between white and gray matter [1,2,5,6]. They are typically smaller than 2 vertebral bodies and are usually located in the dorsolateral cord regions. Unlike cerebral lesions in MS, spinal cord lesions rarely exhibit low signal intensity on T1-weighted MRI images. Most MS plaques in the spinal cord appear as hyperintense. On T2-weighted images, larger active lesions may show cord enlargement and intense edema. Chronic lesions often exhibit focal cord atrophy [1–4].

The etiology of tumor-like inflammatory demyelinating disease is unknown. A possible link exists between viral infections and autoimmune processes leading to myelin damage. This condition typically affects young and middle-aged adults [2,3,4,6]. Most cases present with symptoms of acute or subacute inflammatory demyelinating disease, primarily affecting one side of the posterior and lateral spinal cord regions. Initial symptoms often involve superficial sensory disturbances, followed by concurrent motor disturbances. Motor disturbances can include monoplegia, paraplegia, quadriplegia, and hemiplegia [3,5,6]. Lesions typically appear as single focal lesions in the white matter of the lower cervical and upper thoracic spinal cord. They often have clear borders and edema in the peripheral region. A ring-shaped enhancement pattern on MRI scans with contrast dye is a strong indicator of this condition.

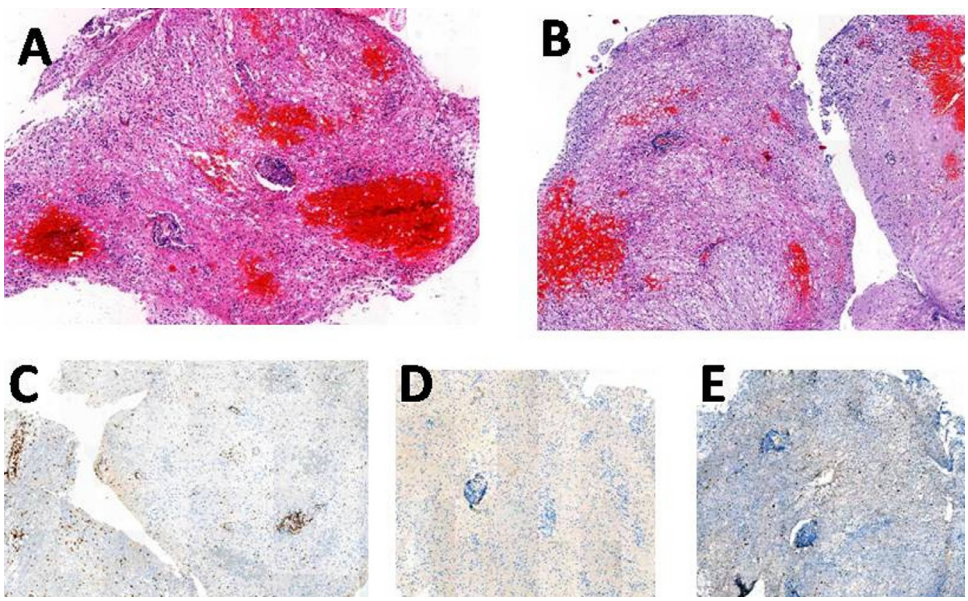

Fig. 3 – (A and B) Numerous macrophages and perivascular lymphocytes are observed within the spinal cord parenchyma. (C) Immunohistochemical staining with CD3 antibody highlights perivascular T lymphocytes (D) Scattered CD 20+ B lymphocytes are also present. (E) The inflammatory infiltrate shows low Ki-67 positivity.

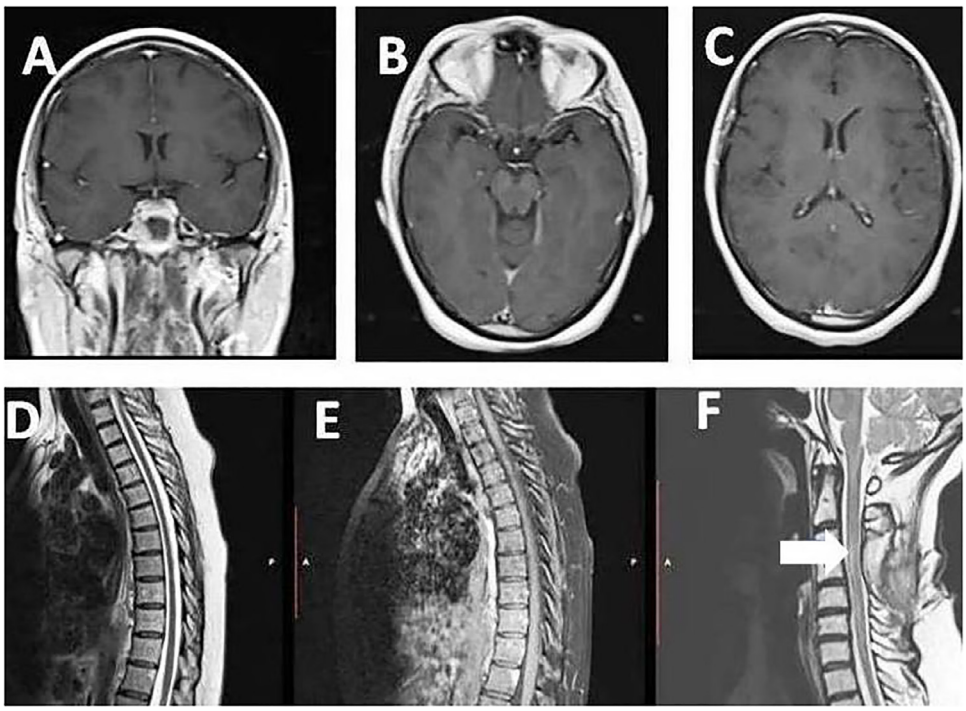

Fig. 4 – (A-C) No demyelinating plaque was identified on contrast-enhanced brain MRI scans performed to screen for such lesions. (D, E) No demyelinating plaque was seen on thoracic MRI scans performed for screening purposes. (F) Postoperative contrast-enhanced cervical MRI shows complete resolution of the previously identified mass in the C3-C4 region (Indicated by the white arrow).

Up to 66% of cases may exhibit this symptom during the acute phase [1–4].

Demyelinating lesions can mimic tumors due to their space-occupying effect. While the surrounding edema in demyelinating lesions may decrease over time, gliomas typically show ongoing growth and a well-defined border. This similarity makes differentiating them from demyelinating pseudotumors challenging [1–5]. However, gliomas are located in the central region of the spinal cord and often present with syringomyelia, while demyelinating pseudotumors tend to be localized in the white matter of the spinal cord and do not usually cause syringomyelia. MRI findings often hold greater weight than clinical examinations in differentiating these conditions. Primary CNS lymphoma lacks the pathological features of a demyelinating pseudotumor, such as demyelination and involvement of the deep gray matter [1–4].

Conclusion

In summary, tumor-like inflammatory demyelinating lesions of the spinal cord can be easily misdiagnosed as true intramedullary tumors due to their space-occupying effect. To differentiate these lesions from tumors, lumbar puncture, immunological examinations, electrophysiological studies, and neuroimaging examinations are needed. Biopsy should be considered if symptoms worsen rapidly and the space-occupying effect persists. Radiotherapy and surgery are not recommended as initial interventions. For some isolated space-occupying lesions, a spinal cord biopsy offers a safe and minimally invasive approach for diagnosis compared to more aggressive procedures.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

CRediT authorship contribution statement

Askin Esen Hasturk: Conceptualization, Data curation, Formal analysis, Writing—original draft, Writing—review and editing. **Taha Eser:** Conceptualization, Data curation, Formal analysis, Writing—review and editing.

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