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Q5 Q1 Tumefactive Demyelination Appearing as Multiple Cystic Brain Lesions

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Key words

Tumefactive demyelinating lesions

Tumefactive multiple sclerosis

Abbreviations and Acronyms

MRI: Magnetic resonance imaging TDL: Tumefactive demyelinating lesion

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CLINICAL BACKGROUND

A 35-year-old female presented to the hospital after 2 right-hand focal sensory seizures, the second of which progressed into a generalized tonic-clonic seizure. On examination she had no motor or sensory deficits. Magnetic resonance imaging (MRI) revealed 5 supratentorial lesions, the 2 largest being cystic—30 \times 30 \times 32 mm within the left superolateral postcentral gyrus and $7 \times 9 \times 6$ mm within the right lateral postcentral gyrus. All lesions were T1 hypointense and T2 hyperintense with mild surrounding vasogenic edema (Figure 1A and C). The 2 cystic lesions demonstrated peripheral contrast enhancement, elevated signal on fluidattenuated inversion recovery, and no diffusion restriction (see Figure 1B and D). To access the prominent left superolateral postcentral gyrus lesion, we dissected down the Rolandic fissure, retracted the Rolandic vein, and made a small corticotomy into the postcentral gyrus. Transparent yellow cystic fluid was aspirated, and samples from the dissected pseudocapsule were for histopathology. Histology revealed gliotic

Tumefactive demyelinating lesions (TDLs) are a rare sequelae of idiopathic inflammatory demyelinating diseases of the central nervous system. Their propensity to mimic tumor and abscess poses a diagnostic challenge for the clinician. Our case depicts TDLs causing right-hand focal sensory seizures in an otherwise healthy 35-year-old female. The differential diagnosis of metastatic disease and infection were excluded on histology. Ensuing magnetic resonance imaging of the cord, in addition to cerebral spinal fluid analysis, supported the diagnosis of idiopathic inflammatory demyelinating diseases. This case highlights the need to consider the rare diagnosis of TDL when imaging shows cystic brain lesions in an otherwise healthy young adult.

demyelinated brain with prominent cystic changes, old hemorrhage, numerous macrophages, and perivascular T-cell cuffing. There was no histologic evidence of cysts, parasites, or malignancy. Ensuing MRI whole spine revealed focal cervical cord lesions, and lumbar puncture obtained cerebrospinal fluid demonstrated oligoclonal bands, distinguishing the diagnosis of tumefactive demyelinating lesions (TDLs). The patient's focal sensory seizures ceased post commencement of levetiracetam, and she was referred to a neurologist for methylprednisolone therapy.

DISCUSSION

Idiopathic inflammatory demyelinating diseases lesions are typically small (<2 cm) and ovoid. On MRI, these lesions are T₂ hyperintense and variably enhance.¹ Infrequently, patients have larger demyelinating lesions, often with edema and mass effect, which appear somewhat similar to central nervous system tumors. These lesions, known as TDLs, are commonly located in the cerebral hemispheric white matter, especially the frontal and parietal lobes.² TDLs are >2 cm in size on MRI and may have associated mass effect and edema, with larger lesions generally having more of both.² Unfortunately, there are no

pathognomonic features on MRI for TDL. However, certain features may point toward the diagnosis including the presence of a T2 hypointense ring around the lesion and presence of other idiopathic inflammatory demyelinating disease-typical white matter lesions.3-5 In addition, almost all TDLs will enhance with gadolinium in a variety of patterns.⁶ Opening enhancement pointing toward the gray matter is an important clue to the presence of demyelination, seen more commonly in TDL than tumors or infections.³ Central diffusion restriction can also be seen in lesions with an intense inflammatory core. and occasionally, diffusion restriction is seen in the periphery of the lesion.⁶

CONCLUSION

This case served as a diagnostic dilemma initially appearing as multiple cystic supratentorial lesions. Initial differentials of a malignant or infective origin were excluded with biopsy and histological analysis. Retrospectively, MRI appearance was suggestive of TDL not initially considered in the differential diagnosis. The additional findings of cervical cord lesions and oligoclonal bands in cerebrospinal fluid aided in confirming the diagnosis of TDL.

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CLINICAL IMAGES

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Figure 1. A 35-year-old female presented after 2 right-hand focal sensory seizures, the second of which progressed into a generalized tonic-clonic seizure. Examination was unremarkable. Magnetic resonance imaging revealed 5 supratentorial lesions. The 2 largest of these lesions appeared cystic—30 imes 30 imes 32 mm within the left superolateral postcentral gyrus and 7 \times 9 \times 6 mm within the right lateral postcentral gyrus (A-D). These were T1 hypointense (A) and T2 hyperintense with mild surrounding vasogenic edema (C). They demonstrated partial signal elevation on fluid-attenuated inversion recovery (C), modest contrast enhancement peripherally (B), and no diffusion restriction (D). Biopsy of the left superolateral postcentral gyrus lesion for histopathology revealed abnormal gliotic brain with cystic change and prominent astrocytosis (E), perivascular lymphocytic cuffing (F), no residual myelin (G), CD163+ macrophages consistent with demyelination (H), and numerous reactive astrocytes (I). The presumptive diagnosis of tumerfactive demyelinating lesions was supported by additional T2 hyperintense cervical cord lesions and oligoclonal bands in the cerebral spinal fluid.

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