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CASE REPORT

Open rings of demyelination – a rare case of tumefactive multiple sclerosis

Parameswaran K. [1], Nemani Sai Manasa [1], Nirmala Devi Chandrasekaran [1], Aswin C. [1], Kumar J.S. [2], Dev Sudersan Venkatesan [3], Dhivakar Ramalingam [3]

[1] Department of Internal Medicine, SRM Medical College Hospital and Research Center, SRM Institute of Science and Technology, Kattankulathur, Chengalpattu District, Tamil Nadu, India
[2] Department of General Medicine, SRM Medical College Hospital and Research Center, SRM Institute of Science and Technology, Kattankulathur, Chengalpattu District, Tamil Nadu, India
[3] Department of Pharmacy Practice, SRM College of Pharmacy, SRM Institute of Science and Technology, Kattankulathur, Chengalpattu District, Tamil Nadu, India

ABSTRACT

Introduction and aim. Tumefactive multiple sclerosis (MS), a rare and atypical subtype of MS, presents with large demyelinating lesions that can mimic acute stroke, leading to diagnostic uncertainty. Stroke-like symptoms in such cases require a thorough neuroimaging. We describe the case of a 35-year-old woman who presented with acute onset of right-sided hemiparesis and hemisensory loss, along with facial weakness of the left upper motor neuron facial weakness and focal seizures. Initially suspected to be a cerebrovascular event, the condition was later diagnosed as tumefactive multiple sclerosis.

Description of the case. Comprehensive neurological assessment with neuroimaging and magnetic resonance peduncle, and bilateral cerebral hemispheres, raising suspicion of a demyelinating process. A differential diagnosis, including neoplastic, infectious, and inflammatory conditions was carefully evaluated before confirming tumefactive MS. The patient's stroke-like deficits improved significantly with high-dose intravenous methylprednisolone therapy. Follow-up imaging demonstrated the resolution of the enhancing lesions, strengthening the diagnosis. The dramatic response to steroids and the absence of progressive deterioration helped differentiate tumefactive MS from gliomas or infectious abscesses.

Conclusion. This case highlights the importance of considering tumefactive MS in acute neurological deficits with ring-enhancing lesions. Advanced imaging techniques are crucial for accurate differentiation that allows for timely and appropriate treatment.

Keywords. case report, demyelination, neuroimaging, open ring-enhancing lesion, stroke mimic, tumefactive MS

Introduction

Ring-enhancing lesions are frequently encountered in neuroimaging and are typically present as isointense or hypointense on noncontrast computed tomography scans. On contrast imaging, a ring or plate of enhancement occurs within the area of hypointensity or isointensity, which may be encircled by vasogenic edema. The differential considerations for ring-enhancing lesions are extensive, including tuberculomas, abscesses, metastases, subacute infarction, and radiation necrosis.[1] An open ring or crescent-shaped pattern of white matter enhancement is rare and often suggestive of a demyelinating or neoplastic lesion, posing a significant diagnostic challenge for healthcare professionals.

Corresponding author: Nirmala Devi Chandrasekaran, e-mail: nirmalac@srmist.edu.in

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Tumefactive multiple sclerosis (TMS) is a rare form of multiple sclerosis, with a reported prevalence ranging from 1.4% to 8.2% among patients with multiple sclerosis. The incidence of this rare subtype is approximately 0.3 per 100,000 individuals.[2] The initial treatment for tumefactive demyelinating lesions involves intravenous corticosteroids for 3 to 5 days, followed by a gradual reduction in oral corticosteroid dosage. If steroids are contraindicated or ineffective, alternative therapies, including plasmapheresis (PLEX), cyclophosphamide, and rituximab, are considered.[3] The prognosis of tumefactive MS depends on a range of factors, such as the size and areas of the lesions, as well as the individual's response to treatment.

Aim

We present a report on a woman who presented with symptoms resembling a cerebrovascular accident. Further investigations confirmed a diagnosis of tumefactive multiple sclerosis, and treatment with corticosteroid therapy was initiated, leading to total remission.

Table 1. Laboratory parameters

Test	Parameter	Result	Normal values
Complete hemogram	Hemoglobin	12.3 g/dL	12.1–15.1 g/dL
	White blood cell	5500 cells/µL	4,500–11,000 cells/µL
	Platelet count	265000 cells/µL	150,000–450,000 cells/µL
	Hematocrit	42%	36.1–44.3%
	Mean corpuscular volume	86 fL	80–100 fL
Liver chemical Panel	Alanine aminotransferase	32 U/L	7–56 U/L
	Aspartate aminotransferase	23 U/L	10–40 U/L
	Alkaline phosphatase	52 U/L	44–147 U/L
	Total bilirubin	0.8 mg/dL	0.1–1.2 mg/dL
	Albumin	3.7 g/dL	3.5–5.0 g/dL
	Total protein	6 g/dL	6.3–7.9 g/dL
Renal function analysis	Blood urea nitrogen	18 mg/dL	7–20 mg/dL
	Serum creatinine	0.6 mg/dL	0.59–1.04 mg/dL
	Glomerular filtration rate	124 mL/min/1.73 m^2	≥90 mL/min/1.73 m^2
Random blood glucose	Glucose	88 mg/dL	70–140 mg/dL
Lipid profile	Total Cholesterol	145 mg/dL	<200 mg/dL
	Low-density lipoprotein	87 mg/dL	<100 mg/dL
	High-density lipoprotein	42 mg/dL	>50 mg/dL
	Triglycerides	64 mg/dL	<150 mg/dL
Vasculitis profile	Extractable nuclear antigen	Negative	

Description of the case

A 35-year-old woman was admitted with seizures involving the right upper and lower limbs, which occurred upon waking in the morning. She experienced 1–2 episodes daily over the past 6 days, accompanied by progressive weakness in the right upper and lower limbs, involving both proximal and distal muscle groups. This was associated with sensory loss on the right side and deviation of the angle of the mouth to the right. There was no history of recent respiratory infections, diarrheal disease, travel, dog bites, or toxin exposure.

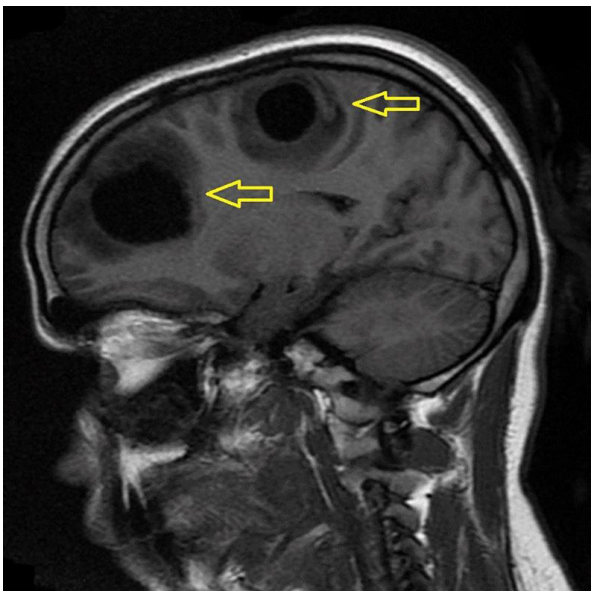

Fig. 1. Sagittal section of T_1 weighted MRI brain plain illustrating well-circumscribed `T_1 hypointense lesions with perilesional edema in the frontal and parietal lobe of the white matter of the cerebral hemisphere

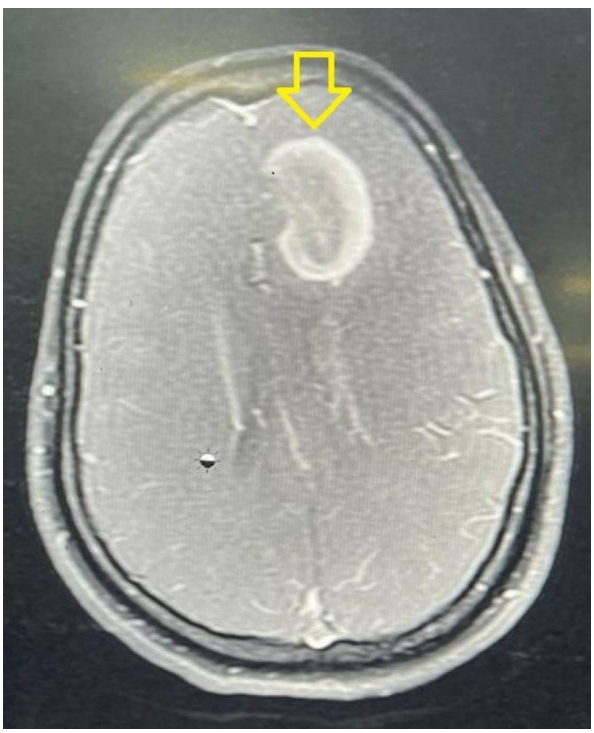

Fig. 2. Magnetic resonance brain imaging with contrast was done, which revealed these ring-enhancing lesions to be broken, classically opening towards the grey matter in the frontal lobe; termed as an open ring-enhancing lesion

On examination, the patient was conscious and well-oriented, with normal vital signs. Carotid pulses were equally palpable. Higher mental functions were in-

tact. Notably, there was a deviation of the angle of the mouth to the right, which clinically suggested left upper motor neuron facial palsy. The assessment of motor function revealed a power of 2/5 in both the right upper and lower limbs, with spasticity and brisk reflexes. Sensory examination showed diminished sensation on the right, particularly for pain and temperature. The clinical presentation resembled a stroke, affecting structures such as the left corticospinal tract, left spinothalamic tract, and left medial lemniscus (Fig. 1).

Complete hemogram, liver function analysis, renal function analysis, random blood glucose levels, lipid profile, and vasculitis profile were all within normal limits (Table 1).

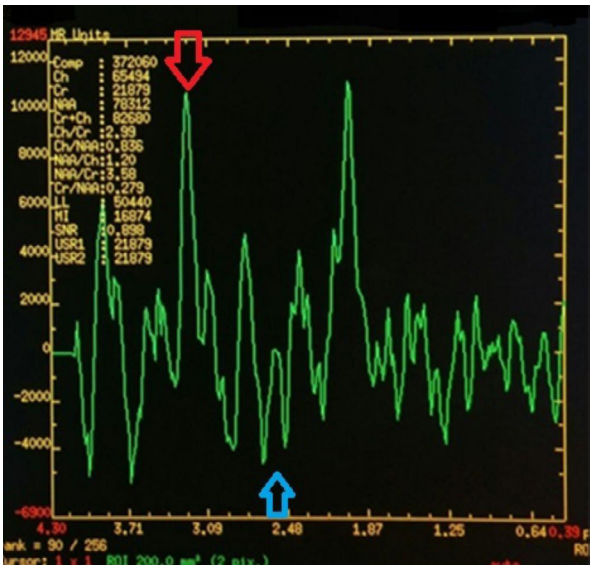

Fig. 3. Magnetic resonance spectroscopy further revealed to have a demyelinating pattern which indicates increased choline-containing peaks (red arrow) and decreased N-acetyl aspartate activity (blue arrow)

Discussion

Central nervous system demyelinating disorders are relatively common; among them, MS is the most prevalent. MS is an autoimmune disease characterized by dissemination in both time and space. Tumefactive multiple sclerosis is an uncommon and atypical form of MS, often presenting initially as a clinically isolated syndrome, which may later progress to develop MS several years afterward.[4] Tumefactive MS can onset at any age, with peak incidence occurring between 20 and 40 years, and it is more prevalent in women.[5] Tumefactive MS can present as an initial presentation of the disease or develop during the course of MS. It can also occur following the discontinuation of fingolimod therapy in MS patients.[6] MS lesions are typically small, numerous, and ovoid. In comparison, tumefactive MS lesions are larger (>2 cm) with focal areas of demyelination, which may or may not exhibit associated edema or mass effect.[7] Although the exact pathology of tumefactive TMS is not fully understood, a large multicenter retrospective study of tumefactive TMS patients found that approximately 10% had concomitant autoimmune disorders.[8]

The clinical presentation of tumefactive MS is often multifocal and varies with the specific areas of central nervous system involvement. The supratentorial areas, predominantly the frontal and parietal lobes, are most commonly involved.[9,10] Motor disturbances are the most common symptoms, followed by sensory, visual, and cognitive disturbances. The atypical symptoms reported in tumefactive MS can, at least in part, be explained by cortical grey matter involvement or by the effects of surrounding edema.[6]

The radiological approach is considered the gold standard for diagnosing tumefactive MS. On magnetic resonance imaging, tumefactive MS lesions typically appear hyperintense on T_2-weighted images and relatively hypointense on T_1-weighted and fluid-attenuated inversion recovery sequences.[11] Ring enhancement with gadolinium is a hallmark of tumefactive demyelinating lesions, observed in 95 to 100% of cases. This ring enhancement may present in various patterns, including open or closed rings, diffuse, homogeneous, punctate, or concentric configurations. The presence of an open-ring pattern, with the open segment connecting to the gray matter of the cortex or deep nuclei, is highly suggestive of a demyelinating disorder (Fig. 2). The enhancing component of the ring is considered an advancing front of demyelination, while the non-enhancing core at the center signifies a more chronic inflammatory process.[9]

Magnetic resonance spectroscopy demonstrated high levels of choline-containing compounds (Fig. 3), indicative of inflammation and the release of choline-containing membrane lipids resulting from active myelin and cell membrane breakdown. Concurrently, reduced levels of N-acetyl aspartate are observed, reflecting neuronal destruction and axonal damage. When combined with conventional magnetic resonance imaging, the Cho/NAA ratio has been demonstrated to enhance diagnostic accuracy.[10]

Histopathological features of tumefactive demyelination are consistent with active inflammatory demyelinating disease and typically include demyelinated areas with relative axonal preservation, infiltration of foamy macrophages, reactive astrocytosis, and perivascular lymphocytic infiltration.[10] Brain biopsy should be reserved for cases where a definitive diagnosis remains unclear and uncertain despite a comprehensive diagnostic approach using multiple modalities.[12]

High-dose corticosteroid therapy is considered the primary therapeutic approach for patients with tumefactive multiple sclerosis; however, the response to corticosteroid therapy is not universal, as approximately 86% of patients exhibit clinical improvement.[8] The standard reg-

imen involves administering a high dose of methylprednisolone (e.g., 1 gram per day for 3 to 5 days) followed by a gradual taper with oral corticosteroids.[3] If steroids are contraindicated or ineffective, PLEX is regarded as the alternative second-line therapy.[6,9] In patients who are refractory to intravenous corticosteroids or plasma exchange, cyclophosphamide may be an effective alternative due to its immunomodulatory properties.[13] The role of disease-modifying therapies in patients with tumefactive MS remains unclear. Interferon-beta and glatiramer acetate are typically preferred; however, these treatments are generally not initiated until a second clinical relapse occurs.[8]

The long-term prognosis of tumefactive MS is variable. Approximately one-third of patients will not experience a subsequent demyelinating episode. Among those who do, two-thirds will exhibit a relapsing-remitting course.[6,8]

In our case, a positive response was observed with pulse methylprednisolone therapy, further supporting the definitive diagnosis of tumefactive MS, as evidenced by the presence of open ring-enhancing lesions.

Conclusion

Tumefactive MS, an uncommon variant of multiple sclerosis, can be difficult to diagnose due to its resemblance to the radiological and clinical features of an abscess, tumor, or stroke. Our findings highlight the importance of including demyelination in the differential diagnosis of ring-enhancing lesions, particularly with a positive response to corticosteroids, thereby minimizing the necessity for biopsy and broadening the spectrum of potential tumefactive MS presentations.

Declarations

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Author contributions

Conceptualization, P.K. and N.S.M.; Methodology, P.K. and A.C.; Software, N.D.C.; Validation, N.D.C. and D.S.V.; Formal Analysis, D.S.V., K.J.S. and D.R.; Investigation, P.K. and D.S.V.; Resources, P.K.; Data Curation, N.D.C.; Writing – Original Draft Preparation, N.S.M, P.K. and N.D.C.; Writing – Review & Editing, N.D.C., D.S.V. and D.R.; Visualization, N.D.C. and A.C.; Supervision, P.K. and K.J.S.; Project Administration, P.K.

Conflicts of interest

The authors declare that they have no conflicts of interest.

Data availability

All data generated or analyzed during this study are included in this published article. Additional data are available from the corresponding author upon reasonable request.

Ethics approval

Ethical approval was not required for this case report. Written informed consent was obtained from the patient for participation in this study.

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