



Case Report

Ganglioglioma of optic chiasma: A case report and review of literature

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ABSTRACT

Background: Gangliogliomas are neoplasms containing both astrocytic and neuronal components. We present a case of gangliogliomas of the optic chiasm, which are extremely rare pathologies.

Case Description: A 16-year-old female patient referred to our clinic with gradual deterioration of vision for the age of 1 year mostly in the right eye. Ophthalmic examination confirmed reduced visual acuity with only perception of light in the left eye. Brain magnetic resonance imaging showed a solid mass lesion involving the hypothalamus and the optic chiasm, which was hypointense on T1-weighted images, hyperintense on T2-WI, and marked homogenous contrast enhancement. The patient was operated and bulging of the optic chiasm and the site of lamina terminalis was seen. Subtotal resection of the tumor was achieved. Histopathological examination revealed ganglioglioma (WHO Grade I). Follow-up of the patient was for 3 years and 8 months with stable neurologic and radiologic findings.

Conclusion: To the best of our knowledge, 20 cases, including ours, have been reported in the literature and a presurgical diagnosis of ganglioglioma is very infrequent with confused radiologically with low-grade pilocytic astrocytomas.

Keywords: Ganglioglioma, Optic chiasm, Suprasellar tumor

INTRODUCTION

First described by Courville in 1930, the term ganglioglioma was introduced to define neoplasms containing both astrocytic and neuronal components.^[5] Although rare, gangliogliomas are the most frequently occurring mixed glioneuronal tumors of the central nervous system (CNS), corresponding to the WHO Grade I or II.^[10] Their incidence ranges from 0.4 to 1.3% of all brain tumors, but they are more common in the pediatric group, with an incidence of 7.6%.^[11] The mean male-to-female ratio is 1.3:1.^[8] The mean age at diagnosis, considering all sites, is approximately 20 years, with a range of 75–80 years.^[11] The temporal lobes are the most common site, but ganglioglioma can occur anywhere in the central neuraxis, including the brain stem and spinal cord.^[10,13]

Gangliogliomas of the optic chiasm are extremely rare pathologies and intensive literature search by the authors showed only 20 cases of optic chiasm.

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Ganglioglioma, including the current case, has been reported so far.^[1,2,3,4,7,9,11,12,13,14,15,16,17,18,19,20] Radiological, clinical, and pathologic data with intraoperative findings are discussed, with a brief review of similar cases in the literature.

CASE REPORT

A 16-year-old female patient referred to our outpatient clinic with gradual deterioration of vision for the age of 1 year mostly in the right eye, with episodes of sharp headaches. Ophthalmic examination confirmed reduced visual acuity with only perception of light in the left eye. Neurological examination showed no other neurologic deficit. Brain magnetic resonance imaging (MRI) showed a solid mass lesion involving the hypothalamus and the optic chiasm. The lesion was hypointense on T1-weighted images, hyperintense on T2-WI, and showed marked homogenous enhancement after the administration of contrast material [Figure 1]. The patient was operated by the right pterional craniotomy and transsylvian approach. Bulging of the optic chiasm and the site of lamina terminalis were seen. Incision with knife was done on the right side of optic chiasm through the exophytic part of the tumor and tumor evacuation done from inside the chiasm [Figure 1e]. Furthermore, resection of the suprachiasmatic part of the tumor was performed and lamina terminalis was opened. Tumor entry sites were planned to minimize the risk of neural injury. Entry to the optic chiasm was performed from the lateral part of the right optic chiasm where tumor was bulging and exophytic

with minimal covering neural tissue. Furthermore, this site is far from the median part of the chiasm in which crossing of optic fibers takes place. In the suprachiasmatic part, we also aimed to the exophytic part of the tumor and planned to enter through the lamina terminalis, which is known for its safety for dissection, to the anterior part of the third ventricle. In general, there was no clear-cut plain of cleavage, and dissection was carried on according the difference of color and consistency between the normal tissue and the friable grayish tumor tissue. Yet, subtotal resection of the tumor was achieved due to the absence of this difference between a part of the tumor and the hypothalamus. Thus, we decided to leave this part rather than inducing hypothalamic injury. Early postoperative period was uneventful, however, the patient showed no change in the visual status.

Histopathological examination revealed ganglioglioma (WHO Grade I). Microscopic examination showed clusters of abnormal neurons which exhibit enlarged hyperchromatic nuclei and some are binucleated. Between the neurons, there are dissecting fibrillary cells highlighted by GFAP. No necrosis, mitosis or endovascular proliferation were seen. The abnormal neurons are immunoreactive for S-100 and Synaptophysin, negative for EMA and CEA. CD68 highlight scattered macrophages in the background. KI67 was less than 1% [Figure 2]. The low KI67 and the low grade of the tumor (WHO Grade I) influenced our decision to follow up the patient than subsection radiotherapy, and until this report it has been 3 years and 8 months with stable neurologic and radiologic findings of the patient [Figure 3].

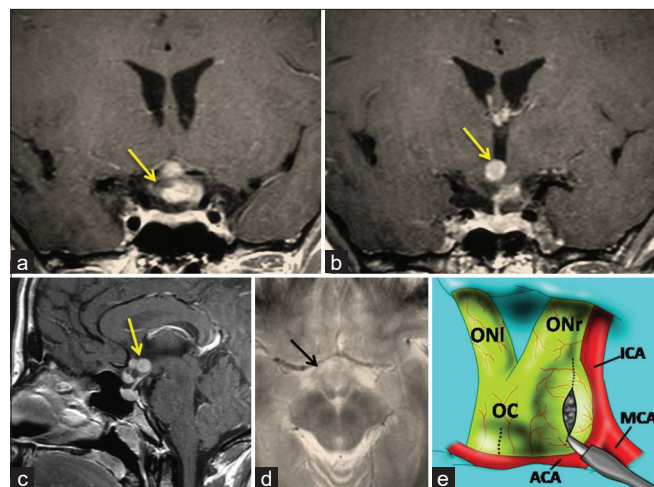


Figure 1: Preoperative brain magnetic resonance imaging showing a mass lesion involving the optic chiasm and hypothalamus (arrows). (a and b) Coronal postcontrast, (c) sagittal postcontrast, and (d) axial T2-weighted images. (e) Schematic drawing demonstrating the enlarged optic chiasm due to tumor and the entry site through the exophytic part of the tumor in the right optic chiasm and lamina terminalis (dotted lines). OC: Optic chiasm, ONr: Right optic nerve, ONl: Left optic nerve, ICA: Internal carotid artery, MCA: Middle cerebral artery, ACA: Anterior cerebral artery.

DISCUSSION

Gangliogliomas involving the optic chiasm are extremely rare. To the best of our knowledge, 20 cases, including ours, have been reported in the literature [Table 1]. Adequate clinical data were available for 19 patients. The mean age was 15 years; 89% of the tumors were diagnosed during the first three decades. There was a slight male predilection with a ratio of 1.38:1. The presenting symptoms varied depending on the extent of the lesions. However, all of the patients had visual impairment, as in our case.

On CT scan, gangliogliomas appear as hypodense lesions with calcification and a variable contrast enhancement pattern. The MRI findings are nonspecific and mimic pilocytic astrocytomas, as at this site they are indolent, slow-growing neoplasms, and their presenting symptoms vary depending on location.^[18] As in our case, the lesions may be hypointense on T1-weighted images and hyperintense on T2-weighted images. Contrast enhancement was reported in 44% of one series and may have either a nodular rim or solid enhancement pattern.^[16] As in our case, 40% of cases may appear as solid lesions only. Cystic tumor components occur in the rest of the cases.^[16] An interesting results in a study

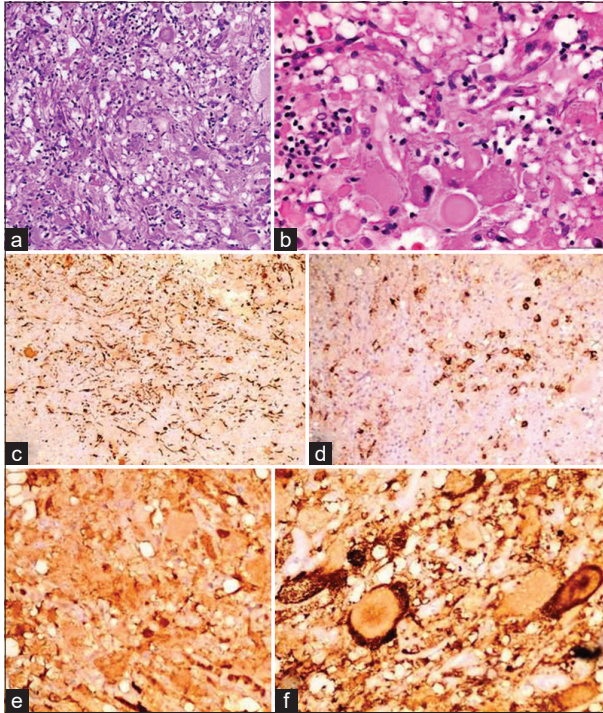


Figure 2: Histopathologic examination of the tumor revealed clusters of abnormal neurons which exhibit enlarged hyperchromatic nuclei and some are binucleated in H&E $\times 10$ (a) and H&E $\times 40$ (b). Between the neurons, there are dissecting fibrillary cells highlighted by GFAP (c). The abnormal neurons are negative for EMA (d) and positive for S-100 (e) and Synaptophysin (f).

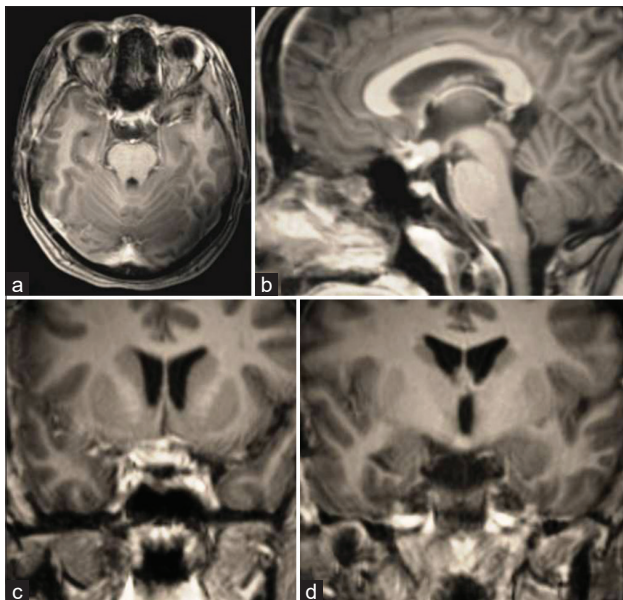


Figure 3: Postoperative postcontrast brain magnetic resonance imaging 3 years after surgery showing small and stable remnant of the tumor. (a) Axial, (b) sagittal, (c and d) coronal images.

performed by Sugita *et al.* measuring the metabolic activity of low-grade gangliogliomas by proton MR spectroscopy

and positron emission tomography (PET) scan, they findings suggestive of high-grade tumors, although all cases were histologically benign.^[18] On proton MR spectroscopy, gangliogliomas show increased Cho/NAA ratio, while they are a low-grade gangliogliomas.^[18] This increased Cho/NAA ratio may indicate high glial and myelin content of gangliogliomas.^[18] 2-deoxy-2-^[18F] fluoro-D-glucose (^{18F}-FDG) PET scan showed a markedly hypermetabolic suprasellar lesion. Hypermetabolism shown by ^{18F}-FDG PET may reflect the larger neuronal cell population of gangliogliomas.^[18] Furthermore, in their study, performing α -[N-methyl-¹¹C]-methylaminoisobutyric acid (¹¹C-MeAIB) PET scan showed that high accumulation in the lesion, with the lesion to gray matter ratio (LGR) of ¹¹C-MeAIB uptake was extremely high (39.01).^[18]

Histologically, gangliogliomas are considered benign tumors, usually classified as WHO Grade 1. The cases of primary atypical (WHO Grade 2) and anaplastic (WHO Grade 3) gangliogliomas are more rare.^[6] There are even rare cases of newly diagnosed gangliogliomas showing Grade IV (glioblastoma) changes in the glial component.^[6] In our case, histopathologic examination revealed clusters of abnormal neurons which exhibit enlarged hyperchromatic nuclei and some are binucleated in H&E. However, the biphasic cellular pattern exhibited by gangliogliomas is not always uniform, and it can range from variants with predominant glial population to variants with prominent neuronal population.^[17] Ganglion cells must be distinguished from non-neoplastic neurons which might be encapsulated into an astrocytoma: neoplastic ganglion cells usually appear as clusters of cells of different size, often enlarged and binucleated, with large nuclei, prominent nucleoli, abundant cytoplasm, and Nissl's substance.^[1] As in our case, the positivity at immunohistochemistry for neurofilament protein and Synaptophysin aids the differentiation of ganglion cells with hyperplastic or neoplastic astrocytes. Glial cells are usually proliferating fibrillary astrocytes with rare mitotic figures.^[1] The origin of the neuronal component in case of gangliogliomas of the optic pathway is still controversial. Moreover, normally pregeniculate optic pathway does not contain ganglion cells, but only the axons originating from the ganglion cells of the retina. There are two theories that have been developed to explain the histogenesis of these tumors. The first is developmental hypothesis. During embryological development, the optic stalk is lined by a layer of undifferentiated cells and is filled by nerve fibers extending from retinal ganglion cells.^[1,13] In subsequent stages, the primitive epithelium disappears, leaving only precursors of the supporting glial cells.^[13] Ganglion cells of ganglioglioma may originate from these elements. Another hypothesis maintains that neoplastic neurons originate from perivascular sympathetic neurons.^[1]

Table 1: Summary of the reported cases of optic chiasm ganglioglioma.

Age (years)/sex	Site	Treatment	Follow-up	Reference
6/M	OC + Hypothalamus	Biopsy + RT	5 years, stable	Liu <i>et al.</i>
10/F	Right ON + OC	ST + RT	4 years, stable	Cogan <i>et al.</i>
13/F	OC	ST	8 months, extensive spread	Liu <i>et al.</i>
15/F	Right PG	Biopsy	6 years	Sugiyama <i>et al.</i>
16/M	Right ON + OC	Resection + RT	3 years	Lowes <i>et al.</i>
21/F	OC + Hypothalamus	ST	3 months	Shuangshoti <i>et al.</i>
23/M	OC + optic tracts	ST + RT	17 years, stable	Liu <i>et al.</i>
24/M	Right ON + OC	Resection and RT	2 years 11 months	Lowes <i>et al.</i>
33/M	OC + Right optic tract	Biopsy + RT	1 year, stable	Chilton <i>et al.</i>
38/M	Right PG	ST	NA	Lu <i>et al.</i>
NA	OC + Hypothalamus	NA	NA	Rodriguez <i>et al.</i>
52/F	Right optic tract	NA	NA	Albayrak <i>et al.</i>
8/F	OC + Optic tract	ST	6 months, stable	Pant <i>et al.</i>
18/M	OC + Left optic tract + Right lateral geniculate body + Right optic radiation	Biopsy	NA	Vajramani <i>et al.</i>
11/M	Right ON +OC +Bilateral optic tracts + Right temporal lobe + Right midbrain + Hypothalamus + Thalamus.	Biopsy	9 years. malignant transformation in atypical teratoid/rhabdoid tumor death	Allen <i>et al.</i>
7/M	OC + Bilateral ON	Biopsy + RT	NA	Gupta <i>et al.</i>
6/M	Bilateral PG	ST + ChT + RT	4 years, stable	Spennato <i>et al.</i>
26/M	OC	ST + Fractionated SRT	NA	Sugita <i>et al.</i>
7/F	OC + Bilateral ON + Hypothalamus	Biopsy + Conformal SRT	NA	Jalali <i>et al.</i>
16/F	OC +Hypothalamus	ST	3 years 8 months, stable	Current case

F: Female, M: Male, OC: Optic chiasm, ON: Optic nerve, PG: Pregeniculate pathway, NA: None available information, ST: Subtotal resection, RT: Radiotherapy, SRT: Stereotactic radiotherapy, ChT: Chemotherapy

Most CNS gangliogliomas are well circumscribed. Therefore, surgical excision is possible, resulting in a generally favorable prognosis.^[11,13] However, complete resection often cannot be accomplished in cases with chiasmatic involvement. The role of postoperative radiotherapy is controversial and usually reserved for those with disease progression or recurrence.^[11,13] Nevertheless, radiotherapy has been applied in those cases in which only biopsy or subtotal resection could be performed.^[2,4,7,9,11,12,14,20] In our case, the low KI67 and grade of the tumor directed our decision to follow-up the patient and preserve the radiotherapy in case of progression of the tumor, which showed stability of the tumor until the time of reporting this case (3 years and 8 months). Follow-up was available for 13 patients. All were alive during 8 months–17 years of follow-up, except one case died after 9 years after rare malignant transformation in atypical teratoid/rhabdoid tumor.^[2] No cases, including ours, showed significant visual improvement.

CONCLUSION

Ganglioglioma of the optic chiasm is extremely rare pathologies and a presurgical diagnosis of ganglioglioma is very infrequent with confused radiologically with low-grade

pilocytic astrocytomas. We report this case due to its relative rarity and to broaden the differential diagnoses of lesions arising in the optic chiasm.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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