Metastatic Craniopharyngioma

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Summary: We report a unique case of metastatic craniopharyngioma. Initially, the patient had a right frontal craniotomy for resection of a suprasellar mass, which was determined to be an adamantinomatous craniopharyngioma. Seven years later, an MR study of the brain showed two peripheral enhancing lesions adjacent to the dura and contralateral to the craniotomy site. Pathologic examination again showed adamantinomatous craniopharyngioma. Although recurrence, both local and along surgical tracts due to implantation of craniopharyngioma tissue, has been reported, this case raises the possibility of meningeal seeding to remote sites.

Craniopharyngiomas are complex epithelial tumors arising from remnants of Rathke’s pouch (1, 2). Total resection of a craniopharyngioma may be difficult, and recurrence has been reported in approximately 25% to 70% of patients (3, 4). Recurrence usually occurs at the primary site, although a few cases of ectopic recurrence along the surgical or needle tracts have been reported (5–7). We present a case of metastatic craniopharyngioma that cannot be attributed to local recurrence or to direct implantation by the surgical procedure. To our knowledge, no other cases of metastatic craniopharyngioma have been reported to date.

Case Report

A 73-year-old man initially presented in March 1990 with symptoms of bitemporal hemianopsia. An MR study of the brain (Fig 1A) showed a 2-cm complex partially enhancing suprasellar mass, which elevated and stretched the optic chiasm and compressed the floor of the third ventricle. This mass had a cystic-appearing area of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, with a small, enhancing mural nodule. No other areas of abnormality were detected at this time. The lesion was surgically excised through a right frontal craniotomy, and pathologic examination revealed adamantinomatous craniopharyngioma. Although recurrence, both local and along surgical tracts due to implantation of craniopharyngioma tissue, has been reported, this case raises the possibility of meningeal seeding to remote sites.

In October 1997, the patient presented with a new onset of partial seizures. An MR examination of the brain (Fig 1B) showed two peripheral enhancing lesions adjacent to the dura, in the left parietooccipital and posterior left frontal convexities. There was no recurrent or residual suprasellar mass. At surgery, these masses were found to be adherent to the dura. The lesions were excised, and histopathologic evaluation of each one revealed adamantinomatous craniopharyngioma (Fig 1C). Histologically, these were identical to the suprasellar craniopharyngioma resected in 1990 (Fig 1D). The patient received no further treatment and is doing well 1 year later.

Discussion

Craniopharyngiomas are neoplasms that arise from squamous epithelial rests along remnants of Rathke’s cleft (1, 2). Most commonly arise in the suprasellar region (90%) and account for 1% to 3% of all intracranial neoplasms. More than half of all craniopharyngiomas occur in children and young adults. A second, smaller peak occurs in middle-aged adults. Patients may present clinically with headaches, visual disturbances, and hypothalamic and pituitary dysfunction (1, 3).

On gross pathology, most craniopharyngiomas are cystic masses with cholesterol-rich fluid and calcifications. Histologically, prominent epithelial lobules with palisading of cells at the periphery of the lobules and a stellate appearance of internal cells (so-called stellate reticulum) are seen (1, 2) (Fig 1C and D).

The hallmark of a craniopharyngioma on imaging studies is a suprasellar cyst with calcification and enhancement. CT scans typically reveal a cystic suprasellar mass with some solid component. Nodular or rim calcification is found in about 90% of cases. Attenuation of the cystic component is variable but is often slightly higher than CSF. Nodular or rim enhancement is seen on contrast-enhanced CT studies. On MR imaging, a heterogeneous suprasellar mass with a highly variable appearance is observed, especially on T1-weighted images. A cyst with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images is the most common pattern (8, 9). High signal intensity on T1-weighted images may be seen, owing to high protein content or blood degradation products or both. Heterogeneous enhancement of the solid component has been noted on contrast-enhanced images (8–11).

Despite benign histopathologic features, craniopharyngiomas are locally invasive, with projections of tumor extending into the adjacent brain (2). Hence, total resection of this tumor is often difficult, and local recurrence at the primary site or in the contiguous brain may ensue, usually within 5 years (3, 4). A few cases of ectopic recurrence di-
rectly related to previous surgical procedures have been reported. These include frontal lobe implantation of craniopharyngioma along needle tracts following repeated suprasellar aspirations, sylvian fissure implantation directly adjacent to the surgical site, and craniopharyngioma recurring in the epidural space contiguous to the craniotomy site (5–7).

Conclusion

Our case represents a unique scenario of multiple ectopic dural-based metastatic foci of craniopharyngiomas remote from the primary site and contralateral to the craniotomy site, suggesting the possibility of meningeal seeding. Case reports of meningeal spread from other benign intracranial tumors, including meningioma, pituitary adenoma, choroid plexus papilloma, and central neurocytoma, have been reported (12–15); however, CSF seeding or distant metastases of craniopharyngiomas have not been reported to date. Long-term follow-up studies with MR imaging may cause this unique mode of tumor spread to be recognized in the future.

References