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Case Report

Glioblastoma multiforme presenting with an open ring pattern of enhancement on MR imaging

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Abstract

Background: Intracerebral ring enhancing lesions can be the presentation of a variety of pathologies, including neoplasia, inflammation, and autoimmune demyelination. Use of a precise diagnostic algorithm is imperative in correctly treating these lesions and minimizing potential adverse treatment effects.

Case Description: A 55-year-old patient presented to the hospital with complaints of a post-concussive syndrome and a non-focal neurologic exam. Imaging revealed a lesion with an open ring enhancement pattern, minimal surrounding vasogenic edema, and minimal mass effect. Given the minimal mass effect, small size of the lesion, and nonfocal neurological exam, we elected to pursue a comprehensive noninvasive neurologic workup because our differential ranged from inflammatory/infectious to neoplasm. Over the next 8 weeks, the patient's condition worsened, and repeat imaging showed marked enlargement of the lesion with a now closed ring pattern of enhancement with satellite lesions and a magnetic resonance (MR) spectroscopy and perfusion signature suggestive of neoplasm. The patient was taken to surgery for biopsy and debulking of the lesion. Surgical neuropathology examination revealed glioblastoma multiforme.

Conclusion: The unique open ring enhancement pattern of this lesion on initial imaging is highly specific for a demyelinating process, however, high-grade glial neoplasms can also present with complex and irregular ring enhancement including an open ring sign. Therefore, other imaging modalities should be used, and close follow-up is warranted when the open ring sign is encountered.

Key Words: Glioblastoma, incomplete peripheral rim enhancement, open-ring sign, radiographic image enhancement, tumefactive demyelination, tumefactive demyelinating lesion

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INTRODUCTION

Intracerebral ring enhancing lesions can be a presentation of neoplasia (including metastases, lymphoma, and gliomas), infections, or autoimmune demyelination mimicking neoplasia (tumefactive demyelination), thus posing a diagnostic and management dilemma.[1] While neoplastic lesions require surgical intervention for diagnosis and sometimes resection, tumefactive demyelination responds well to corticosteroids without the need for surgery. However, use of corticosteroids can mask lymphomas and should be avoided when a tissue diagnosis is required. Incomplete peripheral enhancement, also referred to as an "open-ring" enhancement is thought to be highly specific for tumefactive demyelinating lesions (TDLs), yet can also represent malignancy – we present such a case and underscore the need for close follow-up of patients presenting with open-ring lesions.

CASE HISTORY

A 55-year-old man presented to an outside institution for treatment of a concussion which he sustained after a fall from a ladder while working in his yard. Initial neurologic examination and magnetic resonance imaging (MRI) of the brain were normal. The patient continued to have concussive symptoms and headache 4 months later. Neurologic exam was still normal, however, repeat MRI revealed a right posterior temporal cystic lesion measuring 1.5 × 1.5 cm with incomplete peripheral enhancement characteristic of an open ring sign [Figure 1a], with mild surrounding vasogenic edema [Figure 1b], minimal mass effect, no midline shift, and no restriction on diffusion. Because the differential diagnosis included infectious/inflammatory process, cavernoma, and neoplasm, the decision was made to follow the lesion with a comprehensive, noninvasive neurologic workup. Two months later, the patient began having acute, intermittent confusional episodes, culminating in a witnessed generalized seizure. Repeat MRI showed a 5.3 × 4.2 cm lesion with significant mass effect and surrounding vasogenic edema [Figure 2a]. The mass had a more defined central cystic component, did not show restriction on diffusion and a closed ring pattern of peripheral enhancement, as well as satellite lesions along its posterior aspect [Figure 2b]. MR spectroscopy was performed and showed an increased choline-to-creatine ratio with a decreased NAA signature suggestive of a neoplasm [Figure 3]. MR perfusion showed increased relative blood volume compared to contralateral tissue, strongly suggesting neoplasia [Figure 4].

The patient was taken to surgery for a right-sided temporal craniotomy for resection of the mass. Intraoperatively, the lesion was noted to be hemorrhagic with areas of necrosis and thrombosed vessels suggestive of a malignant glial neoplasm. Surgical neuropathology examination showed the lesion to be an infiltrating

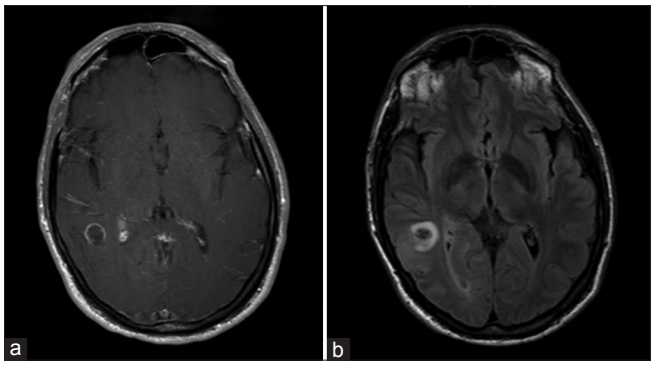

Figure 1: Axial cut MR showing right, posterior temporal cystic lesion measuring 1.5 × 1.5 cm with incomplete peripheral enhancement on T1 contrast-enhanced image (a) with mild surrounding vasogenic edema on T2 FLAIR (b)

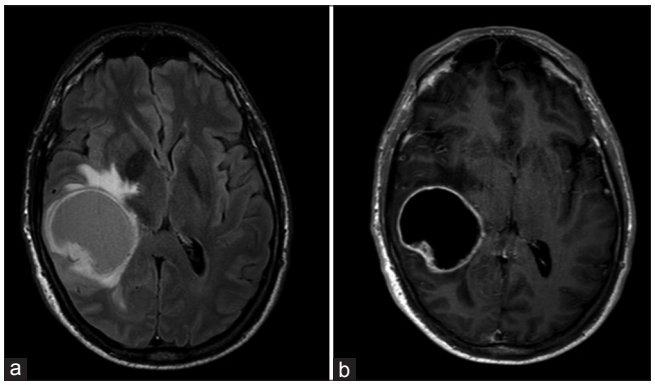

Figure 2: Axial cut MR showing increased size of lesion, measuring 5.3 × 4.2 cm with significant mass effect and surrounding vasogenic edema on T2 FLAIR (a) and a now closed ring pattern of peripheral enhancement on T1 contrast-enhanced image (b). Central cystic component now well-defined and satellite lesions are visible along posterior aspect

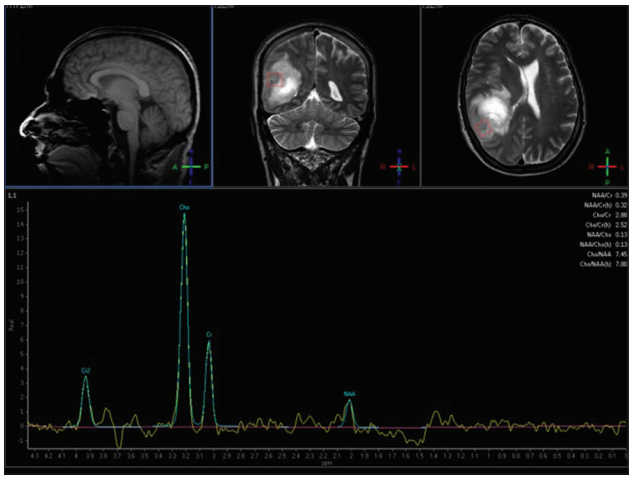

Figure 3: Sagittal, coronal and axial cuts of lesion on T2-weighted MRI above, below MR spectroscopy shows an increased choline to creatine ratio with a decreased NAA signature

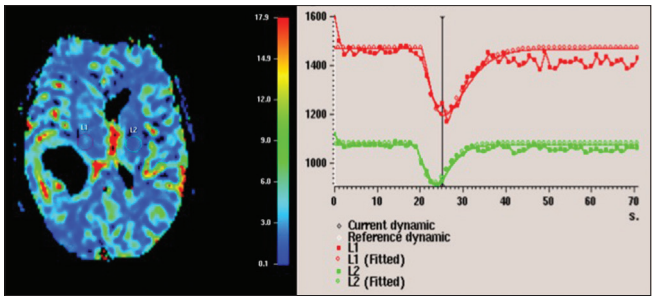

Figure 4: Axial cut MR perfusion scan shows increased relative blood volume compared to contralateral tissue

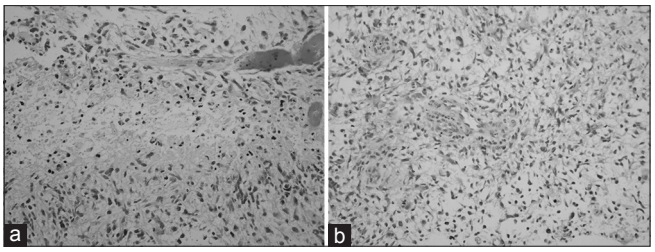

Figure 5: H and E sections from the neoplasm show pleomorphic process-forming astrocytic cells around an area of necrosis (pseudopalisading necrosis) (a). Higher power (b) shows the atypical, pleomorphic astrocytic cells characteristic of high-grade astrocytic tumors

glial neoplasm with a fibrillary background. Tumor cells had elongated, pleomorphic, vesicular nuclei with scant cytoplasm [Figure 5a and b]. Psuedopallisading, necrosis, vascular proliferation, and abundant cellular mitoses and apoptosis were present, confirming the diagnosis of gliobastoma multiforme.

The patient began treatment with standard radiation and temozolomide and was well at the last follow-up 3 months after his diagnosis.

DISCUSSION

Ring-enhancing lesions are usually subcortical or deep. When presenting with associated vasogenic edema and mass effect, these lesions usually favor primary central nervous system neoplasm or abscess. However, demyelinating lesions seen in multiple sclerosis or tumefactive demyelination during the "active phase" of inflammation can also enhance, though the enhancement pattern of these lesions can be faint, with minimal perilesional vasogenic edema, and the enhancing rim may be incomplete.[1,3] The "open-ring" sign has been presented in many case studies in which lesions demonstrating incomplete, peripheral rim enhancement have been proven to be TDLs upon histological examination and/or resolution after steroid administration.[4,6,8,9,13,14] Unfortunately, some of these cases were preemptively treated as high-grade gliomas, and the patients underwent resection surgery only to be diagnosed with atypical demyelinating disease on histopathological examination.[8,10,11] The risks and potential comorbidities resulting from such misdiagnosis and treatment merit serious consideration when managing lesions exhibiting an open-ring enhancement pattern. Masdeu *et al*. reviewed imaging from 32 reported pathology-proven TDL cases showing complete or incomplete ring-enhancement and compared them to the same number of cases of neoplasm and brain abscess.[7] They reported a specificity of 84.4–93.8% of the "open-ring" sign for TDL diagnosis, with a likelihood ratio of 5.2 for demyelination versus neoplasm for a lesion with open-ring enhancement.[7] Although 70% of the reported TDL cases presented exhibited incomplete peripheral enhancement, the authors highlight the increased incidence of neoplasia as compared to atypical demyelination to keep neoplasm as a possible diagnosis.

This case is a rare example of an open-ring enhancing lesion which evolved from initial presentation to a complete ring-enhancing lesion, which was later confirmed to be glioblastoma multiforme. When trying to identify MR and CT features that distinguish TDLs from glioma or lymphoma, Kim *et al*. found 4 out of the 13 tumor patients exhibiting focal rim enhancement to have incomplete rings on MR.[5] They were able to distinguish these patients from TDL with corresponding CT grading of the enhancing portions for each pathology, but underscored that incomplete ring enhancement combined with lack of mass effect alone significantly suggests atypical demyelination.[5] In this case, there was no head CT done concurrently with the MR demonstrating open-ring enhancement to correlate. Another study investigating MR findings of primary CNS lymphoma patients reported 2 out of 26 studied cases demonstrating open-ring enhancement.[15] They note thick and non-uniform quality of the rings when compared to the primarily thin and uniform open-ring sign in TDLs.[15] Nonetheless, the present case shows a thin, uniform pattern of incomplete rim enhancement [Figure 1].

When presented with open-ring enhancing lesions, there is necessity to correlate with other features suggestive of TDL to rule out neoplasm. Such features include T2-weighted iso- and hyperintensity of enhanced regions, absence of mass effect, low relative perfusion, absence of cortical involvement, and CT hypoattenuation of MR enhanced regions.[5] MR imaging of TDLs may also show necrosis and cystic degeneration.[2] In this case, MR spectroscopy demonstrated the characteristic glioma spectrum consisting of elevated choline with suppressed levels of NAA, however, these can also be mimicked by TDLs.[12] The presence of other lesions typical of demyelination or oligoclonal bands in the cerebrospinal fluid are also suggestive of demyelination rather than neoplasia, but not absolute and were absent in our case. Patients with features suggestive of TDL may be

managed acutely with a short course of high dose steroids and if a good clinical and radiographic response was observed a diagnosis of a demyelinating process would be supported.[6] However, because all of these features can be seen in neoplasia, close follow up is required.

CONCLUSION

Open-ring pattern of enhancement is reported to be highly specific for demyelinating lesions, rather than neoplasia. However, high grade glial neoplasms can also present with complex and irregular ring enhancement including an open ring sign. Therefore, other imaging modalities should be used and close follow-up is warranted when the open ring sign is encountered.

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Conflicts of interest
There are no conflicts of interest.

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