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## Case Report

# Intracranial Demyelinating Pseudotumor: A Case Report and Review of the Literature

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## ABSTRACT

Demyelinating pseudotumor is a rare inflammatory demyelinating disease of the central nervous system (CNS) that has a similar clinical presentation and computed tomography (CT) and magnetic resonance imaging (MRI) imaging findings as brain tumors or abscesses. Unlike brain tumors, demyelinating pseudotumors respond well to steroid hormones. There are only a few reported cases of intracranial demyelinating pseudotumors in the literature. In this case report, we present the diagnosis and treatment of demyelinating pseudotumor in a patient whose condition was initially misdiagnosed as an astrocytoma. Based on the literature and our case, we formulated an outline for the differential diagnosis of demyelinating pseudotumor and astrocytoma. A timely and correct diagnosis of demyelinating pseudotumor would avoid blind surgery, radiotherapy and chemotherapy, which are used to treat brain tumors.

**KEYWORDS:** Demyelinating pseudotumor, Diagnosis, Cord-Like enhancement, Enhanced MRI

## ■ INTRODUCTION

Demyelinating pseudotumor of the brain, also called tumor-like demyelinating lesions (TDLs) or inflammatory demyelinating pseudotumor (IDP), is a rare inflammatory demyelinating disease of the central nervous system (CNS) (3,4,7,8). The exact etiology of demyelinating pseudotumor is unclear, however it may associate with multiple sclerosis (MS) (5). Based on the clinical presentation and computed tomography (CT) and magnetic resonance imaging (MRI) findings, it is difficult to distinguish between brain tumors, abscesses and demyelinating pseudotumors (13). Unlike brain tumors, demyelinating pseudotumors respond well to steroid hormones (6). Therefore, a reliable distinction between brain tumors and intracranial demyelinating pseudotumors is important for a targeted therapy. Although the incidence of demyelinating

pseudotumor in the clinic is relative low, delayed diagnosis or misdiagnosis may cause delays in treatment of these patients and subsequently may lead to physical and mental damage (14). The differential diagnosis of demyelinating pseudotumor and brain tumors, especially of low-grade astrocytomas, should arouse more attention to exclude the possibility of a demyelinating process. Despite this, only few reported cases of intracranial demyelinating pseudotumor are known in the literature. In this case report, we present the diagnosis and treatment of a demyelinating pseudotumor patient who was misdiagnosed with astrocytoma.

## ■ CASE REPORT

A female 40-year-old patient was hospitalized due to a progressive disability of the left limb that severely increased over one week. The patient did not have a recent history



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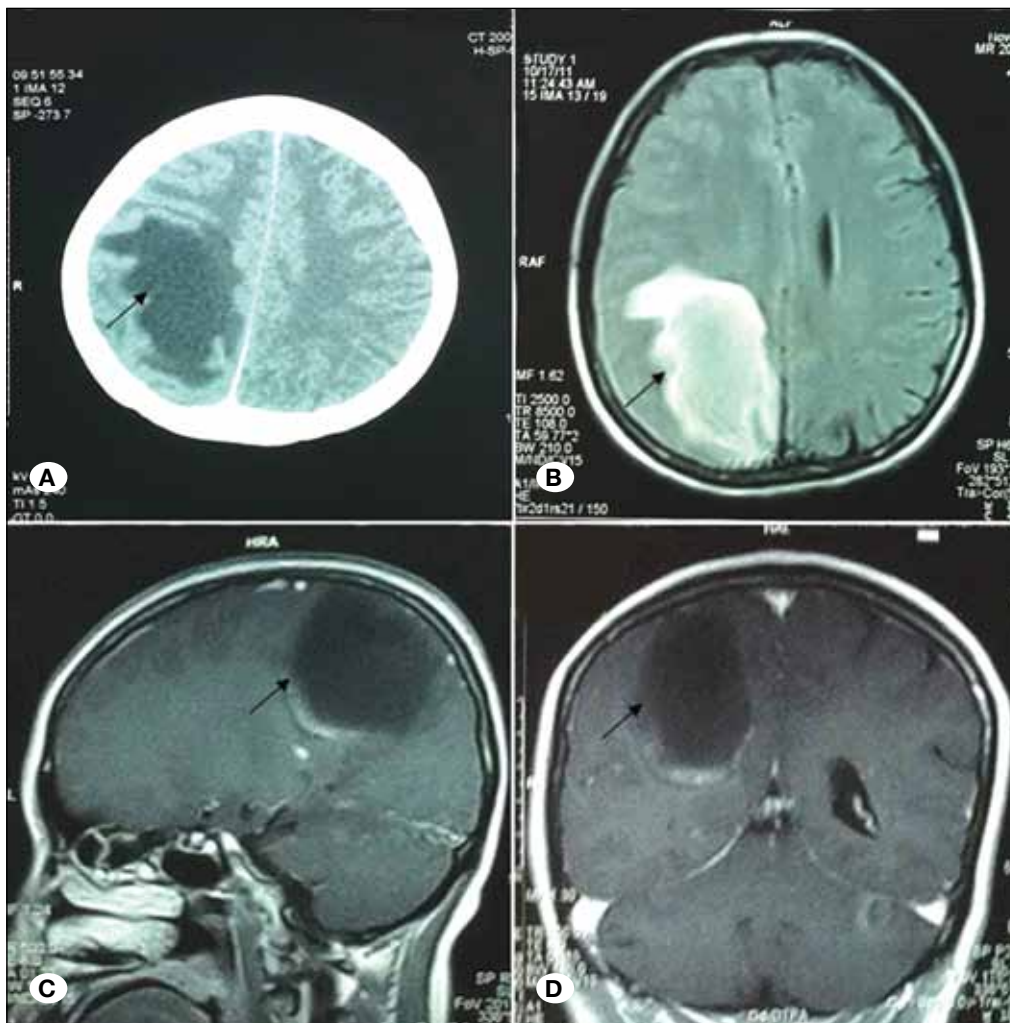
of fever or drug applications. The admission examination revealed that the patient was conscious, fluent, had a soft neck, light reflex sensitivity and hyperreflexia of the left knee, but the right side was normal. Her body temperature was 36.9°C. Her pupils were the same size bilaterally and the pupillary light reflex was unaffected. The strength of the left and right limb muscle were graded as 3 and 5, respectively. A head CT showed a large low density area at the right parietal lobe and compressive deformation of the right lateral ventricle. A brain MRI with T1WI (T1 weighted image), T2WI and fluid attenuated inversion recovery (FLAIR) showed an abnormal signal at the right parietal lobe. Enhanced scans showed that the edge of the “lump” was in the sagittal plane and that a cord-like enhancement was distributed vertical to the lateral ventricles in the coronal plane (Figure 1A-D). A preoperative diagnosis of glioma was made, and craniotomy tumor resection was performed.

Intraoperative findings revealed that the “mass” had a yellowish foam texture, was avascular, and had clear boundaries to the surrounding tissue. The “mass” was completely resected and two weeks following the surgery, pathology showed that the “mass” was a demyelinating pseudotumor. Immunohisto-

chemistry (IHC) revealed the following markers: Cluster of Difference 68 (CD68) (+), Myelin basic protein (MBP) (+), Vimentin (Vim) (+), Glial Fibrillary Acidic Protein (GFAP) (-) (Figure 3A-C). While waiting for the pathology report, the patient did not receive steroid hormone therapy. Three weeks after surgery, the hemiplegia became worse and the patient received steroids. The patient was treated with methylprednisolone pulse therapy (1000 mg/d), followed by oral prednisone (1 mg/kg) for two weeks. The patient was discharged from the hospital after two weeks. At the last examination, the left limb muscle strength was grade 4, the right limb muscle strength was grade 5, and the left knee showed active tendon reflexes and a left Babinski sign. Prednisone was given in tapering doses over a 2-month period. Follow-up was performed at 11 months postoperative, and the imaging is shown in Figure 2A-D. The patient recovered well, the left limb muscle strength was grade 4+ and the right limb muscle strength was grade 5. The left knee tendon reflexes were normal and without a Babinski sign.

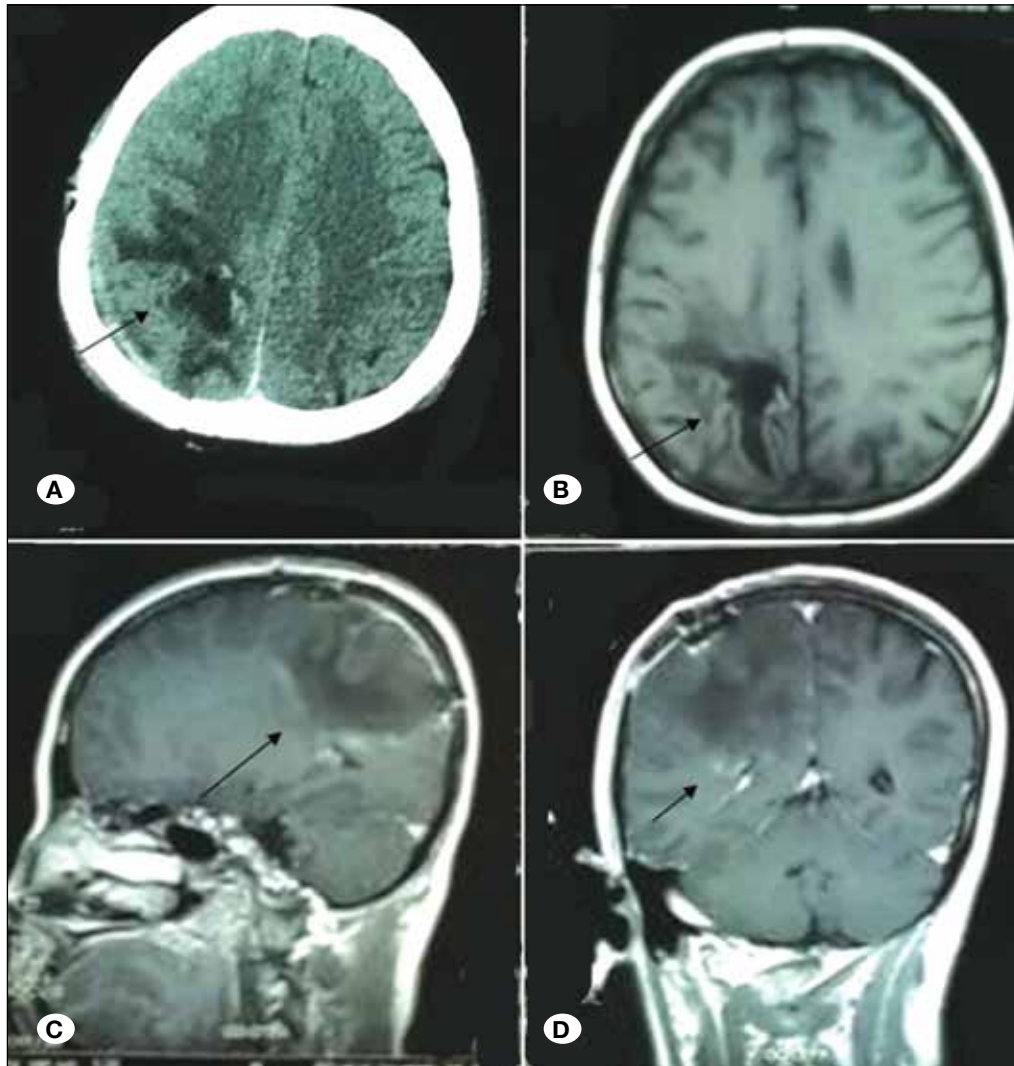
## DISCUSSION

Demyelinating pseudotumor of the brain is a very rare demyelinating disease affecting the CNS. Previous reviews have

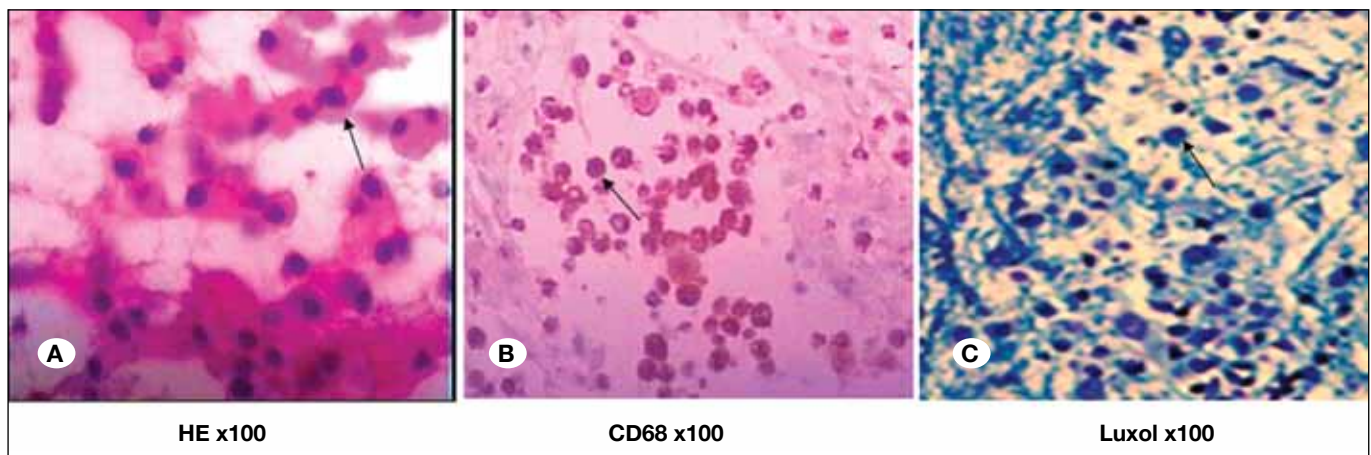


**Figure 1:** CT and MRI before Treatment.

**A)** Axial CT showed a large low density area at the right parietal lobe (arrow).  
**B)** MRI with FLAIR showed an abnormal signal at the right parietal lobe (arrow).  
**C, D)** Enhanced scans showed that the edge of the “lump” was in the sagittal plane and a cord-like enhancement (arrow) was distributed vertical to the lateral ventricles in the coronal plane.



**Figure 2:** CT and MRI after Treatment. **A, B)** Axial CT and MR images showed that the large low density area at the right parietal lobe disappeared (arrow). **C, D)** Enhanced MR image showed that the “lump” and the cord-like enhancement of the edge of the “lump” disappeared (arrow).



**Figure 3:** Pathologic finding revealed demyelinating pseudotumor. **A)** HE staining of the removed brain tissue, which showed accumulations of macrophages (arrow). **B)** Macrophages (arrow), but no swollen astrocytes were detected with CD86 staining. **C)** Luxol fast stains showed some fiber and extensive destruction of myelin with macrophage infiltration (arrow).

shown that demyelinating pseudotumor can occur at any age with an increased tendency in young and middle-aged females (8,20). The patient in this report was a 40-year-old female. There are no specific radiology and clinical manifestations of intracranial demyelinating pseudotumor and the accurate diagnosis relies on pathology results. In 1979, van der Velden et al. (17) first described this disease. Pathological features included different degrees of demyelination in the lesion area, axonal reservation, astrocytosis, and perivascular infiltration with a large number of mononuclear macrophages and lymphocytes. Most of the patients showed a single lesion with or without old lesions at the same site. Demyelinating pseudotumor progresses with an acute and severe onset phase that can later gradually stabilize with prolonged disease duration (7). Quick frozen sections are often misdiagnosed as astrocytoma because it is difficult to distinguish macrophages and swollen astrocytes. Therefore, Xia et al. (19) suggested that intraoperative frozen section analysis was inconclusive in the diagnosis of intracranial demyelinating pseudotumor. Immunohistochemical techniques should be applied when the case is suspicious or uncertain (3). Immunohistochemical examinations of myelin and CD68 staining can be used to confirm the diagnosis of intracranial demyelinating pseudotumor because the CD68 marker discriminates between macrophages and swollen astrocytes (3). The observation of demyelination and axonal preservation, combined with CD68 staining indicates intracranial demyelinating pseudotumor. In the current patient, CD68 staining was positive, there was extensive destruction of myelin with macrophage infiltration, and preserved axonal fibers. Therefore, the patient was diagnosed with intracranial demyelinating pseudotumor and treated with steroids. Based on the last follow-up, the patient had recovered well.

Intracranial demyelinating pseudotumor often presents as a single-subcortical lesion with an unclear boundary mass without obvious surrounding edema and significant disproportion between the mass effect and lesion volume. In most cases, CT imaging shows low or modest densities, while MRI shows abnormal long T1 and T2 signals. In the literature, the majority of demyelinating pseudotumor cases presented with non-closed ring enhancements (also called crescent sign) on CT and (or) MRI scans (11,12). However, intracranial tumors often lack this signal (13,18). Ma et al. (10) suggested that non-closed ring enhancements observed in demyelinating pseudotumor tend to be perpendicular to the surface of the lateral ventricle. In the differential diagnosis of astrocytoma, single demyelinating pseudotumor should always be ruled out.

Low grade astrocytoma usually presents on CT and MRI scans without any enhancement or is only slightly enhanced, however, demyelinating pseudotumor presents with significant enhancement. In addition, complete ring or lace-like enhancements with uneven thickness are found in the acute phase of high grade astrocytoma. Unlike astrocytoma, demyelinating pseudotumor is often associated with light edema and less mass effect.

Demyelinating pseudotumor also should be differentiated from other demyelinating lesion such as multiple sclerosis. Multiple sclerosis often shows multiple and symmetrically located lesions in a periventricular distribution (2). It should be noted, that the patient in our case presented with a cord-like enhancement and non-typical MRI features for demyelinating pseudotumor.

In the clinic, demyelinating pseudotumors are often sensitive to hormone therapy with decreased recurrence after treatment. Kepes described 31 cases of demyelinating pseudotumor treated with hormone therapy, where 28 out of 31 cases showed no recurrence at follow-up 9 months to 12-year post-treatment (8). This finding suggests that the prognosis in the majority of demyelinating pseudotumor patients is very good. Thus, if we can timely and correctly diagnose demyelinating pseudotumor, patients will receive treatment earlier and avoid blind surgery, radiotherapy and chemotherapy that could cause unnecessary physical damage and economic losses.

Although the case was misdiagnosed, we were able to outline how to differentially diagnose demyelinating pseudotumor and astrocytoma based on our case and the literature (1,9,15,16,21).

## ■ CONCLUSION

- 1) Cerebral demyelinating pseudotumor is often found with an acute onset and short duration, while astrocytomas show a slower progression. Consequently, the presented case had a medical history of only one week;
- 2) Patients with demyelinating pseudotumor are often young and middle-aged. Our patient was a 40-year old patient;
- 3) Unlike astrocytoma, demyelinating pseudotumor is often accompanied with light edema and less mass effect, which can be observed on CT and MRI scans;
- 4) Lesion enhancements of demyelinating pseudotumor tend to be perpendicular to the surface of the lateral ventricle;
- 5) Demyelinating pseudotumors are often sensitive to hormone therapy with less recurrence risk after treatment. All the above mentioned features should be kept in mind and should be used when a demyelinating pseudotumor cannot be excluded from the differential diagnosis.

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