Ectopic Recurrence of Craniopharyngioma—Case Report—

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Abstract

A 27-year-old woman presented with ectopic recurrence manifesting as an asymptomatic mass in the left frontal convexity 5 years after subtotal removal of suprasellar craniopharyngioma through a right orbitozygomatic craniotomy. The mass had enlarged gradually over the next 5 years, so a second operation was performed for total removal. The histological diagnoses of both lesions were adamantinomatous type craniopharyngioma with MIB-1 indexes of 4.2% and 7.4%. The second lesion probably resulted from dissemination. Craniopharyngioma is a benign tumor, and ectopic recurrence is rare. Long-term clinical and neuroimaging follow up is recommended.

Key words: craniopharyngioma, ectopic recurrence, dissemination

Introduction

Craniopharyngioma is a benign tumor arising from the persistent fetal craniopharyngeal duct, and accounts for 2% to 3% of primary intracranial neoplasms.15 Squamous cells present in the infundibulum of the hypophyseal stalk are another potential origin for craniopharyngioma, which may be associated with squamous metaplasia of the pituitary cells.15 Craniopharyngioma tends to recur, often at the primary site and contiguous areas, despite the benign histological character. Ectopic recurrence is very rare with only 14 reported cases.1–11,13,14,16 Here we report a case of craniopharyngioma which resulted in long-term ectopic recurrence.

Case Report

A 17-year-old woman presented with symptoms of amennorhea and right temporal hemianopsia in March 1995. Neurological examination also found minor right visual disturbance. Her medical and family history was noncontributory. Magnetic resonance (MR) imaging showed calcification within a heterogeneous enhanced mass with a cystic component in the suprasellar area (Fig. 1A). The preoperative diagnosis was craniopharyngioma.

The lesion was surgically removed through a right orbitozygomatic approach on July 14, 1995. Postoperative MR imaging showed near total removal except for the tumor adhering to the pituitary stalk (Fig. 1B). Histological examination revealed adamantinomatous craniopharyngioma (Fig. 2A). The MIB-1 labeling index was 4.2% (Fig. 2B), but adjuvant therapy was not performed. Postoperatively, the patient’s visual acuity and field had improved, but postoperative complications such as diabetes insipidus developed. She was discharged with desmopressin (DDAVP) medication.

Fig. 1 Sagittal T1-weighted magnetic resonance images showing (A) a heterogeneous enhanced suprasellar mass after administration of gadolinium, and (B) near total removal of the mass except for the adhesion with the pituitary stalk.
She continued in good condition, but re-enhancement at the stalk was detected at 5 years after surgery, and stereotactic radiotherapy (30 Gy) of the stalk was performed in five sessions. At the same time, MR imaging detected a left frontal mass (Fig. 3A). The mass gradually enlarged over the next 5 years (Fig. 3B), so elective left frontotemporal craniotomy was performed for diagnosis and treatment.

She was admitted on January 18, 2005. The resected mass contained a partly cystic component in elastic soft tumor. The tumor was mostly extra-axial and was not attached to the dura mater. The histological diagnosis was adamantinomatous type craniopharyngioma, as found previously (Fig. 4A). Reactive astrocytes were present at the boundary with the brain surface, but no invasion was seen.

The MIB-1 labeling index was 7.4% (Fig. 4B). The patient was discharged in good condition.

**Discussion**

Ectopic recurrence of craniopharyngioma is very rare, with only 15 reported cases including our case. The nine male and six female patients were aged 2 to 73 years (mean 36.3 years). The primary craniopharyngioma was located in the suprasellar region in all cases. The initial operation achieved total or subtotal removal in all cases except one. Craniotomy was used in all cases. Trans-sphenoidal or trans-nasal approaches were not used, possibly because there was little cerebrospinal fluid space for access. Radiation therapy was performed in two cases.

The time between the initial operation and ectopic recurrence ranged from 6 months to 21 years (mean 4.9 years). The most common region of ectopic recurrence was the frontal lobe convexity in nine cases, with one case in the epidural space of the frontal region, and one case in the intradural extramedullary region of the lumbar spinal cord.

The critical pattern of ectopic recurrence occurred along the surgical route and Ommaya tube placed at the initial operation in 11 cases. Only four cases appeared to be dissemination, as in our case. The ectopic recurrence occurred ipsilateral to the tumor but remote from the craniotomy in one case, in the intradural extramedullary region of the lumbar spinal cord in one case, and contralateral to the initial surgical route in two cases including ours.

The histological diagnosis was the adamantinomatous type in seven of the eight reported cases. The MIB-1 labeling index of craniopharyngioma with local recurrence is higher in the presence of shifts and changes, and the border is 7%. The MIB-1 labeling index of ectopic recurrent craniopharyngioma...
pharyngioma is unknown, but the second specimen was 7.4% in our case, which mandates careful follow up.

Ectopic recurrence of craniopharyngioma may develop from the transplantation of tumor cells along the previous surgical route, as well as dissemination. Craniopharyngioma is a benign tumor but ectopic recurrence may occur even after total removal, so long-term clinical and neuroimaging follow up is recommended.

References


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