

Case Report

Optic nerve glioma: A great mimicker

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Abstract**Background:** Arachnoid proliferation, although rare, is known to occur in association with optic gliomas. However, chondroid and chordoid metaplasia has not been reported previously.**Case Description:** A 27-year-old male presented with progressive, painless loss of vision in right eye, associated with vomiting and headache for one and a half months. Computed tomography (CT) scan revealed a contrast enhancing mass arising from planum sphenoidale. Perioperative findings showed the tumor adherent to the right optic nerve and attached to basal dura and falx. A clinical impression of an intradural, optic nerve sheath meningioma was made. Histopathological examination revealed a glial tumor with adjacent areas displaying marked fibroblastic and arachnoid cell proliferation with chondroid as well as chordoid differentiation along with myxoid change and dense collagenisation. Reticulin stain, immunohistochemistry with glial fibrillary acid protein (GFAP), epithelial membrane antigen (EMA), and S-100 helped to arrive at the final diagnosis of optic glioma displaying exuberant arachnoid proliferation with cartilaginous metaplasia.**Conclusion:** We report a case of optic nerve glioma displaying extensive arachnoid proliferation, chordoid, and cartilaginous metaplasia, which mimicked chondrosarcoma or chordoid meningioma, posing a diagnostic dilemma. A clinical feedback, simple reticulin stain, and GFAP staining is of immense value in such cases to arrive at the correct diagnosis.**Key Words:** Arachnoid hyperplasia, chondroid, chordoid, glioma, optic nerve**Access this article
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Quick Response Code:**INTRODUCTION**

Optic nerve gliomas are rare tumors, accounting for 1.5-3% of orbital tumors, 1% of intracranial tumors, 1.7-7% of gliomas, and 3-5% of gliomas in children.^[3,6] Many of these gliomas have been found to have a propensity to invade the leptomeninges and cause arachnoid hyperplasia. Arachnoid hyperplasia is a meningeal response associated with gliomas of anterior optic

pathway. This response may be so exuberant as to camouflage the tumor itself, resulting in diagnostic dilemma. Rarely, such marked meningeal proliferation has resulted in erroneous diagnosis of meningioma, which was later found to be a glioma on subsequent surgery.^[2] We present a case of glioma of optic nerve, with marked meningeal proliferation, chordoid and chondroid metaplasia, which can prove to be a significant diagnostic pitfall.

CASE REPORT

History and Examination

A 27-year-old male presented with progressive, painless loss of vision in right eye, associated with vomiting and headache for one and a half months, without accompanying loss of consciousness or fever. He had a history of seizures 4 years back, for which he was on antiepileptic drugs. On clinical examination, there was no perception of light in the right eye while left eye vision was normal. Computed tomography (CT) scan revealed a contrast enhancing mass arising from planum sphenoidale with mass effect [Figure 1]. The adjacent pituitary fossa and cavernous sinus were normal.

Surgery

Perioperative findings showed a grayish pink, nonsuckable tumor firmly adherent to the right optic nerve and expanding it. The tumor was also attached to basal dura and falx. Debulking surgery was performed. A clinical impression of an intradural, optic nerve sheath meningioma was made.

Pathology

Histopathological examination of the excised tumor revealed a moderately cellular glial tumor intermixed with extensive reactive proliferation of arachnoid cells. The tumor cells were present in a fibrillary background and exhibited minimal nuclear pleomorphism. No mitosis, necrosis or micro-vascular proliferation was noted. Adjacent to the tumor, there was marked fibroblastic and arachnoid cell proliferation with chondroid as well as chordoid differentiation along with myxoid change and dense collagenisation. The sharply outlined tumor was reticulin poor while the adjacent reactive areas were rich in reticulin fibers. On immunohistochemistry using Dako antibody kit, the tumor cells expressed glial fibrillary acid protein (GFAP). [Figure 2] Epithelial membrane antigen (EMA) highlighted the reactive

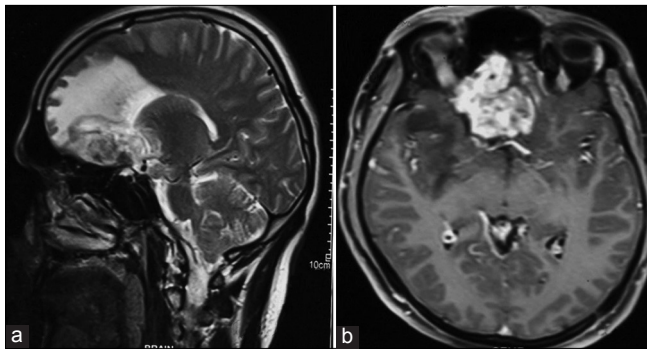


Figure 1: MRI Brain shows a heterogeneously hyperintense mass extending from planum sphenoidale and compressing both frontal lobes on T2WI sagittal images (a) with surrounding edema. The lesion shows heterogeneous postcontrast enhancement and extends to the right frontal sinus and anterior cerebral artery on T1WI image with contrast (b)

arachnoid cells. The chondroid areas were positive for S-100. A final diagnosis of optic glioma displaying exuberant arachnoid proliferation with cartilaginous metaplasia was made.

Treatment and Follow-up

Postoperatively, there was no improvement in the vision. Recurrence of tumor was noted after a period of four months and debulking surgery was repeated. Histopathological examination confirmed the presence of glioma (grade I) having similar morphology as the previous tumor displaying extensive fibro-mesenchymal proliferation of the meninges. The patient is on regular follow-up, however, there is still no improvement in the vision.

DISCUSSION

Optic nerve gliomas account for 1.7-7% of gliomas.^[3,7] Approximately 75% of patients with these tumors present during the first decade of life and 90% become symptomatic before 20 years of age.^[1,5] The presenting symptoms usually include slow unilateral visual loss, proptosis, decreased color vision or symptoms of raised intracranial pressure. Associated neurological symptoms like headache, eye pain, hemiplegia and dementia may be seen in up to one-third of the adult population having malignant optic glioma.^[6,8]

Arachnoidal cell proliferation causing distension of the perineural space is a common observation in optic gliomas and there have been case reports where glioma has been mistaken for meningioma due to such marked

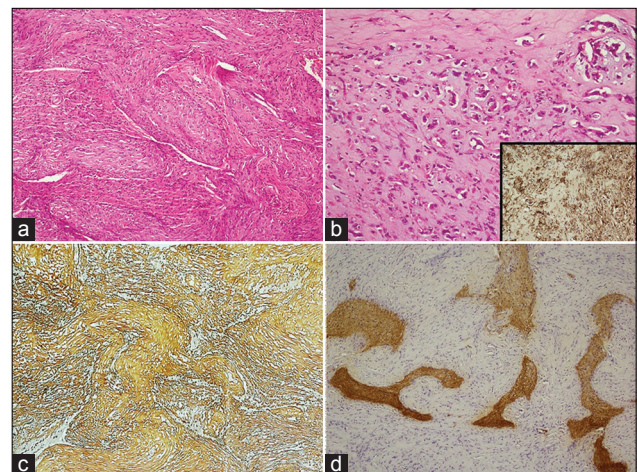


Figure 2: Photomicrograph showing (a) Islands of gliomatous component scattered among areas of marked fibroblastic proliferation (H and E, $\times 100$); (b) Areas of cartilaginous and chordoid metaplasia (H and E, $\times 200$). Inset showing positivity for S-100 in cartilaginous areas; (c) Sharp demarcation between reticulin poor gliomatous component and reticulin rich fibroblastic area (Reticulin stain, $\times 100$); (d) Glioma showing strong positivity for GFAP ($\times 40$)

proliferation of meningotheial cells.^[2] Connective tissue trabeculae of the optic nerve, capillaries within the glioma and supporting tissue of the arachnoid mater also undergo proliferation. Our case displayed exuberant arachnoid cell proliferation with large areas showing cartilaginous differentiation. These cartilaginous areas posed a diagnostic difficulty and were mistaken to be meningioma with chondroid metaplasia or a well differentiated chondrosarcoma. However, no nuclear atypia or increase in mitosis was noted within the chondroid areas. Chordoid meningioma was also considered as a differential diagnosis due to presence of chordoid like areas. Foci of well demarcated gliomatous component (as highlighted by GFAP) interspersed within the hyperplastic arachnoid tissue as well as clinico-radiological correlation helped us to arrive at the correct diagnosis of optic glioma. To the best of our knowledge, such extensive chondroid and chordoid differentiation accompanying meningotheial proliferation has not been described previously in an optic nerve glioma. It is important to distinguish meningeal hyperplasia from optic nerve sheath meningiomas as the latter are potentially lethal tumors associated with orbital recurrences with or without intracranial involvement, even after surgical excision. The arachnoidal proliferation is intradural, whereas most of the meningiomas show invasion through the dura into the surrounding orbital tissues.^[4]

CONCLUSION

Optic nerve gliomas are usually low grade tumors with variable clinical course. Arachnoidal hyperplasia is a common phenomenon associated with optic gliomas. However, chordoid and chondroid metaplasia has not been reported previously. Clinico-radiological consultation, simple reticulin stain and immunohistochemistry can be of great help in such a situation to distinguish these tumors from a chondrosarcoma or chordoid meningioma.

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