Malignant optic pathway glioma

While optic pathway gliomas (OPGs) are a frequent finding in pediatric patients with neurofibromatosis type 1, sporadic malignant optic pathway gliomas (mOPGs) in adult patients are more rare with 72 cases described in the literature. MOPGs are classified as either anaplastic astrocytoma (WHO grade III) or glioblastoma (WHO grade IV). Treatment is based on radiation therapy and chemotherapy with temozolomide. Here we report a highly unusual case of mOPG classified as a pilocytic astrocytoma (WHO grade I).

Patients with malignant optic nerve pathway gliomas had unilateral or bilateral visual dysfunction, which was often accompanied by periorbital discomfort in an otherwise asymptomatic adult. Regardless of origin, the clinical course invariably was bilateral blindness rapidly followed by death. Occasionally, the diagnosis was made preoperatively and was confirmed by surgical exploration and biopsy. [1].

A 71 year-old woman was referred to our institution with progressive concentric, predominantly left-sided, visual field loss for six weeks. Gadolinium-enhanced T1-weighted magnetic resonance tomography (MRI) showed contrast enhancement of the optic pathway that included both optic nerves, the optic chiasm and the left optic tract. Under the initial suspicion of neurosarcoidosis, the patient received immunosuppressive treatment with high-dose corticosteroids for another two weeks without improvement of her symptoms. The subsequent MRI three weeks later showed enlargement of the contrast-enhancing lesions, and the patient developed amaurosis on her left eye. Hence, an open biopsy of the left optic nerve was performed.

Microsurgery was carried out in a standard fashion via pterional craniotomy. The postoperative course was uneventful. In the absence of necrosis or vascular proliferation, with a low mitotic activity, the tumor was classified as pilocytic astrocytoma (WHO grade I) which to some extent contrasted with the observed clinical course. Genetic testing for BRAFV600 mutation showed wild-type status. After discussing treatment options, including radiotherapy and chemotherapy, the patient opted for best supportive care. A follow-up MRI six weeks after the biopsy showed tumor progression and the patient died another six weeks later. An autopsy was declined by the patient´s family.

This case illustrates that mOPGs are a diagnostic and therapeutic challenge. The diagnosis should be established and treatment instituted as soon as reasonably achievable [2].

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Spoor TC, Kennerdell JS, Martinez AJ, Zorub D. Malignant gliomas of the optic nerve pathways. Am J Ophthalmol. 1980 Feb;89(2):284-92. PubMed PMID: 6243868.
2)
Heiland T, Karpel-Massler G, Scheuerle A, Wirtz CR, Halatsch ME. A rare case of sporadic malignant optic pathway glioma in a 71-yearold woman. World Neurosurg. 2019 Jul 20. pii: S1878-8750(19)32014-5. doi: 10.1016/j.wneu.2019.07.108. [Epub ahead of print] PubMed PMID: 31336173.