

An Infraselar Craniopharyngioma Involving the Sphenoid Sinus and Clivus

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Craniopharyngiomas are rare, benign, epithelial neoplasms that account for about 5% of all intracranial neoplasms. Craniopharyngiomas are mainly confined to the intraselar and suprasellar regions, with the suprasellar location being the most common. Exclusively infraselar craniopharyngiomas with no sellar involvement are extremely rare.^[1,2] We reported an infraselar craniopharyngioma involving the sphenoid sinus and clivus in a 16-year-old girl, and discussed the clinical features and management of this tumor.

A 16-year-old girl presented with a 6-month history of progressive headaches and nasal obstruction. Neurological examination revealed no cranial nerve abnormality. No signs or symptoms of pituitary dysfunction were evident. The results of laboratory studies, including hypophyseal function, were normal.

Radiologic examinations included computed tomography (CT) and contrast-enhanced magnetic resonance imaging (MRI). Cranial CT showed a well-defined infraselar mass invading the entire sphenoidal sinus and the space just anterior to the clivus. CT scan using the bone window revealed a clear edge of the lesion. The peripheral sclerotin was compressed and thinned, but without the destruction of the clivus [Figure 1a]. MRI demonstrated a large, well-defined, heterogeneous, infraselar mass confined to the sphenoid sinus. The mass consisted of cystic and solid tissues with contrast enhancement of some of the solid parts after administration of gadolinium-containing contrast medium. Contrast-enhanced sagittal T1-weighted images showed the sella turcica, suprasellar region, pituitary gland, brain parenchyma, cerebrospinal fluid spaces, and pituitary gland to be normal [Figure 1b-1d].

The lesion was resected via an endoscopic endonasal transsphenoidal approach. After removal of the compressed and thinned anterior wall of the sphenoid sinus, the tumor was visualized within the sphenoid sinus. The tumor was greyish-red, moderately vascular, and firm, with cystic areas. The tumor was removed completely in a piecemeal fashion. The entire tumor was extradural, and the sella was intact, with no extension superiorly into the sella or posteriorly through the clival dura, confirming an exclusively infraselar location. A postoperative MRI scan confirmed complete excision of the tumor [Figure 1e-1g]. Histologic examination revealed an adamantinomatous craniopharyngioma [Figure 1h]. The patient was discharged from hospital with improvement of her nasal obstruction and headache-free. The patient was neurologically intact and disease-free at 2-year postoperatively.

Craniopharyngiomas are benign but locally aggressive tumors. They are generally located intracranially and occur with similar frequencies in children and adults. Ninety percent of these tumors arise in the sellar or suprasellar region and then extend to the anterior, middle or posterior fossa, though rare cases of infraselar craniopharyngioma have been reported. Infraselar craniopharyngiomas are generally thought to originate from the embryologic development of the adenohypophysis. In 1904, Erdheim postulated that craniopharyngiomas developed from squamous remnants of the obliterated craniopharyngeal duct, suggesting that these tumors may arise at any point along the tract of migration of Rathke's pouch from the vomer, the roof of the nasopharynx, or through the midline sphenoid bone beneath the floor of the sella turcica.^[2]

The symptoms and clinical findings of these lesions are related to their size and location, and the degree of compression of surrounding structures. Patients with suprasellar lesions commonly present with visual field defects, pituitary

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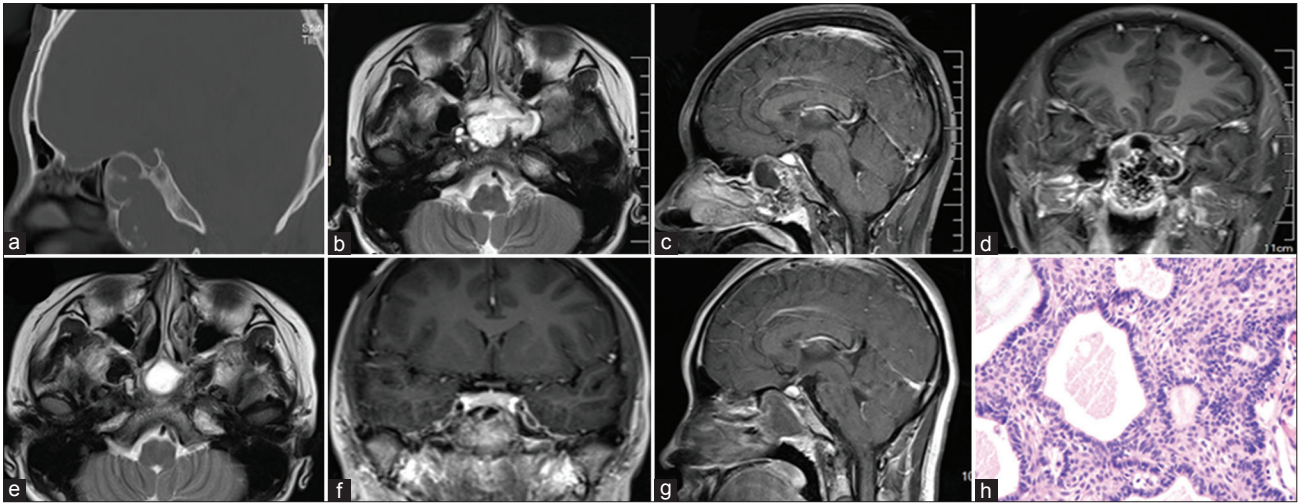


Figure 1: Pre- and post-operative imaging, and microscopic appearance. Computed tomography (CT) scan showing the intact sella floor. A heterogeneous mass is located in the sphenoid sinus and beneath the clivus. CT bone window image (a) revealing calcification of the tumor tissue. Magnetic resonance image (MRI) scans showing partial enhancement in the sphenoid sinus. Note the normal appearance of the pituitary stalk and gland with no relationship between the sella and infrasellar mass (b-d). Postoperative MRI at 3 months postoperatively revealed that the tumor had disappeared (e-g). Typical appearance of adamantinomatous type of craniopharyngioma (h) (H and E Staining, $\times 100$).

insufficiency, and symptoms of raised intracranial pressure. In contrast, infrasellar craniopharyngiomas present with symptoms of headache, cavernous sinus syndrome, frontal headache, nasal obstruction, epistaxis, and nasopharyngeal and nasal fossa masses. The current case presented with progressive headaches and nasal obstruction, without pituitary insufficiency, suggesting that the tumor had appeared in the sphenoid sinus and subsequently grown extradurally and superiorly.

The most common location of infrasellar craniopharyngiomas is the sphenoid sinus, either alone or combined with other sites such as the nasopharynx, sella turcica, suprasella, ethmoid sinuses, or maxillary sinus.^[1] We suspected that the tumor in the present case originated in the infrasellar region because the mass was confined to the sphenoid sinus, and the dura mater of the sellar floor was intact. Pituitary hormone levels were also found to be normal both pre- and post-operatively.

The treatment of choice for infrasellar craniopharyngioma is complete surgical excision, with the exact surgical approach determined by the anatomical location of the tumor. The surgical approaches that can be used for infrasellar craniopharyngiomas include the lateral rhinotomy approach, Denker's medial maxillectomy approach, the transpalatal approach, and the transsphenoidal approach.^[1]

We used an endoscopic transsphenoidal approach to achieve complete resection in our patient. This approach provides excellent exposure of the sphenoid sinus, clival area, and posterior ethmoid area, and is, therefore, commonly used for lesions in this region. We recommend the aggressive surgical resection of infrasellar tumors such as the one reported here, because of the lack of involvement of the pituitary gland, optic apparatus, hypothalamus, and the suprasellar vasculature.

The risk of local recurrence can be as high as 50% in the event of incomplete excision,^[3] however, only one case of recurrent infrasellar craniopharyngioma has been

reported to date.^[4] Radiotherapy is currently the most frequently used adjuvant treatment and plays a major role in preventing recurrence and improving survival. Postoperative radiotherapy has been shown to increase survival and is recommended when complete surgical removal of the tumor is not possible.

The reported overall 10-year disease-free survival rate for craniopharyngiomas ranges from 60% to 96%.^[5] However, this is based purely on patients with suprasellar craniopharyngiomas because of the lack of reported cases of entirely infrasellar craniopharyngiomas. It is, therefore, not possible to predict the prognosis of infrasellar craniopharyngiomas. The patient has been following-up with serial scans closely.

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