Isolated low grade pilocytic astrocytoma of the optic nerve in the elderly: case report

Astrocitoma pilocítico de baixo grau do nervo óptico em adultos: relato de caso

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INTRODUCTION

Gliomas of the optic nerve are uncommon, comprising 1-5% of intracranial astrocytomas and 1.5-3.5% of all orbital tumors. They occur most frequently in children and young adults, 75% of them in the first decade and 90% in the first two decades of life (1-3). They may be confined to the optic disc and nerve (25%) or may involve the chiasma with or without the optic nerves in 20-40% of cases (1-2). We describe here a case of an isolated pilocytic astrocytoma of the optic nerve in an aged adult, which was incompletely excised and recurred as an extensive mass in the orbit.

CASE REPORT

A 68-year-old man presented with a history of a right optic glioma. Eighteen months ago he underwent a lateral orbitotomy at another institution for removal of an optic nerve mass. At that time histology revealed that the tumor was an optic nerve glioma with a pilocytic pattern. No further treatment was instituted and one year after surgery he noticed that his right eye was proptotic again. Magnetic resonance imaging of the orbit showed that his right orbit was almost completely filled with a mass which extended through the optic canal to the chiasma. The tumor was excised by a combined neurosurgical and orbital approach. Histology proved that the neoplasm was a low grade pilocytic astrocytoma of the optic nerve.

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There was no light perception and the intraocular pressure was zero. An MRI of the orbits showed an extensive mass in the right orbit extending through an enlarged optic canal (Figures 1C e D). The patient underwent combined neurosurgical and orbital surgery. The optic nerve was incised through a transcranial approach at the chiasma level and the mass was removed by an eyelid- and conjunctiva-sparing orbital exenteration. The orbit was reconstructed with a temporalis transfer and dermis-fat graft (Figure 2D). Histology showed the same pattern as observed in the previous biopsy, i.e., a low-grade pilocytic astrocytoma of the optic nerve.

**COMMENTS**

Histologically, gliomas are divided into four types: pilocytic astrocytomas (WHO grade I), low-grade diffuse astrocytomas (WHO grade II), anaplastic astrocytomas (WHO grade III), and glioblastomas (WHO grade IV). Pilocytic astrocytomas tend to have a favorable prognosis and usually occur in patients less than 20 years of age with neurofibromatosis type I. In patients older than 40 years of age optic nerve gliomas usually differ from the gliomas of childhood. Most adult tumors are highly infiltrative anaplastic astrocytomas or glioblastomas. These neoplasms tend to be aggressive and fatal. Our patient was an elderly male who presented the low grade pilocytic pattern of glioma usually observed in children. A Medline search revealed only one report of isolated pilocytic astrocytomas of the optic nerve in an aged adult, as described in this case.

Pilocytic astrocytomas of the optic nerve are typically low-grade neoplasms with a favorable long-term prognosis. Despite incomplete surgical resection, some tumors fail to...
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Figure 2 - Histological photographs revealing the optic nerve with neoplastic glial cells, piamater enlargement (asterisk), and Rosenthal fibers (arrow). HEX40 (A); HEX100 (B). Preoperative (C) and postoperative (D) clinical photographs.

recur; suggesting arrest of tumor progression or deceleration of tumor growth\(^5\). In the present case, however, the previous partial excision elicited a rapid recurrence of the tumor in the orbit.

Complete surgical resection is successful in tumors confined to an optic nerve. For disease that has progressed into or beyond the chiasma, total resection is not feasible. Partial resection may provide symptomatic relief and also tissue diagnosis. Radiotherapy (RT) and radiosurgery might be more effective in posterior tumors than surgery alone in preventing relapse, but their morbidity should be taken into consideration. RT alone or after partial resection shows relapse-free rates of 56-90%\(^3\). Long-term prognosis, however, apparently does not change after any adjuvant treatment\(^3\).

In summary, an isolated pilocytic astrocytoma of the optic nerve in the elderly is a rare event. In the present case, although the tumor had a benign histopathological pattern, partial removal was followed by a rapid growth that warranted a combined neurosurgical and orbital approach.

RESUMO

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REFERENCES