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Case Report

Glioblastoma multiforme that unusually present with radiographic dural tails: Questioning the diagnostic paradigm with a rare case report[☆]

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ABSTRACT

Glioblastoma multiforme (GBM) is both the most common as well as one of the most aggressive primary intracerebral tumors. It classically presents on magnetic resonance imaging as a heterogeneous ring-enhancing lesion in the brain parenchyma with central necrosis. This type of neoplasm can also rarely present, however, as a mass with meningeal attachment and radiographic evidence of a dural tail, which was until recently thought to be specific to meningiomas. Here we present a case of a central nervous system neoplasm that on imaging was initially suggestive of meningioma based on its presence of a dural tail. Final pathology, however, revealed desmoplastic GBM. It is, therefore, important to include GBM on the differential diagnosis of a patient presenting with a dural-based lesion on imaging, especially since the overall survival rate of GBM is much worse than that of a suspected meningioma.

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Introduction

Glioblastoma multiforme (GBM) is the most common and one of the most aggressive primary intracerebral tumors, with a yearly incidence of about 32 per 1,000,000 population [1]. GBM has been used to describe grade IV astrocytic tumors with great heterogeneity in tissue histology and imaging [2,3]. On magnetic resonance imaging (MRI), glioblastomas typically have irregular enhancing margins with a central heterogeneous signal and are surrounded by vasogenic edema. This heterogeneous signal represents a necrotic core, which is usually secondary to the coexistence of tumor, necrosis, and hemorrhage.

What is not commonly found on MRI imaging, however, is enhancement of the leptomeninges to create a “dural tail” [4]. Here we report an interesting and rare finding of primary extracerebral meningeal GBM presenting with a distinct radiographic finding of a dural tail.

Case

A 63-year-old male with a past medical history of developmental delay, diabetes, hyperlipidemia, and peripheral vascular disease presented to the emergency room with a 2-week

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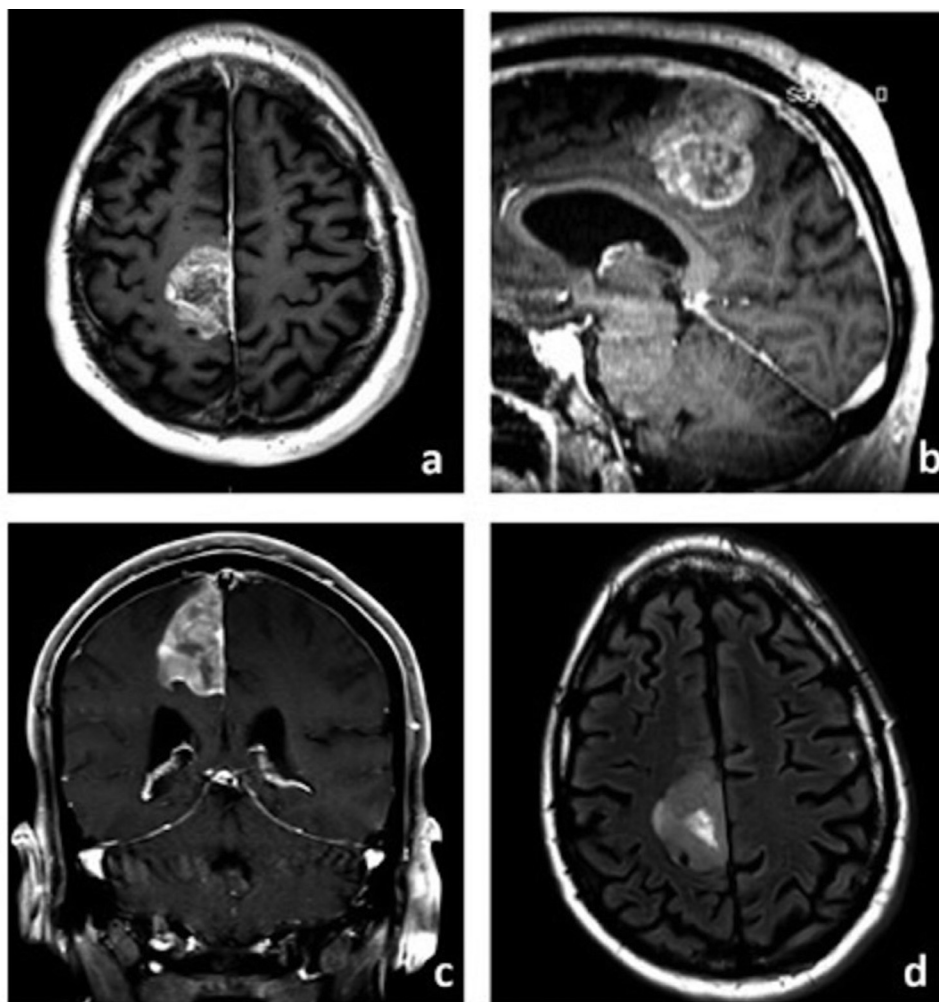


Fig. 1 – Preoperative MRI. (a) Axial T1 MRI after administration of gadolinium contrast demonstrating a right frontal-parietal lesion with heterogeneous enhancement. (b) Sagittal T1 with contrast. (c) Coronal T1 with contrast. The broad dural attachment to the falx is well visualized. (d) Axial T2 fluid attenuated inversion recovery (FLAIR) sequence demonstrates a small amount of surrounding edema.

history of falls and new-onset seizures. The seizure episodes consisted of involuntary movements of his left arm and tonic contraction of his neck lasting several minutes. Physical exam was significant for 4/5 left hand weakness. A computed tomography (CT) scan of the head revealed a right frontal mass with increased attenuation involving the parafalcine frontal lobe with surrounding hypodensity suggestive of vasogenic edema. MRI of the brain with gadolinium confirmed a large, heterogeneously enhancing mass with evidence of central necrosis. There appeared to be a broad dural attachment with a “dural tail” feature. The diagnosis was initially felt to be most consistent with a dural-based neoplasm, such as a meningioma (Fig. 1a-d).

The patient was admitted and, because on the progression of his symptoms, underwent a craniotomy for surgical resection. The mass was noted to have a gray, firm consistency, which extended directly into the overlying dura. There was no clear demarcation between the tumor and the underlying brain parenchyma. Frozen section was concerning for a high-

grade parenchymal tumor. After surgery, he was admitted to the hospital for routine monitoring and recovery. His hospital course was unremarkable, and he was discharged on post-operative day 5.

Final pathology revealed a WHO grade IV astrocytoma with meningeal invasion. Tumor cells were typical of glioblastoma, consisting of primitive astrocytic forms growing in sheets with frequent necrosis, endothelial proliferation, and mitosis. Concurrently, there were columns of tumor cells infiltrating areas of extensive desmoplastic reaction (Fig. 2a and b). The tumor cells were glial fibrillary acidic protein positive, and Ki67 positivity was 40%. Gene analysis showed IDH wild-type, epidermal growth factor receptor (EGFR) amplification, homozygous loss of CDKN2A, loss of chromosome 10, and positive MGMT methylation. The patient underwent radiation therapy with concurrent chemotherapy with temozolomide. Two and a half years after diagnosis, the patient was doing well neurologically with a stable disease burden on follow-up MRI.

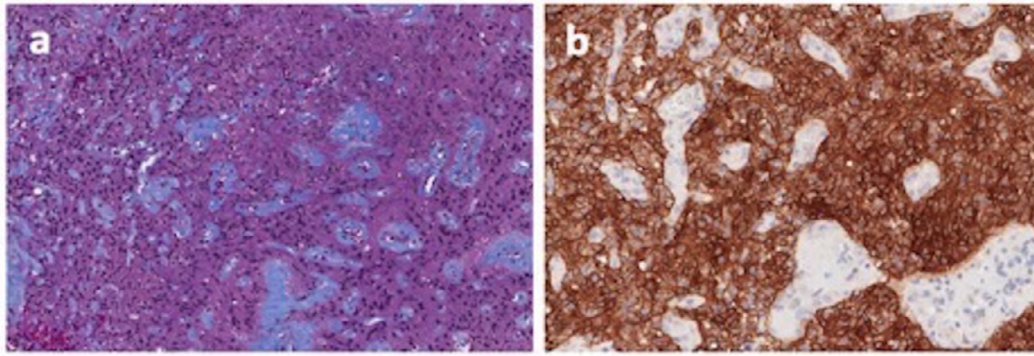


Fig. 2 – (a) The abundance of collagen, associated with blood vessels and fibrous bands, is seen as blue material in this photomicrograph (trichrome stain, 225 ×). (b) In this photomicrograph, the neoplastic astrocytes stain brown and are admixed with numerous vessels and collagen bands, which are unstained (glial fibrillary acidic protein [GFAP] immunohistochemistry, 350 ×).

Discussion

The dural tail sign was first described in 1989 and was initially used to diagnose meningiomas with the assumption that this radiographic finding was almost entirely specific to this condition [5]. Recent evidence has shown, however, that the dural tail sign is also present in several other neoplasms. In fact, a study by Rokni-Yazdi and Sotoudeh concluded that the dural tail was 94% specific, suggesting that more data are often necessary to differentiate true meningiomas from other neoplasms that carry a more fatal prognosis such as GBMs [4,6]. It also prompts discussion as to the utility of the dural tail sign in the future. Currently, primary staging and a presumptive diagnosis are achieved by noninvasive radiographic imaging such as MRI and CT scans. MRI is often used to differentiate brain tumors from non-neoplastic lesions and to stratify into low-grade versus high-grade lesions [7]. Advanced MRI can provide independent and complementary prognostic information [8]. This in turn, influences surgical decision making and other important aspects of patient care. A more extensive surgical resection can potentially be associated with a better prognosis [9].

A noninvasive tool that can be used to help distinguish GBM from meningioma is 3T proton MR spectroscopy. This technique, which utilizes the spin properties of individual hydrogen atoms, can help analyze the biochemical components of living tissue to further improve diagnosis. One study found that the presence of a signal intensity at 3.8 ppm was able to reliably distinguish meningiomas from high-grade gliomas or metastases [10]. This unique signal is thought to represent a combination of phosphoethanolamine and amino acids such as alanine, glutamate, glutamine, glutathione, lysine, arginine, and serine, found in this subset. Lack of a distinct peak of 3.8 ppm signal intensity is suggestive rather of a metastasis or high-grade glioma, as would be found in a GBM.

In a recent literature review, the first account of an extracerebral GBM, consisting of neoplastic ependymal cells intermixed with glial fibrillary acidic protein tumor cells, was reported in 1984 [11]. Since then, there have been 5 other reports of extracerebral primary GBM neoplasms with dural-

based tails, in addition to the one that we are presenting here [11–15]. It is important to highlight this unique feature of GBMs and encourage its inclusion on the differential diagnosis given its prognostic significance. It has been reported that the median survival of glioblastoma is between 6 and 12 months [16]. A recent population-based study in Switzerland reported that only 3% of patients survived more than 2 years [17]. Another study found that only 6.4% of GBM patients who are 71 years of age or older survived longer than 2 years [18]. This is in stark contrast to meningiomas, with which patients can live asymptotically often throughout their lives. The set of prognostic factors that influence the course of this terminal brain tumor are still poorly understood and likely include size, molecular markers, and adjuvant treatment. This subclassification of GBMs warrants further study in order to be able to provide patients with the most accurate date to help them with treatment and life decisions.

Our case report represents an important addition to this new emergency of extracranial GBMs presenting with dural tails. Altogether, there are still only a handful of cases of extracerebral GBMs and this emerging trend warrants further discussion and investigation, especially with regards to the utility of the dural tail sign in terms of prognostication. This imaging finding was previously thought to be almost exclusive of meningiomas. It is important, however, to highlight that the differential diagnosis can and should include GBM, which implies a significantly lower chance of survival.

REFERENCES

- [1] Ostrom QT, Gittleman H, Fulop J, Liu M, Blanda R, Kromer C, et al. CBTRUS Statistical Report: Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2008–2012. *Neuro Oncol* 2015. <https://doi.org/10.1093/neuonc/nov189>.
- [2] Nicholas MK, Lukas R V, Chmura S, Yamini B, Lesniak M, Pytel P. Molecular heterogeneity in glioblastoma: therapeutic opportunities and challenges. *Semin Oncol* 2011. <https://doi.org/10.1053/j.seminoncol.2011.01.009>.

- [3] Zygogianni A, Protopapa M, Kougioumtzopoulou A, Simopoulou F, Nikoloudi S, Kouloulas V. From imaging to biology of glioblastoma: new clinical oncology perspectives to the problem of local recurrence. *Clin Transl Oncol* 2018;20:989–1003.
- [4] Rokni-Yazdi H, Sotoudeh H. Prevalence of “dural tail sign” in patients with different intracranial pathologies. *Eur J Radiol* 2006. <https://doi.org/10.1016/j.ejrad.2006.04.003>.
- [5] Wilms G, Lammens M, Marchal G, Van Calenbergh F, Piets C, Van Fraeyenhoven L, et al. Thickening of dura surrounding meningiomas: MR features. *J Comput Assist Tomogr* 1989. <https://doi.org/10.1097/00004728-198909000-00003>.
- [6] Sotoudeh H. A review on dural tail sign. *World J Radiol* 2010. <https://doi.org/10.4329/wjr.v2.i5.188>.
- [7] Fink JR, Muzi M, Peck M, Krohn K. Multimodality brain tumor imaging: MR imaging, PET, and PET/MR imaging. *J Nucl Med* 2015. <https://doi.org/10.2967/jnumed.113.131516>.
- [8] Young RJ, Gupta A, Shah AD, Graber JJ, Zhang Z, Shi W, et al. Potential utility of conventional MRI signs in diagnosing pseudoprogression in glioblastoma. *Neurology* 2011. <https://doi.org/10.1212/WNL.0b013e31821d74e7>.
- [9] Brown TJ, Brennan MC, Li M, Church EW, Brandmeir NJ, Rakszawski KL, et al. Association of the extent of resection with survival in glioblastoma a systematic review and meta-Analysis. *JAMA Oncol* 2016;2:1460–9. <https://doi.org/10.1001/jamaoncol.2016.1373>.
- [10] Kousi E., Tsougos I., Kapsalaki E. Proton magnetic resonance spectroscopy of the central nervous system. *Novel frontiers of advanced neuroimaging*. Kostas N. Fountas, editors IntechOpen2013 Jan 9. <https://doi:10.5772/53892>.
- [11] Shuangshoti S, Kasantikul V, Suwanwela N, Suwanwela C. Solitary primary intracranial extracerebral glioma. Case report. *J Neurosurg* 1984;61:777–81. <https://doi.org/10.3171/jns.1984.61.4.0777>.
- [12] Hsieh CT, Liu MY, Tang CT, Sun JM, Tsai WC, Hsia CC. Problem of dural tail sign in glioblastoma multiforme? *Acta Neurol Belg* 2009;109:310–13.
- [13] Stavrinou P, Magras I, Stavrinou LC, Zaraboukas T, Polyzoidis KS, Selviaridis P. Primary extracerebral meningeal glioblastoma: clinical and pathological analysis. *Zentralbl Neurochir* 2010;71:46–9. <https://doi.org/10.1055/s-0029-1225652>.
- [14] Wu B, Liu W, Zhu H, Feng H, Liu J. Primary glioblastoma of the cerebellopontine angle in adults. *J Neurosurg* 2011. <https://doi.org/10.3171/2010.12.JNS10912>.
- [15] Patel M, Nguyen HS, Doan N, Gelsomino M, Shabani S, Mueller W. Glioblastoma mimicking meningioma: report of 2 cases. *World Neurosurg* 2016;95:624. <https://doi.org/10.1016/j.wneu.2016.08.048>.
- [16] Smoll NR, Schaller K, Gautschi OP. Long-term survival of patients with glioblastoma multiforme (GBM). *J Clin Neurosci* 2013. <https://doi.org/10.1016/j.jocn.2012.05.040>.
- [17] Ohgaki H, Dessen P, Jourde B, Horstmann S, Nishikawa T, Di Patre PL, et al. Genetic pathways to glioblastoma: a population-based study. *Cancer Res* 2004. <https://doi.org/10.1158/0008-5472.CAN-04-1337>.
- [18] Krex D, Klink B, Hartmann C, von Deimling A, Pietsch T, Simon M, et al. Long-term survival with glioblastoma multiforme. *Brain* 2007. <https://doi.org/10.1093/brain/awm204>.