

A potential pitfall in neuro-oncological surgery

Tumefactive multiple sclerosis

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Summary

Tumefactive lesions in multiple sclerosis (MS) can mimic brain tumours and, thus, may become a pitfall in neuro-oncological surgery. In the current case, an atypical closed-ring enhancing tumefactive MS lesion was mistaken for a malignant tumour. Ultrasound real-time imaging was advantageous when intraoperative management was adjusted in response to a lack of obvious tumour tissue and inconclusive histopathological results from fresh frozen sections. Measures to identify tumefactive MS, in order to avoid operating, are briefly discussed. These recommendations include specifically inquiring about MS clues in the patient's history as well as the integration of new neuroimaging techniques for preoperative diagnostics.

Key words: brain tumour; biopsy; multiple sclerosis; pitfall; tumefactive; ultrasound

Introduction

Tumefactive lesions represent a rare radiological manifestation of multiple sclerosis (MS) seen in only a few cases per one thousand patients [1]. Often, their pattern appears to be open ring-enhancing, with the incomplete portion abutting cortical gray matter or the basal ganglia [2]. As implied by the name, tumefactive MS can be challenging to diagnose correctly prior to obtaining tissue samples [3]. Mistaking tumefactive MS for a brain tumour is a pitfall that should not be underestimated [3], as in these cases microsurgical resection carries unnecessary risks for the patient.

We present a case of an unusual closed ring-enhancing appearance of tumefactive MS along with an outline of our patient management that accounted for this rare event.

Case report

A 53-year-old otherwise healthy right-handed woman presented with a 6-month history of morning headaches and nausea, as well as recent transient episodes of confusion and spatial disorientation. Furthermore, she reported that while teaching in school she repeatedly missed grasping items from the desk drawer with her left hand. She also noted some left-sided coordina-

tion difficulties and gait instability with a shift to the left side. We specifically inquired about similar or other neurological symptoms in the past, but there were none. During examination, she had slowed diadochokinesis, atactic heel-shin tests on the left and finger perimetric testing revealed left inferotemporal quadrant anopia, which was confirmed with Goldmann perimetry.

Magnetic resonance imaging (MRI) with contrast medium showed a 2.4 cm ring-enhancing lesion with extensive perifocal oedema in the right parietal lobe (fig. 1). An extracranial primary tumour was ruled out by thorax/abdomen/pelvis computed tomography, rendering a cerebral metastasis unlikely. With the expectation of a high-grade glioma as the top of our list of differential diagnoses, we obtained her consent for an open ultrasound-guided microsurgical resection via a minicraniotomy. Dexamethasone was not given until after surgery to prevent a false-negative biopsy result in the event of lymphoma.

Surgery was without complications; however, lesion site visualisation did not reveal any obvious tumour, although intraoperative ultrasound confirmed an intralésional location. Fresh frozen sections were not consistent with the suspected diagnosis, either. Therefore, the operation was limited to a biopsy, demonstrated on postoperative MRI (fig. 1, inset).

Definite histology revealed fragmented myelin sheaths with preserved axons accompanied by a massive infiltration of phagocytosing macrophages with lymphocytic infiltration, in agreement with the diagnosis of MS (fig. 2).

The neurology service followed up on patient. One day after finishing postoperative dexamethasone tapering, a lumbar puncture revealed 38 mononuclear cells/ml, intrathecal IgG synthesis and no oligoclonal bands. No further demyelinating lesions were found on complete imaging work-up of the neuraxis. Treatment with high-dose glucocorticoids was then initiated and resulted in substantial symptom improvement. Neuropsychological assessment 1 week after surgery, however, still demonstrated a severe visual-spatial processing disorder, a persistent visual field defect and left kinetic apraxia.

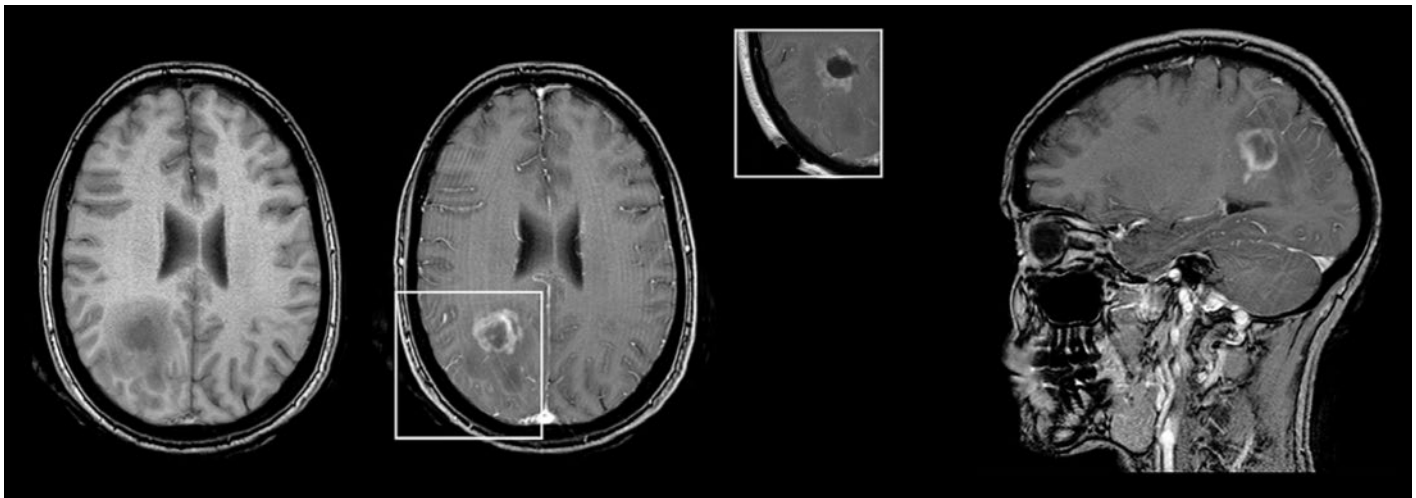


Figure 1: T1 magnetic resonance tomography with contrast showing a 2.4 cm closed ring-enhancing lesion with extensive perifocal edema in the right parietal lobe. The inset shows the postoperative ultrasound-guided open biopsy site.

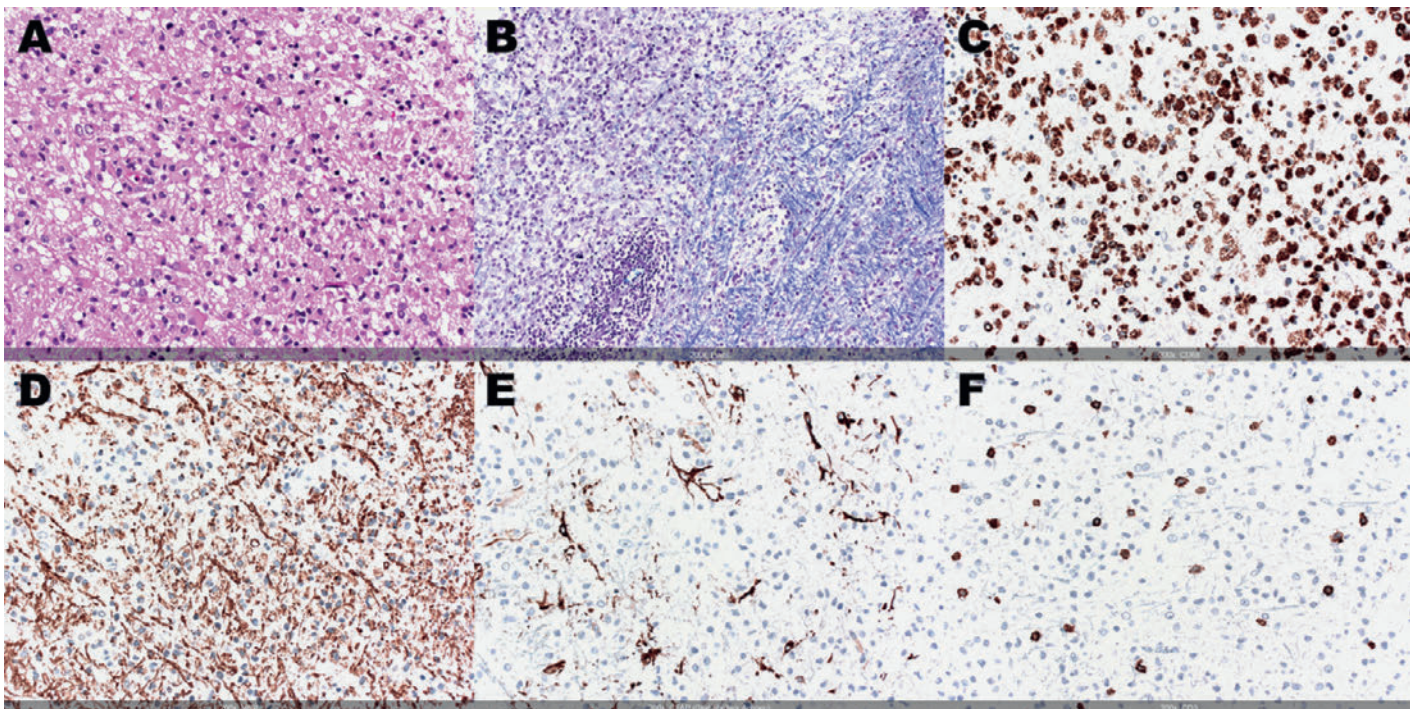


Figure 2: A. Haematoxylin-eosin (HE) staining revealing an abundance of eosinophilic macrophages and few glial cells with a paucity of axon remnants without myelin sheaths. B. Luxol fast blue staining showing areas with (right side) and without myelin sheaths (left side) together with macrophages. C. CD68 immunohistochemical staining showing dense infiltrations of macrophages that contain myelin debris (cytoplasmic 'gaps'). D. Myelin basic protein (MBP) immunohistochemical staining showing fragmented myelin sheaths imbedded in a mass of myelin-phagocytosing macrophages. E. Glial fibrillary acidic protein (GFAP) immunohistochemical staining showing predominantly astroglial cells in the midst of (nonstaining) macrophages. F. CD3 immunohistochemical staining showing concomitant reactive T-lymphocytes infiltration.

Discussion

In this case report, we present a pitfall in operating on a closed ring-enhancing lesion: tumefactive MS. Unlike in the presented MRI, tumefactive MS usually is reported to be reminiscent of a crescent [2] with only mild perifocal oedema [1]. This rendered preoperative differential diagnosis difficult and, in retrospect, it is

therefore understandable that we decided for an "open" approach rather than a stereotactic biopsy. The latter carries the risks of surgical complications; these may be obviated when a microsurgical full-resection is highly likely to follow, such as when the lesion in question is a glioma or metastasis. Whenever the pre-

operative diagnosis is uncertain, however, we would recommend a biopsy over an upfront open resection. When tumour tissue is not visualised under the operative microscope and fresh frozen sections are not helpful in establishing the diagnosis, verification of the correct intralesional location by ultrasound can be reassuring when terminating the operation. To avoid operating on tumefactive MS, it is important to bear in mind the importance of taking a thorough patient history. In Lucchinetti's review of 168 biopsied tumefactive MS cases [1], as many as 29% had prior neurological episodes. Also, adjunctive new neuroimaging techniques may aid in establishing the correct diagnosis [3]. For instance, magnetic resonance spectroscopy reveals a higher n-acetyl aspartate/creatinine ratio, and dynamic contrast-enhanced T2-weighted MRI shows a lower regional cerebral blood volume value in demyelinating lesions as compared with neoplasms. Also, transfer magnetisation MRI showing decreased values might represent a useful diagnostic means to pick up tumefactive MS when conventional MRI diagnosis is thought to be misleading [3].

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Conclusion

Tumefactive MS is rare and can be misleading in preoperative decision making for neuro-oncological surgery, as it can atypically present as a closed ring-enhancing lesion. Previous neurological episodes should be sought in the patient's history as these may give clues to a MS diagnosis. Adjunctive, new neuroimaging techniques may also be considered prior to invasive diagnostic measures. The utility of intraoperative ultrasound should not be discounted when there is doubt about correct lesion location because of its advantage of real-time imaging.

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Disclosure statement

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Informed consent

The patient presented in this case report provided written consent for publication.

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