Radiology Case Reports 17 (2022) 729–734

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journal homepage: www.elsevier.com/locate/radcr

Case report

Glioblastoma in the optic chiasm: A case report☆,☆☆

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ARTICLE INFO

Article history:
Received 19 November 2021
Revised 4 December 2021
Accepted 13 December 2021

Keywords:
Magnetic resonance imaging
Optic glioma
Optic nerve
Glioblastoma

ABSTRACT

Malignant optic gliomas are an uncommon pathology, with around 67 cases reported worldwide in the literature. We present the case of a 77-year-old-male with a two-month history of progressive vision loss, ultimately leading to bilateral blindness. The initial clinical suspicion was a non-inflammatory ischemic optic neuropathy. Stereotactic biopsy was performed on the optic chiasm, and the histopathological diagnosis was confirmed as Glioblastoma.

Introduction

Optic gliomas represent 0.6%–1.2% of all brain tumors detected in general [1]. Most are benign and occur in the pediatric population before age 15 (85%) and are strongly associated with Type I neurofibromatosis [2,3]. Malignant optic gliomas rarely occur in adults; this entity was defined for the first time in 1973 by Hoyt et al. [4]. There have been 67 cases reported (70) in the medical literature, of which 26 [5] correspond to Glioblastoma (GBM).

Patients are usually middle-aged men presenting with progressive deterioration of vision, which leads to blindness within a few weeks. The initial diagnostic impression is often wrong due to symptoms misinterpreted as ischemic neuropathy, optic neuritis, or retinal vascular occlusion [3].

The tumor may originate in the optic chiasm, the optic tract, or the optic nerves and may occur as multifocal neoplasia. Magnetic resonance imaging (MRI) of the brain is the primary diagnostic tool. Nevertheless, the findings are nonspecific. A biopsy is needed to confirm the diagnosis. Pathologic features can be consistent with Anaplastic Astrocytoma (grade III - WHO) or GBM (grade IV- WHO) [6].

We present the case of a patient diagnosed with GBM in the optic chiasm and optic nerves. We also briefly review the clinical manifestations, neuroradiological findings, and clinical outcomes of the optical GBM cases reported in the literature to this date.

☆ Acknowledgments: The author(s) received no financial support for the research, authorship, and/or publication of this article. The authors thank Dr Camila Ospina Jiménez for her initial collaboration.
☆☆ Competing interests: The authors declare no conflicts of interest in the publication of this article.
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https://doi.org/10.1016/j.radcr.2021.12.020

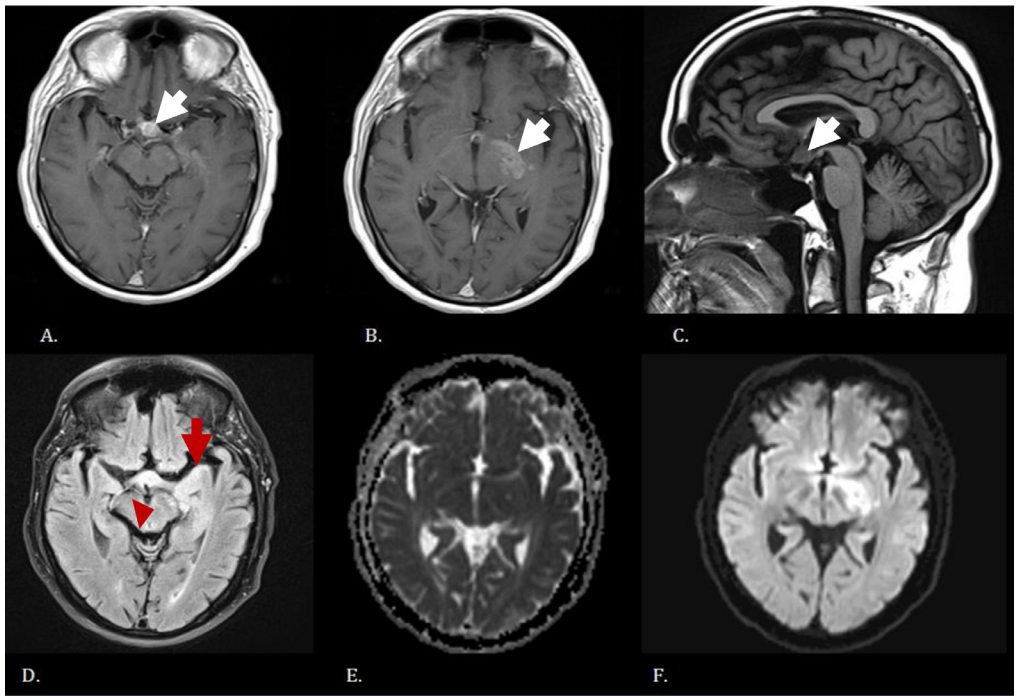

Fig. 1 – (A) and (B) Axial T1 contrast-enhanced sequence with evidence of nodular thickening of the optic chiasm (white arrow on A) and left optic tract (white arrow on B) associated with enhancement of these structures. (C) Sagittal T1 sequence demonstrating evidence of nodular thickening of the optical chiasm (white arrow). (D) Axial FLAIR sequence with hyperintensity of the temporal lobe's mesial region (red arrow) and the midbrain's left lateral margin (red arrow head). (E) and (F) Axial ADC, and DWI sequences show no evidence of restriction of the previously described structures.

Case report

A 77-year-old-man with a two-month history of progressive bilateral vision loss, more prominent on his left eye, presented to the ophthalmologist's office. The initial presumptive diagnosis was a non-arteritic ischemic optic neuropathy, for which the patient was referred to the emergency room. The patient was assessed by a clinical neurologist, who did not find headache, mandibular claudication, or pain associated with eye movements or in the temporal area. During the physical examination, pupils were hyporeactive, and visual acuity was diminished with dyschromatopsia on the right eye and absence of light perception on the left eye. The patient had no further neurologic compromise.

Given the clinical manifestations, a brain and orbit MRI was ordered, revealing evidence of a non-uniform enhancement in the optic chiasm and the left optic tract. Thickening and cortical enhancement were also found in the left mesial temporal lobe region and left lateral margin of the midbrain (see Fig. 1). These findings suggested an inflammatory compromise or demyelination, though it was not possible to rule out neoplastic infiltration. Under the suspicion of demyelinating disease, an analysis of oligoclonal bands was requested. The patient was treated with methylprednisolone and showed modest clinical improvement. He was discharged and sent for ambulatory studies in the Neurology Department.

The patient returned one month later due to bilateral blindness with no additional neurologic symptoms. A new brain contrast-enhanced MRI was performed and demonstrated progressive thickening and enhancement of the same structures compared to the previous study and a heterogeneously enhancing mass in the left optic tract with similar features (see Fig. 2). Due to these new findings, the patient was referred for a stereotactic biopsy of the lesion. This procedure was made following a pallidotomy to ensure that the optic tract was reached precisely. Pathology reported a glioblastoma with a Ki67 marker of 25%.

Due to the lesion's location, neurologic status, and clinical deterioration, surgical resection was not possible. He was offered palliative care. The patient died five months after the initial presentation.

To be granted access to clinical information and diagnostic images, the institutional ethics committee was requested to waive the case report from informed consent.

Discussion

Optic malignant gliomas are exceedingly rare. Wabbels et al. (2004) undertook a literature review and described 45 cases [7]. More recently, Traber et al. (2015) reported five patients and identified 21 additional cases published after 2004 [6]. Subsequently, Nagaichi et al. (2015) reported a new case [1]. After this report, and to our knowledge, there are at least 71 cases of optic gliomas malignancies in the medical literature, 30 corresponding to GBM (see Table 1).

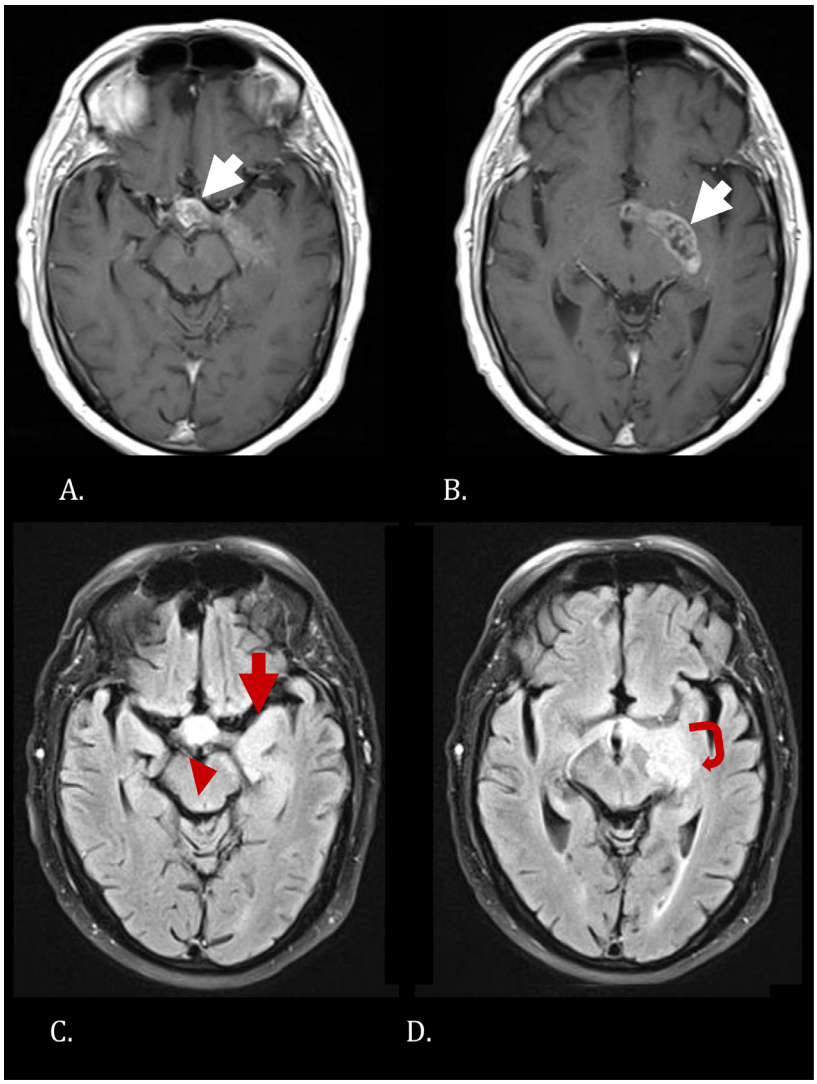

Fig. 2 – (A) and B) Axial contrast-enhanced T1 sequence demonstrates increased thickening and heterogeneous enhancement of the chiasm (white arrow on A) and left optic tract (white arrow on B). (C) and (D) Axial FLAIR sequence shows increased hyperintensity of the mesial portion of the temporal lobe (red arrow con C), midbrain's left lateral margin (red arrowhead on C), optic chiasm and tracts (curved red arrow on D).

Our patient presented with imaging findings and clinical characteristics like those reported in other cases of GBM, in which the median age of presentation was 61, and there was a slight predominance of male gender (11 women and 16 men). The clinical picture is characterized by rapidly progressive vision loss, either unilateral or bilateral, in conjunction with headache or periorbital pain [3]. The average time to complete vision loss in the reported cases was 5–6 weeks. Proptosis is exceptionally rare in these cases. In 1949, Saebo reported the case of a 43-year-old-woman with four months of vision loss and simultaneous proptosis of the left eye, for whom the final diagnosis was optical GBM [8].

GBM is frequently confused with another diagnosis due to the unspecific clinical picture. Non-arteritic ischemic optic neuropathy, for example, was suggested as the initial presumptive diagnosis in our case since this usually occurs in patients over their 50′s and presents as a sudden, painless loss of vision which is either unilateral or bilateral [9,10]. Another differential diagnosis is optic neuritis, which usually presents as a unilateral loss of vision; this is, however, usually found in patients younger than 50 years of age and improves with the administration of corticosteroids [11,12]. GBM can also be confused with other diagnoses such as retinal vein occlusion and temporal arteritis [3].

The tumor's site of origin determines whether the initial vision loss is unilateral or bilateral and the presence or absence of ophthalmic symptoms. Tumors originating in the optic nerve initially cause unilateral vision loss and bilateral changes in fundoscopy (disc edema and venous stasis). On the other hand, tumors in the optic chiasm present bilateral visual alterations and normal fundoscopy. The latter is more likely to infiltrate the hypothalamus and the walls of the third ventricle [3,13]. If the tumor is in the optic tract, there will be a visual field deficit contralateral to the side of the lesion.

Table 1 – Characteristics of the reported cases of Glioblastoma.

Author	Age/Sex	Time to blindness	Initial diagnosis	MRI	Treatment	Time to death after dx
Saebo 1949 [7]	43/F	-	Tumor of the optic nerve	-	RT and surgical resection	One year
Mattson 1966 [16]	59/ F	1 mo	Optic neuritis	-	No treatment	4 mo
Hoyt et al. 1973 [4]	55 / M	-	-	-	RT	9–10 mo
Harper and Stewart-Wynne 1978 [17]	75 / M	-	Brain tumor	-	No treatment	3 mo
	79/ F	-	Brain tumor	-	No treatment	3 mo
Spoor et al. 1980 [18]	60 / F	2 mo	-	-	RT	6 mo
Barbaro et al. 1982 [19]	26 / M	2 wk	Craneopharyngioma – pituitary adenoma cyst	-	RT y CT	8 mo
Evens et al. 1987 [20]	61 / F	1 mo	-	-	RT	9 mo
Albers et al. 1988 [14]	51/ M	2 wk	-	T2 (Hyperintense)	No treatment	20 mo
Woiciechowsky et al. 1995 [21]	76 / M	-	Malignant optic glioma	T1 + Contrast (Enhancement)	Surgical resection and RT	-
Pallini et al. 1996 [22]	59 /F	-	-	-	RT	7 mo
Hahn et al. 2004 [23]	53 /M	-	Neurosarcoidosis	T1 + contrast (Enhancement) T2 (Hyperintense)	RT and CT	-
Hartel et al. 2006 [24]	59 /M	<1 mo	Unknown	-	None	8 wk
Dinh et al. 2007 [3]	48 /F	10 wk	Tumor of the optic nerve	T1 + contrast (Enhancement)	RT	14 mo
Abou - Zeid et al. 2008 [25]	56 /M	<1 mo	Metastatic brain disease	T1 + contrast (Enhancement)	RT	3 mo
Brar et al. 2009 [26]	68 / F	2 mo	-	T1 hypointense - isointense T1 + contrast (Enhancement) T2 (Hyperintense)		-
Matloob et al. 2011 [12]	63 / F	<1 mo	Optic neuritis	T1 + contrast (Enhancement) T2 (Hyperintense)	CT	6 mo
Lincoff et al. 2012 [27]	83 / M	<1 mo	-	T1 + contrast (Enhancement) T2 (Hyperintense)	-	-
Ashur- Fabian et al. 2013 [15]	64/ M	6 mo	-	T1 + contrast (Enhancement)	RT and CT	54 mo
Colpak et al. 2014 [28]	47 / M	<1 mo	Optic neuritis or infiltrative	T1 + contrast (Enhancement)	-	3 mo
Traber et al. 2015 [5]	65 / M	5 wk	Tumor of the optic nerve	T1 + contrast (Enhancement) T2 (Hyperintense)	RT and CT	4.5 mo
	54 / M	7 mo	Optic neuritis	T1 + contrast (Enhancement) T2 (Hyperintense)	RT and CT	18 mo
	64 / F	6 wk	Tumor of the optic nerve	T1 + contrast (Enhancement) T2 (Hyperintense)	RT	6 mo
	75/ M	6 wk	Optic neuritis	T1 + contrast (Enhancement) T2 (Hyperintense)	RT	12 mo
	76 / F	4 wk	Non-artieric ischemic optic neuritis	T1 + contrast (Enhancement) T2 (Hyperintense)	RT	7 mo
Nagaishi et al. 2015 [1]	64 / F	-	-	T1 + contrast (Enhancement) T2 (Hyperintense)	Radiosurgery and CT	10 mo
Mastorakos et al. 2017 [30]	66/M	12 mo	Meningioma	T1 + contrast (Enhancement)	Surgical resection and CT	16 mo
Lin et al. [31]	21/M	4 mo	Optic neuritis	T1 + contrast (Enhancement)	Srugical resection and CT	-
Alabiad et al. 2020 [29]	18/F	4 wk	Unknown	T1 + contrast (Enhancement)	Surgical resection, CT and RT	Alive (>7 y)
Current case	77/ M	3 mo	Optic neuritis	T1 + contrast (Enhancement) T2 (Hyperintense)	None	4 mo

Wabbels et al. (2004) revised 45 cases, and Traber et al. (2015) reported 26 cases.

The most common affected sites are the chiasm, and optic nerves are the hypothalamus, the temporal lobe, and the basal ganglia [7]. Symptoms of hypothalamic involvement include polyuria and polydipsia and may appear in the late course of the disease, suggesting infiltration [14].

The neuroimaging features are non-specific in the early stages of the disease, as noted by Albers et al. when they described the first case, including the results from an MRI study [15]. The reported findings were enhancement in the post-contrast T1 sequence and thickening of the nerve, chiasm, or the optic tract in the T1 weighted images. T2 hyperintensity has also been reported to affect the anterior visual pathway, initially described by Albers et al. in 1988; this last characteristic is still debatable [6]. The imaging findings reported in the

medical literature are like those observed in our case, including enhancement of the optic nerve in the post-contrast T1 images and T2 hyperintensity.

There are similar imaging findings in conditions such as multiple sclerosis, peri-optic neuritis, low-grade primary optic glioma, lymphoma, leukemia, and metastasis. Infiltrative processes produce a fusiform thickening, while inflammatory processes produce a more tubular enhancement [13].

Some diseases thicken the optic nerve without affecting the sheath (optic neuritis, optic nerve ischemia). In contrast, other processes affect the sheath without any nerve swelling (meningioma, pseudotumor). The pathologies that affect both nerves and the sheath include optic glioma, peri-optic neuritis secondary to infection and granulomatous disease, leukemia, lymphoma, and metastatic disease [14]. In recent years it has been proposed that Color Doppler Flow Imaging might help the diagnosis of malignant optic gliomas.

The prognosis is poor for GMB, with an average survival time of 8–9 months of all 27 cases found in the literature. Al-abiad et al. (2020) reported the case with the most extended survival (7 years, and still alive) [5].

Conclusion

Optic nerve GBM is a rare neoplasm with rapid evolution; early diagnosis is difficult due to the lack of a clear clinical presentation and characteristic imaging features. It is crucial to consider malignant optical glioma as a differential diagnosis in cases similar to that reported.

Data sharing statement

The relevant anonymized patient-level data are available via request from the authors.

Ethical approval and patient consent

The report case was reviewed and approved. Following our institutional guidelines, all protected health information was removed, and individual patient consent was not required for the analysis.

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