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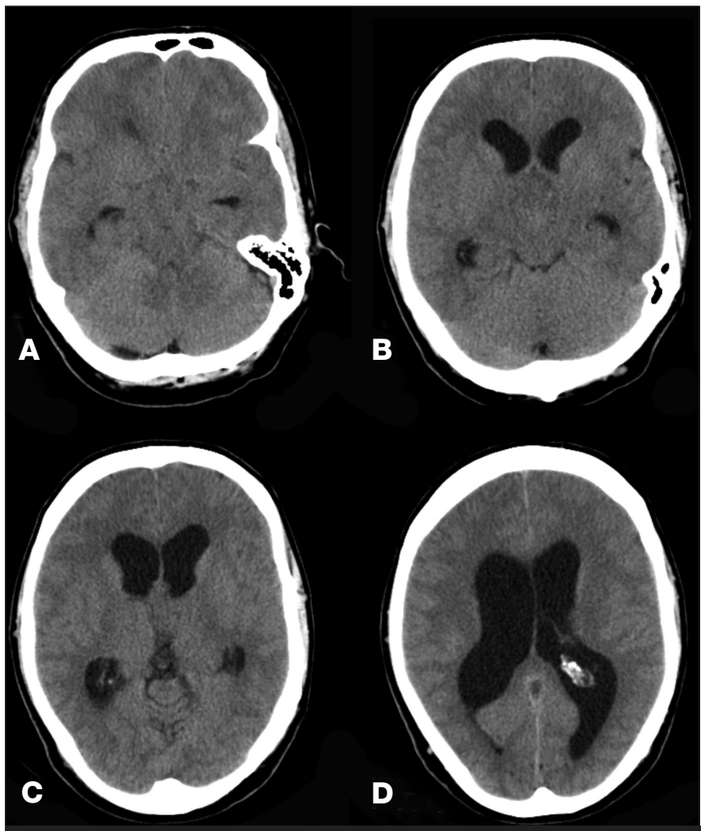

Figure 1

Cranial CT upon initial presentation, demonstrating an isodense third ventricular mass with lateral ventricular dilatation

Abstract

The use of radiation treatment (RT) is usually reserved for residual or recurrent craniopharyngiomas, and the role of RT alone and not as an adjunctive therapy to surgery has not been clearly defined. The authors describe a case of a 50-year-old man presenting with a large suprasellar craniopharyngioma with extension into the third ventricle, producing acute hydrocephalus. A ventriculoperitoneal shunt was performed concurrently with an endoscopic biopsy. Treatment with fractionated stereotactic radiosurgery (FSR) resulted in near resolution of the lesion with no evidence of recurrence over six years. A review of RT for the treatment of craniopharyngiomas without surgical resection is performed.

Introduction

Craniopharyngiomas are histologically benign extraaxial epithelial tumors that arise form embryologic remnants of Rathke's pouch.[12] These rare lesions have an estimated incidence of 1.5 per million people per year, but comprise 10-15% of all pediatric brain tumors.[7,21] Despite their benign histology, craniopharyngiomas cause significant morbidity from damage to the hypothalamus, optic apparatus, and endocrine system. Aggressive treatment is advocated, but the optimal treatment is often debated.

Radical resection is often utilized as a first line treatment due to the frequently large size of these lesions at presentation and associated mass effect.[6,24] Such surgery, however, can carry a high risk of morbidity with hypothalamic and endocrine dysfunction.[26] For this reason, many favor subtotal resection with preservation of adjacent anatomical structures and adjuvant therapies for residual tumor.[11,18] The use of radiotherapy in isolation for the treatment of craniopharyngiomas is infrequent.

Case Report

A 50-year-old man suffering from two months of headache and neck pain presented to the emergency department with a dramatic deterioration of his vision, limb paresis, and seizures. Cranial imaging demonstrated a 3.7 x 2.5 X 3.2 cm, solid suprasellar mass with extension into the third ventricle, producing acute hydrocephalus (Figures 1 and 2). Through a right frontal burrhole, placement of a ventriculoperitoneal shunt was performed concurrently with an endoscopic biopsy of the third ventricular mass tumor (Figure 3). Intraoperatively, a yellow-colored frond-like mass with a consistency similar to choroid plexus was seen filling the right foramen of Monro. Pathology was consistent with a papillary craniopharyngioma.

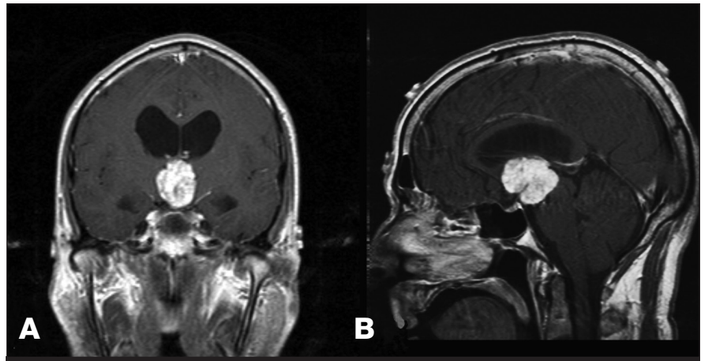

Figure 2

(A) Coronal and (B) sagittal cranial T1WI MRI with gadolinium showing an enhancing suprasellar mass consistent with craniopharyngioma

The patient was subsequently treated with fractionated stereotactic radiosurgery (FSR) for a total of 54 Gy to the 88% isodose line in thirty 1.8 Gy fractions. Within a month of FSR completion, the tumor volume was reduced by nearly half and continued to diminish on each following imaging study. With six years of follow-up, the lesion continues to demonstrate near resolution with no recurrence and further treatment has not been necessary (Figure 4).

Discussion

The treatment of craniopharyngiomas is highly controversial. This controversy is further fueled by the myriad therapeutic modalities available: cystic drainage, intracavitary chemotherapy, limited resection or gross total resection (GTR), and radiation therapy. The greatest debate exists between those favoring radical surgical excision and those believing that subtotal resection followed by adjuvant therapy is best to spare the potential morbidity associated with aggressive surgery. While criticisms of surgical treatment are largely based on the results of open approaches, the role of endoscopic endonasal resection in limiting that morbidity is unclear (Figure 5).

Radiotherapy is most often used as an adjunctive treatment for craniopharyngiomas, either in the setting of residual tumor after a subtotal resection or for tumor recurrence. Published series report a local control rate of 79-95% in these patients.[2,13,14,18,19,22,23] In a review of their craniopharyngioma patients with long-term follow-up, Karavitaki et al. divided 121 patients into four groups: 1) GTR, 2) GTR plus RT, 3) sub-total resection (STR), and 4) STR plus RT. In this cohort, the ten year recurrence-free survival rates were 100%, 100%, 38%, and 77%, respectively.11 These results have been reproduced in the literature examining the newer techniques of fractionated radiation therapy. Typically, 45-55 Gy in 1.8-2.0 Gy fractions are utilized and ten year local control rates for surgery with postoperative FSR are 57-89% compared to 31-42% with surgery alone.[25]

The effect of radiotherapy for craniopharyngiomas appears to be affected by the consistency of the lesion with more solid tumors having the highest average control rate of 90%. Tumors that are either cystic or mixed have rates of 88% and 60%, respectively.[25] The impact of histology on radiation effect is somewhat less clear. While some groups have not found a significant difference between adamantinomatous and papillary craniopharyngiomas, Inoue et al. did find a better response in the latter.[4,10,15]

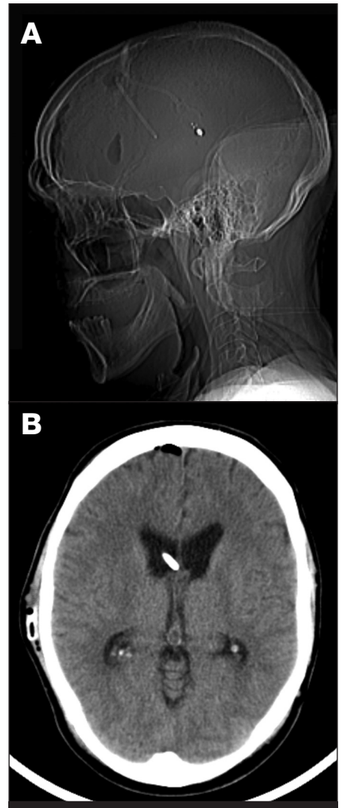

Figure 3

Cranial CT following ventriculoperitoneal shunt placement and endoscopic biopsy

Most reports on radiotherapy discuss craniopharyngioma stability and local tumor control. Few studies describe the effect on tumor size. Significant lesion reduction has been documented after fractionated external radiation[3,8] and stereotactic radiosurgery.[27] In our reported patient, we observed significant tumor shrinkage within one month and this effect has been sustained currently for six years, demonstrating FSR as a possible alternative for initial therapy in select cases. Despite this result, we continue to utilize and recommend surgery as the first line treatment for craniopharyngiomas. We reserve FSR for patients with a significant residual tumor residual or with recurrence. Despite its appeal as a

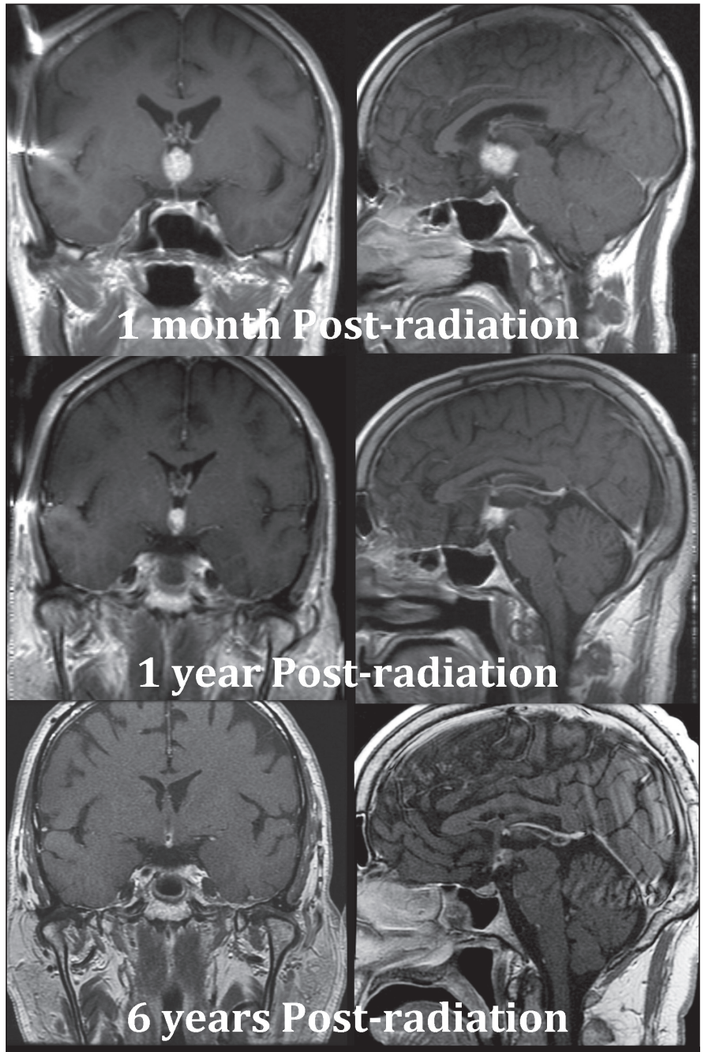

Figure 4

Sequential coronal and sagittal T1WI MRIs with gadolinium demonstrating progressive tumor regression at 1 month, 1 year, and 6 years after radiation

"less invasive" treatment, radiation is not a benign treatment. It can result in growth hormone deficit in pediatric patients, neurocognitive impairment, and optic neuropathy.[9,17,20] Radiation effects to the hypothalamic-pituitary-adrenal axis may produce diabetes insipidus, panhypopituitarism, hypogonadism, hypothalamic obesity, and sleep disturbance.[1,5,16]

Conclusions

While surgery remains the initial treatment of choice for craniopharyngiomas, the debate between GTR versus STR and even open versus endoscopic endonasal approaches continues to persist. Radiotherapy is often reserved for residual or recurrent disease, but we have been able to show that, when necessary, FSR can be effective as an initial treatment. After establishment of a tissue diagnosis, solid papillary craniopharyngiomas may be the most suitable for radiotherapy if surgical resection is not feasible.

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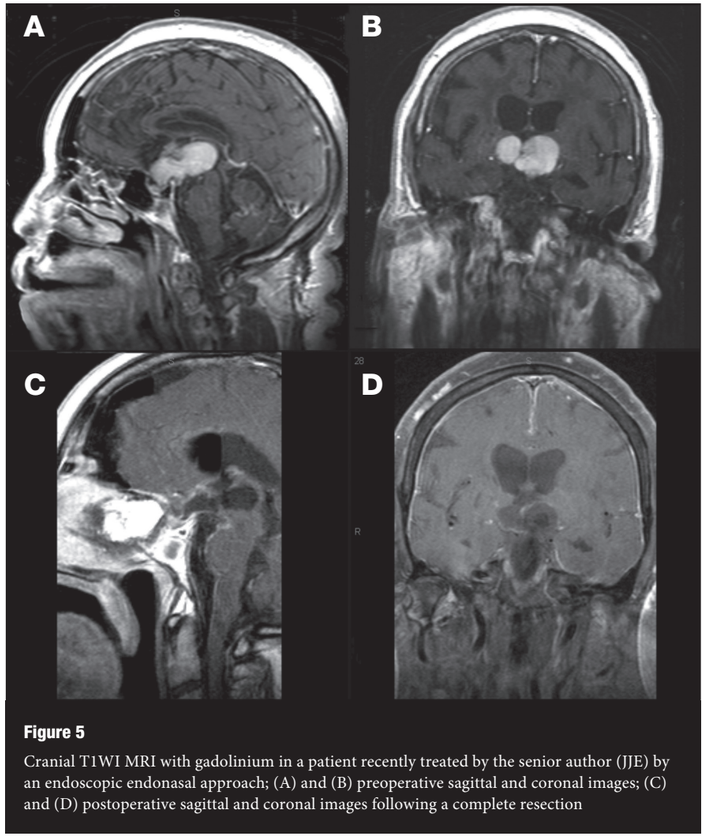

Figure 5

Cranial T1WI MRI with gadolinium in a patient recently treated by the senior author (JJE) by an endoscopic endonasal approach; (A) and (B) preoperative sagittal and coronal images; (C) and (D) postoperative sagittal and coronal images following a complete resection

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