

SHORT COMMUNICATION

Suprasellar germinoma and late perioptic seeding

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PURPOSE. *To report an unusual case of optic nerve seeding 12 years following treatment of a suprasellar germinoma.*

METHODS. *Observational case report.*

RESULTS. *A 34-year-old woman presented with a 3-week history of subjective right eye visual loss. She had been diagnosed with a suprasellar germinoma at 22 years of age, which had been partially excised and radiated (5400 cGy). The tumor had shown complete radiographic regression without neurologic sequelae. Pertinent findings on current examination included right eye visual acuity of 20/150, right relative afferent papillary defect, and optic nerve pallor in the right eye. In addition, partial left facial paralysis was noted. Examination was otherwise unremarkable including 20/20 acuity in the left eye. Magnetic resonance imaging demonstrated abnormal enhancement and thickening of both optic nerves and along the course of the left V and VII cranial nerves. Serum levels of α -fetoprotein and β -HCG were abnormal, consistent with metastatic germinoma. Following two cycles of chemotherapy (VIP-etoposide, ifosfamide, and cisplatin), visual acuity returned to 20/20 bilaterally, with corresponding radiographic improvement and normal cerebrospinal fluid cytology.*

CONCLUSIONS. *Perioptic subarachnoid seeding may occur over a decade after presumed successful treatment of germinomas, suggesting the importance of lifelong observation. (Eur J Ophthalmol 2008; 18: 159-61)*

KEY WORDS. *Germinoma, Perioptic seeding, Recurrence, Metastasis*

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INTRODUCTION

Germinomas constitute 0.1 to 3.4% of all intracranial neoplasms, the vast majority occurring in midline structures (pineal gland, suprasellar and basal ganglia regions) (1). In recurrent cases, seeding of spinal column is fairly common, but spread to the optic nerve is exceedingly rare (2, 3). In the few described cases, recurrence occurred within a few years of treatment (3). In this report we describe an unusual case with optic nerve seeding 12 years following treatment of a suprasellar germinoma.

Case report

A 34-year-old woman was referred for decreased visual acuity in the right eye and partial left facial paresis. She

had been treated for a suprasellar germinoma at 22 years of age. The primary tumor was identified during an evaluation for hypopituitarism and diabetes insipidus. At that time her ophthalmologic examination was unremarkable. Brain magnetic resonance imaging (MRI) revealed mild expansion and enhancement within the pituitary stalk. MRI of the cervical, thoracic, and lumbar spines was normal. Cerebrospinal fluid (CSF) β -human chorionic gonadotropin (β -HCG) was abnormal (6 UI/L). Serum α -fetoprotein (AFP) and β -HCG levels were normal. Trans-sphenoidal biopsy revealed a germinoma. Following focal radiation therapy (5040 cGy) there was a complete radiographic regression. Whole brain radiation and chemotherapy were not administered. For 10 years annual neuroimaging (MRI) failed to detect recurrence and surveillance was discontinued.

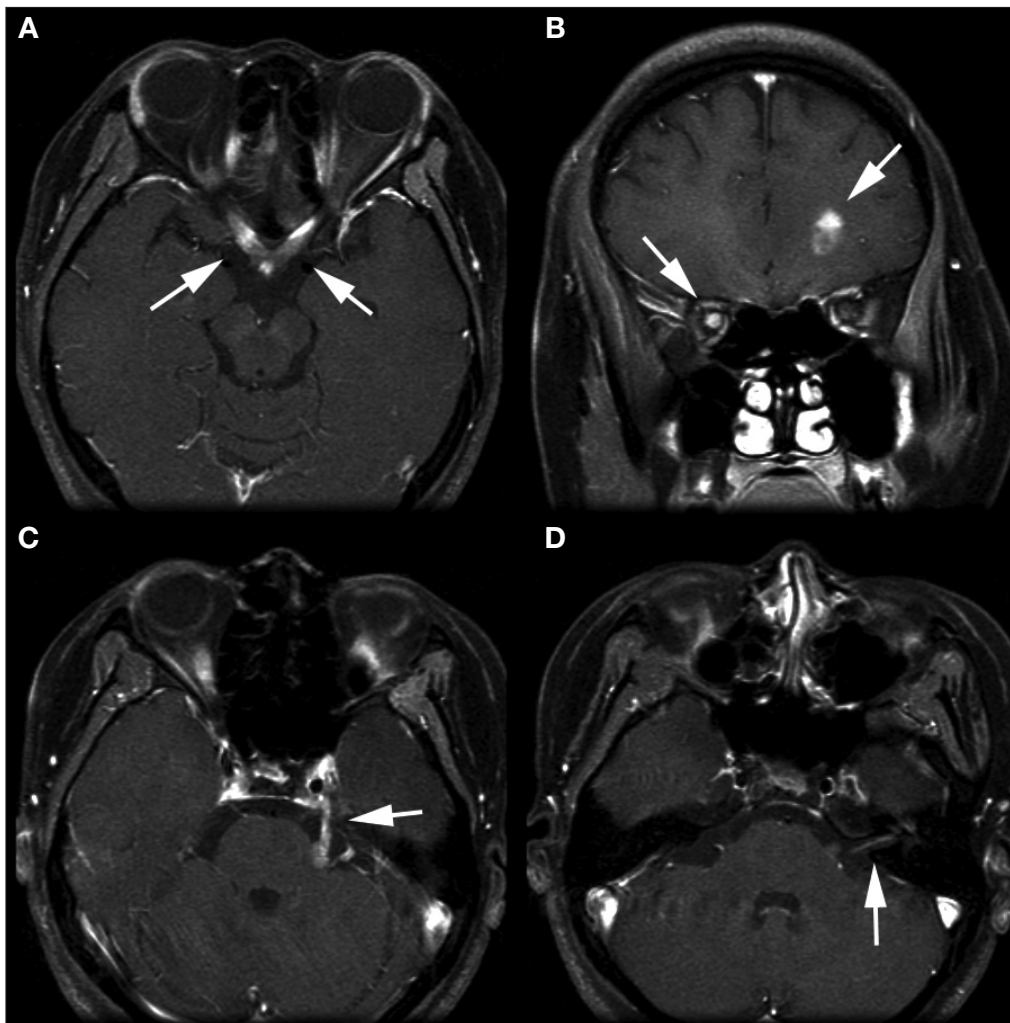


Fig. 1 - Recurrent germinoma with perioptic seeding: gadolinium enhanced T1-weighted magnetic resonance imaging demonstrating infiltration of (A) the intracranial portion of the optic nerves, (B) left frontal lobe and right intra-orbital optic nerve, (C) left trigeminal nerve, and (D) left facial nerve.

Two years later she returned to medical attention with complaints of insidious decline in right eye visual acuity over 3 months, and concurrent left facial weakness. She was found to have a visual acuity of 20/150 in the right eye, a moderate relative afferent pupillary defect, abnormal visual field, and optic disc pallor. Left eye examination was unremarkable. Neurologic evaluation was notable for left facial weakness and was otherwise normal. MRI demonstrated abnormal enhancement and thickening of both optic nerves, despite no visual dysfunction in the left eye (Fig. 1). Additional involved areas included the left facial and trigeminal nerves, left inferior frontal lobe, left lateral ventricle, and a dropped metastasis involving the rostral cauda equina. Serum AFP (9.5 $\mu\text{g/L}$) and $\beta\text{-HCG}$ (179 UI/L) were elevated consistent with recurrent germinoma. Following two cycles of chemotherapy (VIP-etoposide, ifosfamide, and cisplatin), all lesions demonstrated a re-

duction in size, and repeat CSF cytology has been normal. Visual acuity improved to 20/20 bilaterally but her facial paralysis has persisted.

DISCUSSION

Seeding of the perioptic subarachnoid space was first described by Manor et al in 1990 (2). In this case, the optic nerve lesion was identified at the same time as the primary pineal lesion. Nakajima et al reported two cases of metastatic germinomas to the optic nerve, 2 and 3 years following initial diagnosis (3). Although spread within spinal column is common and late recurrences up to 17 years following presumed successful treatment have been described (4), we are unaware of any such cases which involved the optic nerves. Moreover, in all the reported

cases of optic nerve involvement, only slight to moderate improvement of visual function was noticed after treatment. Our patient is unique in that recurrence was not observed until 12 years following initial treatment and significant vision recovery was observed.

The infrequent occurrence of CNS germinoma has resulted in a lack of controlled investigations addressing management and subsequent controversy regarding proper treatment. First line therapy is considered by most to be radiation therapy alone, which has been reported to carry 5-year survival rates between 80 and 100% and 5-year disease-free survival from 70 to 90% (5). Because of the infiltrating and disseminating nature of the disease, whole brain and craniospinal irradiation (CSI) have often been recommended. The intent of primary treatment of entire CSF space is to treat undetected micro foci, the presumed source of recurrent disease. Others reserve this for patients with evidence of CSF dissemination, thus avoiding late irradiation adverse effects. A minority of authors defend chemotherapy as an adjuvant or even as primary therapy (4).

Although the optimal treatment strategy remains uncertain, as demonstrated by this case, lifelong follow-up should be considered as recurrences may occur over a decade after presumed successful treatment. Whether adjunct whole brain radiation would have prevented recurrence in our patient can only be speculated, but cases such as this bolster the argument for primary treatment of the entire CSF space.

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