Papillary Craniopharyngioma: Redefining the Role of Surgical Intervention

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ABSTRACT

A 75-year-old female presented with worsening bitemporal vision loss, normal pituitary function, and a solid-cystic suprasellar mass lacking calcification suspicious for papillary craniopharyngioma (PCP).

Because recent clinical series in the treatment of PCP have demonstrated excellent response to BRAF/MEK inhibition, a transtubercular transsellar endoscopic endonasal approach (EEA) for tissue diagnosis and neurovascular decompression only was performed.

Intraoperatively, the cyst was decompressed, and the tumor nodule was biopsied for intraoperative pathology. Pathology confirmed the PCP diagnosis, so the surgery was stopped. Postoperatively, she retained normal pituitary function, while targeted therapy is reserved for recurrence and radiation avoided.

INTRODUCTION

Management of craniopharyngioma (CP) centers on maximal safe resection, often combined with stereotactic or proton beam radiotherapy, though aggressive gross total resection carries a high risk of panhypopituitarism.[1]

Genetic studies show that ~95% of papillary craniopharyngiomas harbor the BRAF V600E mutation.[2] In addition, a recent trial of neoadjuvant BRAF/MEK inhibitors demonstrated a 94% partial or complete volumetric response after four 28-day cycles with favorable tumor control.[3] However, 50% of patients still received adjuvant radiotherapy, which increases long-term pituitary dysfunction and affects quality of life.[4]

These findings support shifting surgical goals toward confirming diagnosis and decompressing neural structures when PCP is suspected, leveraging targeted therapy for recurrence to potentially avoid radiotherapy and preserve endocrine function.[5]

METHODS

The authors reviewed a case of a single patient who underwent EEA with ultrasound guidance for resection of a suspected papillary craniopharyngioma. Consent was obtained for surgical intervention.

CASE

We present the case of a 75-year-old female with 9 months of worsening bitemporal vision loss. An MRI revealed a 12.7 x 11.4 x 11.0 mm suprasellar solid-cystic lesion with a small nodular component.

The lesion was suspicious for a papillary craniopharyngioma due to the lack of calcification on CT and MRI. Other than mild bitemporal vision loss on visual field testing, the patient's neurological exam and neuroendocrine lab evaluation were unremarkable. She underwent transtubercular transsellar EEA for cyst decompression and lesion biopsy.

Postoperative course was unremarkable. The patient's vision normalized, and she retained baseline endocrinologic function. Pathology was positive for BRAF V600E (+) papillary craniopharyngioma.

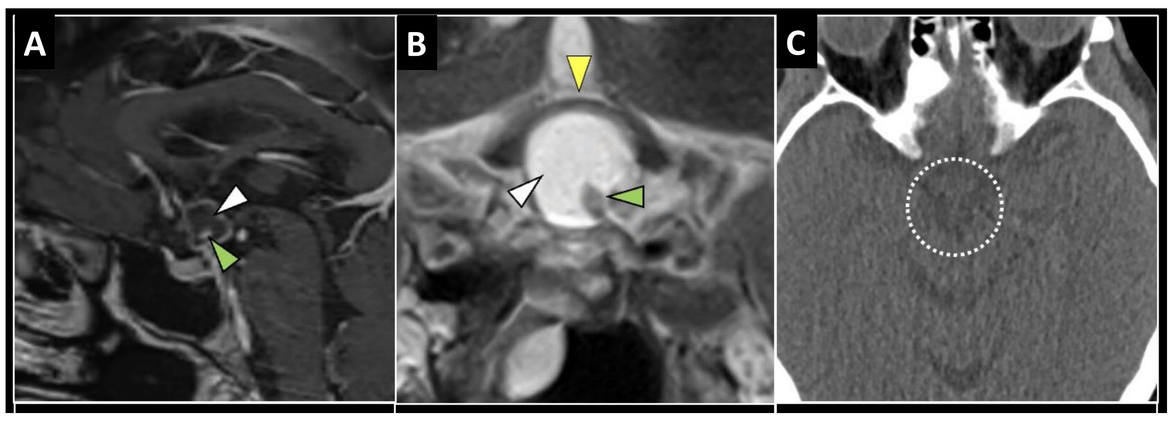

Figure 1: Preoperative Sagittal T1 C+ (A), Coronal T2 (B), Axial CT (C) demonstrating a solid-cystic mass (white and green arrowheads) extending to the base of the 3rd ventricle, suspicious for craniopharyngioma. The lesion caused compression and displacement of the optic chiasm (yellow arrowhead). Lack of calcifications on CT was suggestive of papillary craniopharyngioma

OPERATIVE DETAILS

Following exposure, the sellar dura was opened. Adhesions were dissected sharply to access the suprasellar space. After dividing the superior intercavernous sinus, the arachnoid was opened to expose the pituitary stalk and cyst wall. An incision at the infundibulum–cyst junction allowed entry into the cyst, where a posterior solid nodule was identified, dissected free, and sent for pathology. Ultrasound and inspection revealed residual adherent membrane and a small nodule. Frozen pathology confirmed papillary craniopharyngioma. Given the dense adherence to adjacent neural structures and the availability of targeted therapy if progression occurs, further resection was deferred to minimize neurologic and pituitary morbidity.

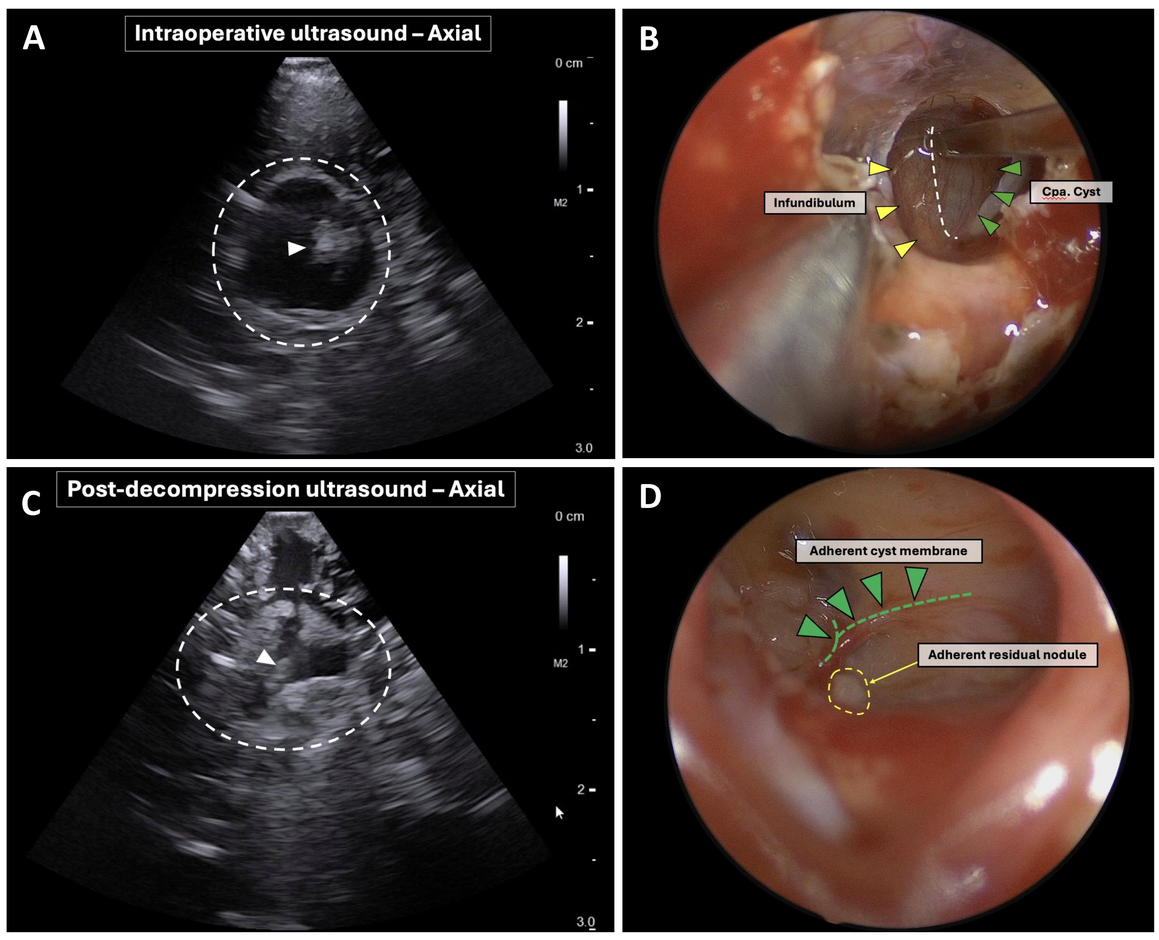

Figure 2: Intraoperative ultrasound view of nodular-cystic lesion pre-decompression (A) and post-decompression (C). A small nodular residual was left behind due to adherence (white arrowhead). Intraoperative endoscopic view of the infundibular incision (B) and post-decompression view (D).

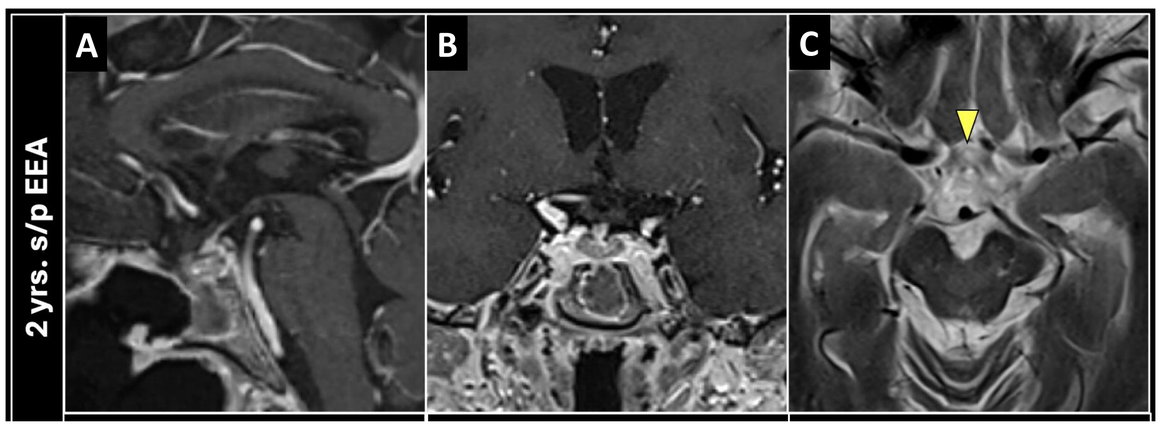

Figure 3: 2-year postoperative sagittal (A) and coronal (B) T1 C+ MRI. Postoperative axial T2 (C) demonstrates decompression of the optic chiasm (yellow arrowhead).

CONCLUSIONS

Targeted inhibitor therapies are reshaping the management of papillary craniopharyngioma and redefining surgical goals. The traditional approach of maximal resection with adjuvant radiotherapy carries a high risk of panhypopituitarism, whereas recognition of a targetable genetic profile supports a more conservative strategy aimed at minimizing neurologic and endocrine morbidity.[5]

This model, however, depends on reliable responses to therapy recurrent cases and requires consideration of BRAF/MEK inhibitor toxicities and patient-specific factors.[3] For these reasons, we favor reserving targeted therapy for progression or recurrence rather than routine adjuvant use, as in our case.

Furthermore, our experience also suggests that adjuvant radiotherapy may not be necessary to optimize progression-free survival in papillary craniopharyngioma, unlike in adamantinomatous craniopharyngioma. Prospective studies are needed to define optimal treatment duration and the value of single versus combination regimens.

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