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# GIANT CRANIOPHARYNGIOMA: A RARE CASE REPORT

**Richi Sinha[1], Rohit Kumar[2], Rakesh Kumar Singh[3]***

[1, 3]Department of Otolaryngology, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, India.
[2]Department of Medicine, Katihar Medical College, Kathar, India.

Corresponding Author:- **Rakesh Kumar Singh**
**E-mail:** rksent5@gmail.com

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**ABSTRACT**
Craniopharyngiomas are relatively benign neoplasms of sellar or suprasellar region. The diagnosis is clinical and radiological. It is confirmed by characteristic histological findings. These unusual tumors should ideally be managed surgically in specialized centers and there is no role of systemic chemotherapy. We have discussed here the diagnosis and management of one such case of Craniopharyngioma with extensive spread and encroachment of vital structures. Careful surgical approach with expertise in these cases can save the patient from the associated morbidity and mortality.

## INTRODUCTION

Craniopharyngioma is a benign tumor of the embryonic tissue of pituitary gland. They account for nearly 1-5% of primary brain tumours [1]. They are derived from Rathke cleft, hence also known as Rathke's pouch tumour. Other synonyms for this condition are craniopharyngeal duct tumour, adamantinoma and dysodontogenic epithelial tumour. They arise from the cells along the pituitary stalk, specifically from nests of odontogenic epithelium within the suprasellar region and therefore contain deposits of calcium, which are evident radiologically. It can be seen along the infundibulum anywhere from the floor of third ventricle to the pituitary gland. Craniopharyngiomas are commonly seen in paediatric age group with variable presentation depending on its location and size. Presenting complaints may include headache, visual symptoms, hormonal imbalances etc. The diagnosis is clinical and radiological. It is confirmed by characteristic histological findings. The management of patients presenting with these unusual tumors is ideally surgical, in a specialized center that has a particular interest. There is no role of systemic chemotherapy [2]. Here we present a case of Craniopharyngioma with extensive spread to vital structures and also discuss the diagnosis and management of such cases.

## Case Report

A 20 years-old female presented to the department of otorhinolaryngology with the complaints of progressive bilateral nasal obstruction and frontal headache of 6 months duration. The visual acuity of the patient was normal with no other visual disturbances. There were no symptoms of any hormonal imbalance. Clinical examination with the help of endoscope showed smooth protuberance in the posterior part of right nasal cavity and extending up to the nasopharynx, obstructing bilateral choana probably arising from base of skull. All routine investigations were normal. T1-weighted (plain and gadolinium enhanced) and T2-weighted magnetic resonance imaging (MRI) showed a large iso-intense mass in the sphenoid sinus with destruction of sella turcica and the ethmoid sinus (Fig 1). It extended to the optic nerve without eroding it, and also spread up to the cortex. Post-contrast imaging showed enhancing, heterogeneous mass in the sphenoid region and peri-sphenoid (Fig 2).

On the basis of MRI findings the trans- sphenoid approach was adopted to remove the lesion. At the end of tumor resection, bilateral nasal packing was done which was removed on postoperative second day. The postoperative period was uneventful with full recovery.

Histopathalogica examination confirmed the diagnosis of craniopharyngioma. After 1 year follow-up, neurological examination and hormonal studies were within normal limits. On endoscopic examination there was no recurrence or residual tumor seen in the sphenoid sinus. Postoperative computed tomography also showed complete clearance of disease with no recurrence (Fig 3).

**Fig 1. T1 waited MRI showed a large iso-intense mass in the sphenoid sinus with destruction of sella turcica.**

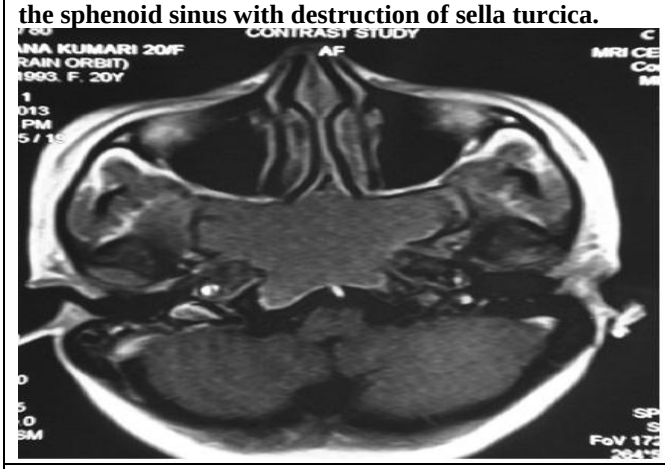



**Fig 2. Post-contrast MRI showed enhancing, heterogeneous mass in the sphenoid and peri-sphenoid region.**

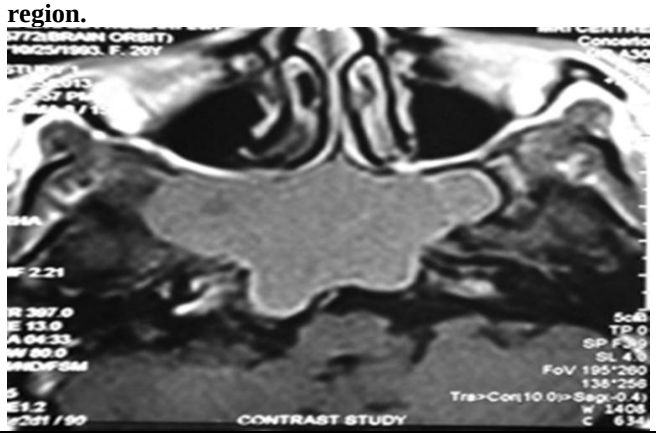
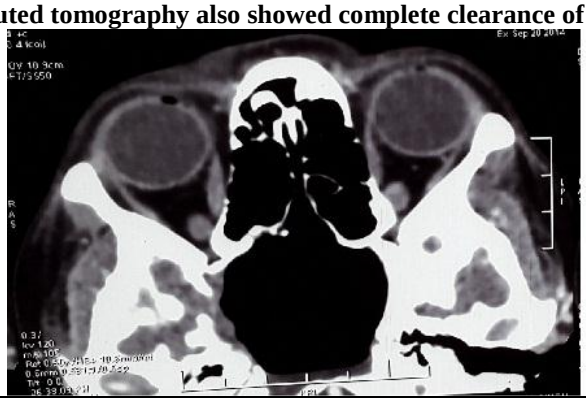



**Fig 3. Postoperative computed tomography also showed complete clearance of disease with no recurrence**



## DISCUSSION AND CONCLUSION

Craniopharyngiomas are rare embryogenic malformations of the sellar area. Initial nasopharyngeal presentation is rare. This patient presented with a nasopharyngeal bulge. They are benign on histology but are locally malignant [3]. The incidence ranges from 0.13 to 2 per 100,000 populations per year. There is equal distribution in both the gender and bimodal age distribution, in children at 5–14 years and in adults at 65–74 years of age. In children, craniopharyngiomas account for 5% of all tumours and 50% of all sellar/parasellar tumours [2, 4]. It has two histological subtypes, adamantinous and papillary craniopharyngiomas. The first type is predominantly seen in children while the other is almost exclusively seen in adults. These are typically slow growing tumors. Symptoms usually develop insidiously and become obvious only after it attains a size of 3cm. The most common symptoms include headache, hormonal imbalances and visual impairment. Most women complain of amenorrhea. Neurological examination reveals signs indicating raised intracranial pressure. Visual field examination mostly shows bitemporal hemianopia. Signs of various endocrinopathies like hypothyroidism, adrenal insufficiency may be seen. Unlike this typical presentation our patient showed no such symptoms other than frontal headache. Rarely tumors such extensive can present with symptoms depending on the region it spread to, like nasal obstruction in this case. Like most tumors, the cause of this tumor is unknown. The classical radiographic appearance of a craniopharyngioma is a partly solid and cystic, calcified mass of sellar or parasellar reagion. But in this case there was further advancement towards optic nerve and cortex. Magnetic resonance imaging very well defines the tumour extent and involvement of the hypothalamus. Therefore preoperative MRI is essential [2]. CT angiography or Magnetic resonance angiography can be done to trace the course of any vessels through the tumor, and help to differentiate a tumor from vascular malformation. The differential diagnosis includes arachnoid cyst, Rathke's cleft cyst, Pituitary macroadenoma, metastasis, meningioma, epidermoid and dermoid tumour, hypothalamic-optic pathway glioma, hypothalamic hamartoma, teratoma, etc [2]. Management

of craniopharyngiomas has been controversial. The treatment strategy is a multidisciplinary approach involves both surgery and radio-oncology. The first involves attempted gross total or subtotal resection of the tumor. The second approach is based on further limited resection and debulking the tumor to merely reduce the mass effect on optic pathway followed by radiation therapy [5]. The second pathway was developed because of high risk of death, endocrinologic complications, and behavioral dysfunction experienced with the gross total resection of tumors. In our case we used a third combined approach were the patient underwent subtotal resection of the tumor followed by postoperative radiotherapy for optimum results. Being radiosensitive tumors, radiotherapy is used as adjuvant treatment modality these days. It helps prevent recurrence and reduces associated morbidity and mortality. Recent multicenter cooperation in this area has already led to beneficial results. The surgical strategy at initial diagnosis should aim at a maximal degree of resection, respecting the integrity of optical and hypothalamic structures. This prevents severe sequelae and therein minimizes consequences that could negatively exacerbate patient's quality of life.

## CONCLUSION

Craniopharyngiomas are rare benign tumors of the sellar area with low-grade histological malignancy. Despite high survival rates quality of life is frequently impaired in long-term survivors due to sequelae caused by the anatomical proximity of the tumor to the optic nerve, pituitary gland, and hypothalamus. But multicenter approach with surgical expertise in such extensive cases can save the patient from the associated morbidity and mortality.

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