Oligodendroglioma of the pineal region

Case report

SUNIT DAS, M.D., PH.D., JAMES P. CHANDLER, M.D., ANIA POLLACK, M.D.,
EILEEN H. BIGGIO, M.D., LESLIE DIAZ, M.D., JEFFREY J. RAIZER, M.D.,
AND H. HUNT BATJER, M.D.

Departments of Neurological Surgery, Pathology, and Neurology, Northwestern University, Feinberg School of Medicine, Chicago, Illinois

The authors describe an oligodendroglioma of the pineal region in a 59-year-old woman. The patient presented with intermittent confusion, memory disturbance, and headache associated with a cystic pineal region mass demonstrated on magnetic resonance imaging. Gross-total resection was performed via a suboccipital supratentorial approach. Pathological and genetic evaluation showed the tumor to be an anaplastic oligodendroglioma. Although the spectrum of tumors arising within the region of the pineal gland is broad, to the authors’ knowledge this is the first report of an oligodendroglioma occurring in this area.

KEY WORDS • oligodendroglioma • pineal region • pineal gland • gene deletion

Tumors of the pineal region are a rare but diverse group and include germ cell tumors, pineoblastomas, meningiomas, primitive neuroectodermal tumors, teratomas, and pineocytomas. Gliomas are rarely found in this region, although the literature does contain reports of astrocytomas and mixed oligoastrocytomas of the pineal gland. We report the case of a 59-year-old woman who presented with intermittent confusion, memory disturbance, and headache associated with a cystic pineal region mass demonstrated on MR imaging. Gross-total resection was performed via a suboccipital supratentorial approach. On immunohistochemical and immunocytogenetic evaluation the tumor was found to be positive for glial fibrillary acidic protein, S100, and synaptophysin, with deletion of chromosome arm 1p as well as chromosome 19 polysomy with deletion of 19q, which is consistent with a diagnosis of anaplastic oligodendroglioma. To our knowledge, this is the first report of an oligodendroglioma occurring in the pineal region.

Case Report

History and Examination. This 59-year-old woman was admitted to our hospital with a 5-day history of intermittent confusion, memory disturbance, and headache. The patient’s medical history was notable for thyroid cancer that had been treated with total thyroidectomy followed by neck irradiation 14 years before. Neurological examination failed to reveal any abnormality other than nystagmus on lateral gaze. Admission MR imaging demonstrated a heterogeneous irregularly enhancing cystic lesion in the region of the pineal gland, with compression of the cerebral aqueduct (Fig. 1). Mild ventriculomegaly was noted. Imaging of the remainder of the neuraxis revealed no other lesions. Results of serological testing for \( \alpha \)-fetoprotein and \( \beta \)-human chorionic gonadotropin were negative. Because the patient showed no clinical signs of hydrocephalus, the decision was made to proceed to surgery without performing cerebrospinal fluid diversion.

Operation. The patient was placed in the sitting position. Frameless stereotaxy was used to assist with tumor localization. A suboccipital, supracerebellar approach was used, and it revealed a dark, gelatinous neoplasm lying just behind the tentorial veins. The tumor was initially detached superiorly from the veins of Rosenthal and Galen, then it was rolled off laterally from the tectorial plate. Finally, the inferior aspect of the tumor was slowly detached from the tectorial plate, revealing the third ventricle through the veil of the velum interpositum.

Histological and Immunocytochemical Studies. The intraoperatively frozen section was initially diagnosed as pineoblastoma as opposed to pineocytoma. The permanent section showed a tumor composed of relatively small, densely packed cells containing little cytoplasm, interspersed with moderately sized cells with abundant, eosinophilic cytoplasm (Fig. 2). Nuclei were mildly to marked-

Abbreviations used in this paper: FISH = fluorescence in situ hybridization; MR = magnetic resonance.
ly pleomorphic, and mitoses were frequent. Gross invasion of the pineal gland was noted, and proliferating vasculature was seen. Neither Homer–Wright nor Flexner–Wintersteiner rosettes were observed. The tumor was found to be positive for glial fibrillary acidic protein, S100, and synaptophysin staining. Neurofilament staining was negative. A preliminary diagnosis of a high-grade glioneuronal tumor was assigned; given its complexity, however, the case was sent for outside consultation. After receiving the suggestion that the diagnosis of anaplastic oligodendroglioma be considered, FISH for chromosome arms 1p and 19q was performed. The FISH results demonstrated deletion of 1p, as well as chromosome 19 polysomy with deletion of 19q, which is consistent with a diagnosis of anaplastic oligodendroglioma.

Postoperative Course. Follow-up MR images obtained the day after surgery demonstrated no evidence of residual tumor, with improvement of the patient’s hydrocephalus (Fig. 3). Clinically, the patient was found to have an upward gaze palsy that resolved with administration of high-dose dexamethasone. Neuropsychiatric testing performed in the immediate postoperative period was remarkable for attention dysfunction and a delayed rate of information processing. The patient has since returned to her baseline neurological status. Fractionated external-beam radiation therapy to the tumor bed with a concurrent course of temozolomide was used as an adjuvant therapy. An MR image obtained at her 9-month follow-up visit demonstrated no evidence of recurrent disease.

Discussion

A wide array of tumor subtypes can occur in the pineal region, a feature that complicates the approach to optimal...

![Image of sections cut from different regions of the tumor]

Fig. 2. Photomicrographs of sections cut from different regions of the tumor. A: Section showing tumor infiltration into normal pineal gland tissue. B: Magnified image showing multiple mitotic figures. C: Densely cellular region composed of small cells with generally round nuclei, frequent mitoses, and microvascular proliferation. D: Myxoid region containing numerous cells with intensely eosinophilic cytoplasm reminiscent of microgemistocytes. H & E, original magnifications ×40 (A and D), ×100 (B), and ×20 (C).
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Fig. 3. Gadolinium-enhanced MR image obtained on Day 1 postoperatively, demonstrating gross-total tumor resection.

clinical management. This diversity is a ramification of the normal cell types that reside in the pineal gland and its adjacent structures. The pineal gland is composed of pineal parenchymal cells, astrocytes, and sympathetic neurons. Adjacent to the gland are ependymal cells lining the third ventricle, cells forming the choroid plexus, arachnoid cells forming the velum interpositum, and glial cells from the brainstem and thalamus. Neoplastic transformation in the pineal region can lead to ependymomas, astrocytomas, choroid plexus papillomas, other gliomas, pineal parenchymal tumors, and meningiomas. Additionally, primitive germ cell rests are frequently retained in midline structures such as the pineal gland, resulting in the full complement of germ cell tumor subtypes, of which germinomas are the most common. The pineal region may also be the site of miscellaneous histological subtypes including metastases, neuronal tumors, endothelial tumors, and lymphomas.

To our knowledge, this report is the first dealing with an oligodendroglioma occurring in the pineal region. Our patient presented with a 5-day history of intermittent confusion, amnesia, and headache, and a distant but significant history of neck irradiation for thyroid cancer. Individuals with tumors of the pineal region commonly present with symptoms and signs of increasing intracranial pressure with hydrocephalus, including headache and Parinaud syndrome. It is possible that our patient’s confusion and amnesia were actually a manifestation of the attention deficit noted during postoperative neuropsychiatric evaluation, and possibly a result of diminished transhemispheric processing secondary to her splenial lesion. A transient upward gaze palsy developed in our patient after surgery, probably the result of surgical manipulation of the tectorial plate.

The discrepancy between the initial diagnosis of pineoblastoma as opposed to pineocytoma rendered on frozen section and the final histological finding of anaplastic oligodendroglioma speaks to the difficulty of characterizing tumors of the pineal region. Our report adds another lesion to the wide array of tumors found within this region. The role of FISH analysis was integral to our final diagnosis. It happened that the intraoperative diagnosis of pineoblastoma directed us surgically to pursue the tumor in an aggressive manner; in the end, such an approach served our patient well.

In retrospect, it is feasible that evaluation of the frozen section would have led to a diagnosis of oligodendroglioma if the entity had been considered within the differential for a neoplasm occurring in this region. Regardless, the patient in this case underwent gross-total tumor resection followed by external-beam radiation to the tumor cavity. Analyses of multiple studies have shown the extent of resection to be important for time to tumor progression as well as total survival; in this case, gross-total resection was feasible without concerns about causing neurological impairment. Although the role of adjuvant radiation therapy in the treatment of low-grade oligodendroglioma after gross-total resection is unclear, its use in patients with anaplastic oligodendroglioma does appear to have a beneficial effect on survival.

In the European Organisation for Research and Treatment of Cancer Study, investigators found an objective response rate of 54%, with a significantly lower incidence of toxicity than treatment with procarbazine, lomustine, and vincristine. The efficacy of temozolomide against anaplastic oligodendroglioma was found to be greater in tumors with 1p and 19q deletions. In our patient a functionally equivalent tumor profile was found, characterized by 1p deletion and chromosome 19 polysomy with relative deletion of 19q. After surgery, our patient underwent fractionated external-beam radiation therapy to the tumor bed with a concurrent course of temozolomide. She subsequently received four cycles of temozolomide to complete a 5-month course of therapy. She remains in clinical remission, with no evidence of tumor recurrence on MR images obtained at her 9-month follow-up visit.

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References


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