Extracranial Infraselar Ectopic Craniopharyngioma: A Case Report and Review of the Literature

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ABSTRACT

We present a case of a purely infraselar craniopharyngioma that initially presented as a sphenoid sinus mass. Craniopharyngiomas are usually located within the sella. Purely infraselar craniopharyngiomas have only rarely been reported in the literature. A 25-year-old woman presented with 6-month history of progressive headaches. Initial neuroimaging revealed the presence of a sphenoid sinus mass. Initially, she underwent an endoscopic biopsy of the mass by our ENT service. Pathology was consistent with craniopharyngioma and she was referred to neurosurgery for further surgical management. She then underwent an endoscopic transsphenoidal approach for complete resection of the purely extracranial, infraselar craniopharyngioma. The Rathke pouch arises from the roof of the primitive mouth and grows toward the brain at the fourth week of gestation. Normally, it loses its attachment with the stomadeum completely by the eighth week of gestation. The craniopharyngeal canal (CPC) extends from the floor of the sella to the vomer and may rarely give rise to ectopic craniopharyngiomas. This case shows that such ectopic tumors may arise anywhere along the CPC. Endoscopic endonasal approach provides an excellent route for the resection of these tumors.

KEYWORDS: Craniopharyngioma, ectopic, Rathke pouch, infraselar

Craniopharyngiomas comprise 6% of all intracranial tumors in childhood. The typical tumor is a mixed solid cystic spherical mass, which often has outpouchings which may invade the surrounding hypothalamus.¹ Symptoms vary from visual dysfunction (unilateral or bilateral, field cuts or acuity deficits) to pituitary insufficiency (dwarfism, anorexia, diabetes insipidus, amenorrhea, etc.) depending on the size/location of the tumor and the age of the patient. Symptoms of increased intracranial pressure often occur late in the

disease course, usually due to obstructive hydrocephalus from a tumor with intraventricular extension. There is a bimodal peak age distribution with the initial peak being in childhood and the second peak in the fourth and fifth decade of life.²

Craniopharyngiomas usually occupy the supra and intraselar areas. Only 10% only occupy the intraselar and 20% are purely suprasellar.³ They are usually located above the pituitary gland and the chiasm and within the suprasellar space (combined sellar/suprasellar

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Figure 1 CT scan of the sphenoid sinus, showing the intact sella floor. The heterogeneous mass is located in the sphenoid sinus, posterior ethmoid air cells and beneath the clivus. (A) Sagittal view. (B) Axial view.

type). Ectopic craniopharyngioma have been reported in the third ventricle,⁴ nasopharynx,¹ and cerebellopontine angle.⁵ Purely extracranial infrasellar craniopharyngiomas have only rarely been reported in the literature.^{1,6-8}

CASE REPORT

Our patient is a 25-year-old African-American woman referred to us from our otolaryngology clinic. The patient initially presented to the ENT clinic due to headache without any neurological deficit. Computed tomography (CT) scan showed a heterogeneous mass involving the sphenoid sinus, posterior ethmoid air cells, and the space just anterior to the clivus measuring 27×52 mm (Fig. 1). Magnetic resonance imaging (MRI) scan revealed a mixed solid cystic lesion with

the presence of some contrast enhancement (Fig. 2). The sellar region and the pituitary gland appeared normal on both CT and MRI. Initially, sinonasal neoplasm or fungal sinusitis was suspected and the patient underwent an ethmoidectomy and sphenoidotomy for biopsy of the lesion by the ear, nose, and throat service. Surprisingly, the pathology was consistent with craniopharyngioma (Fig. 3).

Given the unexpected tissue diagnosis in this region, the patient was subsequently referred to neurosurgery for definitive surgical management of this lesion. Follow-up MRI demonstrated mild enlargement of the mass (Fig. 4), but still revealed a normal appearing sella/suprasellar region. We then performed an endoscopic endonasal transsphenoidal resection of the mass. Pathology was again confirmed to be craniopharyngioma.

Figure 2 MRI scan with contrast of the head, showing the partially enhanced craniopharyngioma in the sphenoid sinus, posterior ethmoid air cells. (A) Sagittal view. (B) Axial view.

Figure 3 Histopathologic features of the tumor: (A) Sheets of wet keratins with calcification (black arrows). (B) Whorls of wet keratins separated by fibroblastic stroma and multinucleated giant cells (white arrows) around the keratin whorls. (C) Cholesterol granuloma. (D) Basophilic squamous tumor cells with delicate trabeculae and peripheral palisading. (Original magnifications: 200 $\times$).

Figure 4 MRI scan with contrast of the head sagittal view, showing the increased size of the tumor following the first operation.

Intraoperatively, no breach was noted in either the sellar floor or the anterior clivus. Postoperative MRI demonstrated complete resection of the tumor (Fig. 5). The patient did well postoperatively with improvement in headaches and no new symptoms.

DISCUSSION

Craniopharyngiomas comprise 3% of all intracranial neoplasms and are mainly found in the first two decades of life, though a second peak occurs in the fourth and fifth decades of life.⁹ They are composed of epithelial cells arising from the cell nests at the anterior surface of the pituitary or beneath the capsule of the anterior lobe. Ninety percent of these tumors arise in the sellar/suprasellar area and then extend to the anterior (2 to 5%), middle (2%) or posterior fossa (1 to 4%).^{9–11} Craniopharyngiomas consist of a stratified epithelium that gradually enlarges into a mixed solid cystic mass that may be partially calcified.¹²

Figure 5 MRI scan with contrast of the head sagittal view, showing the complete resection of the mass with intact sella floor and the clivus.

The pituitary gland develops both from the ectoderm of the stomadeum and the neuroectoderm of the diencephalon. The Rathke pouch arises from the roof of the primitive mouth and grows toward the brain during the fourth week of gestation. The stalk of the Rathke pouch lies in the chondrification centers of the presphenoid and basisphenoid bones. It typically loses its attachment with the stomadeum completely by the eighth week of gestation. However, a remnant may persist and give rise to the pharyngeal hypophysis at the roof of the nasopharynx. The most common location for this vestigial structure, called the craniopharyngeal canal (CPC), is the vomerosphenoid junction. The craniopharyngeal canal extends from the floor of the sella to the vomer and may give rise to ectopic craniopharyngiomas.⁷

The persistent CPC has been classified¹³ into two groups based on size: large (several millimeters in diameter) and small (diameter less than 1.5 mm). The former type, also called transsphenoid canal, differs from the latter not only in its size but also in its propensity to be associated with craniofacial anomalies such as a transsphenoidal meningoencephalocele. The small persistent CPC, as in this case, is seldom seen on skull films because of its size, but is more easily detected on CT with bone windows. Anatomically, the canal runs from the anterior part of the sella turcica to the junction of the soft palate and nasal septum. The frequency of a small persistent CPC in adults is up to 0.42%.¹⁴ The vast majority of these are asymptomatic and never are the source of neoplasia.

Though most people believe that the CPC is the remnant of Rathke pouch, Arey¹⁴ suggested that it represents the remnant of a vascular channel formed during osteogenesis. However, review of the literature

revealed that a persistent CPC has been observed in cases of sphenoid teratoma¹⁵ and ectopic pituitary gland,¹⁶ which probably originated from Rathke pouch. It therefore seems more likely that the CPC is in fact a remnant of Rathke pouch.

Purely extracranial infrasellar craniopharyngiomas have rarely been reported in the literature (Table 1).^{1,6-8} The majority of such cases presents in the third decade of life without any sex predilection. On the other hand, the suprasellar tumors show a slight male predominance and they usually present under the age of 20.⁹ The infrasellar tumors are less likely to demonstrate calcification and contrast enhancement as compared with the sellar/suprasellar tumors.

Drummond,¹⁷ Cooper and Ransohoff,¹⁸ Prasad and Kwi¹⁹ and Podoshin et al²⁰ reported several cases of extracranial craniopharyngioma. However, all of these demonstrated some intracranial component. This made determination of a definitive point of origin difficult. It has been previously noted that craniopharyngiomas with sellar/suprasellar origin have an enlarged or destroyed sella while arising from the infrasellar structures like the nasopharynx or the sphenoid sinus and show a sella with normal size.⁷ The position of the intrasellar dura might provide some further evidence as to the primary origin of the tumor. If the dura is being pushed inferiorly into the sella, it might indicate a suprasellar origin for the craniopharyngioma.

Fitz et al mentioned that any suprasellar tumor with two of the following characteristics can reliably be diagnosed as a craniopharyngioma: contrast enhancement, cyst formation, and calcification.²¹ The cystic component of these tumors typically appears hyperintense compared with normal CSF due to the protein and cholesterol content of the tumor. They can uncommonly be isointense in the presence of high keratin content and bony trabeculae.^{22,23} In Hald et al's study on craniopharyngiomas, preoperative CT and MRI were evaluated in nine patients. MRI showed both cystic and solid components in all cases, contrast enhancement in seven cases, and calcification in four patients. On CT images cysts and calcification were depicted in seven lesions, and contrast enhancement in six of the seven lesions.²⁴ The differential diagnosis of extracranial craniopharyngiomas on the basis of CT and MRI scan consists of rhabdomyosarcoma, lymphoma, squamous cell carcinoma, juvenile angiofibroma, and pharyngeal chordoma.⁷

Surgical resection is the mainstay of treatment for these tumors. This is especially true for the extracranial variants as they do not intimately involve the optic apparatus or hypothalamus. We utilized an endoscopic, endoscopic transsphenoidal approach to achieve complete resection in our patient. Obviously the most appropriate approach varies from a case-to-case basis. Other approaches which may be utilized for extracranial,

Table 1 The Previous Reports of Purely Infraseellar Extracranial Craniopharyngioma in the Literature

Year	Author	Age	Sex	Symptoms	Physical exam	Imaging	Treatment	Outcome
1988	Benitez et al ⁷	29	M	Three-year history of nasal stuffiness and pressure behind the nose	Obstruction of the left nasal cavity and erosion of the floor of the middle cranial fossa and clivus	High intensity on T2, lower intensity than brain on T1 on MRI scan	Lateral rhinotomy, ethmoidectomy, and partial removal of the maxillary sinus; radiation therapy	Discharged uneventfully
1984	Lewin et al ¹	27	F	Intermittent epistaxis, headache	1.5-cm pedunculated mass attached to the vomer	CT scan demonstrated the mass	Transpalatal approach	Recurrence free for 2 years
1990	Byrne and Sessions ⁸	29	M	Nasal obstruction	1-cm mass in the left nasal fossa; nasopharynx was totally occupied by the tumor	Sphenoid, ethmoid, maxillary sinus opacification on the x-ray and CT scan	Lateral rhinotomy and removal of the tumor; radiation therapy	Discharged uneventfully
2007	Arndt et al ⁶	15	F	Nasal obstruction, headache	5-cm clival mass filling the sphenoid and posterior ethmoid sinuses	MRI showed a partly solid, partly cystic mass with contrast enhancement of some of the solid parts	Transnasal endoscopic sphenoidectomy	Complete resection of tumor

CT, computed tomography; MRI, magnetic resonance imaging.

infraseellar craniopharyngiomas included the lateral rhinotomy approach, Caldwell-Luc approach (if lateral extension is noted) or rarely the transpalatal approach. Transpalatal approach gives a good exposure of the nasopharynx and posterior nasal cavity but in extensive tumor it is not feasible. The lateral rhinotomy approach is more applicable in tumors occupying the nasopharynx, nose, and paranasal sinuses.⁸ As the transsphenoidal approach provides excellent exposure to the sphenoid sinus, clival area, and posterior ethmoid area, it is a commonly used workhorse approach for lesions in this region. The choice of endoscopic or microscopic resection of the lesion is based mainly on the preference of the surgical team. We prefer the endoscopic option as it provides excellent panoramic visualization of the involved anatomy. With increased experience, we have found that this advantageous exposure outweighs the limitation of a two-dimensional working space.

Recurrence of the craniopharyngioma is a long-term concern even with total surgical removal. However, extent of surgical resection is certainly important in the long-term recurrence rates of these tumors. After total removal, the chance of tumor recurrence after 5 years is 13%. However, that rate increases to 58% with subtotal resection.²⁵ This is because of hidden “fingers” of tumor which often invade the hypothalamic region. Such recurrence usually occurs in the vicinity of the primary tumor site.²⁶ We certainly advocate the aggressive surgical resection of extracranial, infraseellar tumors such as that in our case. Tumors in these locations afford the surgeon the luxury of aggressive resection without risking the integrity of the pituitary gland, optic apparatus, hypothalamus, or the suprasellar vasculature.

CONCLUSION

We present a case report of a purely extracranial infraseellar craniopharyngioma mimicking a sinonasal mass. We also provide a review of the literature with emphasis on the craniopharyngeal canal, a remnant of Rathke pouch, as the likely source of these extracranial, infraseellar craniopharyngioma. Aggressive surgical resection is the mainstay of treatment of these extracranial, infraseellar variants. We advocate the endoscopic endonasal approach as it provides excellent panoramic visualization of this region. Even with complete surgical resection, long-term radiographic follow-up is recommended as tumor recurrence can be seen.

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