Craniopharyngioma Type 0: A new addition to the tumor classification scheme

Introduction
Craniopharyngiomas are challenging tumors that often require multimodal treatments due to their recurrence rate. As such, clinicians caring for patients with these lesions need multiple strategies for their management. In order to help formulate a surgical strategy, Kassam et al. published a seminal paper in 2008 describing a classification system for craniopharyngiomas based upon their location relative to the infundibulum. This proposed scheme organized these neoplasms into three categories: pre-infundibular (Type I), trans-infundibular (Type II), and retro-infundibular (Type III), and isolated third ventricular or optic recess tumors (Type IV). In addition, the surgical corridor and the limitations of the endoscopic, endonasal approach (EEA) are discussed. Because craniopharyngiomas arising purely from the sphenoid sinus were not elaborated upon in that publication, the authors describe their experience with this specific subset of tumors and propose an addition to the classification system.

Materials and Methods
The authors retrospectively reviewed the clinical presentation, radiographic features, intraoperative findings, and pathologic diagnosis for craniopharyngiomas. The authors have obtained an IRB to review this data.

Results
The patients who obtained a final pathologic diagnosis of craniopharyngioma with an epicenter inside the sella were identified. Three patients met this criteria. The patient and tumor characteristics are described as separate cases and the patients’ endocrine function are noted as well in order to reveal any salient patterns that can be seen between cases. Because of their location, these tumors were entirely managed with EEA.

Case #1
A 15 year old male presented with a recurrence of his craniopharyngioma which was initially diagnosed 4 years prior. Initially, he was treated with two craniotomies with adjuvant cyberknife radiation. On routine, follow-up imaging, he was found to have a recurrence of his tumor confined entirely to the sphenoid sinus. B. Post-op MRI showing excision of pathology proven craniopharyngioma with enhancement of the stalk and possibly the pituitary gland.

Case #2
A 20 year old male presented with a chief complaint of headache and vision changes with associated optic atrophy. Imaging revealed a heterogenous sellar/suprasellar mass with a significant sellar component. He underwent an EEA and no CSF leak was encountered. He presented with hypopituitarism but has not required testosterone or steroid replacement.

Case #3
A 16 year old female presented with history of headache, nausea, vomiting and visual field cuts. Imaging revealed a solid and cystic tumor arising from the sella, expanding the sphenoid sinus and extending into the suprasellar area. She underwent two EEs due to recurrence of her disease in her right lateral sellar compartment. She required hydrocortisone, desmopressin, levodopa, and estradiol replacement pre-operatively.

Discussion
Craniopharyngiomas arise from squamous cell rests of the hypophyseal-pharyngeal duct in the infundibulum and pituitary gland. The cases we have described suggest a subdiaphragmatic origin to a subset of these tumors that expand the sella and have a large component expanding into the sphenoid sinus; much like giant adenomas, these tumors can invade the suprasellar space as well. In these cases, because the diaphragma is intact, no high-flow CSF leak was encountered and a nasoseptal flap was not needed for reconstruction.

Conclusion
The authors believe these tumors, which will be referred as Type 0 craniopharyngiomas, should be approached in a similar fashion to adenomas or Rathke cleft cysts. An endoscopic, endonasal transellar approach is generally sufficient for the surgical management of these tumors.

Selected References