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Transphenoidal Skull Base Excision of a Unique Pituitary Adenoma

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Abstract: Ectopic nasopharyngeal pituitary adenoma is an extraordinarily rare entity that is often misdiagnosed. They have a very indolent presentation, more so in non-functioning adenomas. We present a case report of a patient who presented with nasal obstruction to the Otolaryngology OPD and was eventually diagnosed to have an ectopic pituitary adenoma in the nasopharynx.

Keywords: Endoscopic Surgery; Pituitary Adenoma; Skull Base Surgery; Ectopic Pituitary.

INTRODUCTION

Pituitary adenomas are a diverse group of tumours arising from the pituitary gland with a reported prevalence rate of 16.7% (Ezzat, S. *et al*., 2004). Historically, these tumours have been classified based on size into microadenomas (<1cm) which geberally present with systemic effects of hormone excess and macroadenomas (>1cm) which present as space-occupying lesions that grow into surrounding structures. Most pituitary macroadenomas grow superiorly and present with visual symptoms, headache, or other neurological symptoms (Hyrcza, M.D. *et al*., 2017) Upto 2% of these tumours may have an infrasellar extension but very rarely erode the floor of the sphenoid sinus to present as a nasopharyngeal mass (Panchani, R. *et al*., 2013). Pituitary ectopia is a very unusual presentation, most of them being located in the sphenoid sinus or suprasellar region. About 27% of cases are located sporadically along the clivus, cavernous sinus, petrous bone, sphenoid bone, mid nasal ductus and the third ventricle. (Ali, R. *et al*., 2010) Although a definitive diagnosis is only possible with CT and MR Imaging and histopathological study, a careful history taking and detailed clinical examination needs to be done to rule out functional pituitary adenomas. Immunohistochemical analysis along with staining for pituitary hormones is required for confirmation of diagnosis.

CASE REPORT

A 47 year old male patient presented to the out-patient department with progressive bilateral nasal obstruction of 6 months duration.

General Head and Neck examination and anterior rhinoscopy showed no abnormal findings. On posterior rhinoscopy, a smooth pinkish large bulge was noted arising from the posterior wall and occupying the entire nasopharynx with bilateral choanal obstruction was noted.

A contrast enhanced CT scan was done, which showed a well-defined heterogenous mass eroding into the sphenoid sinus, body of sphenoid and clivus. There was no intracranial extension and the bony floor of the sella appeared intact.

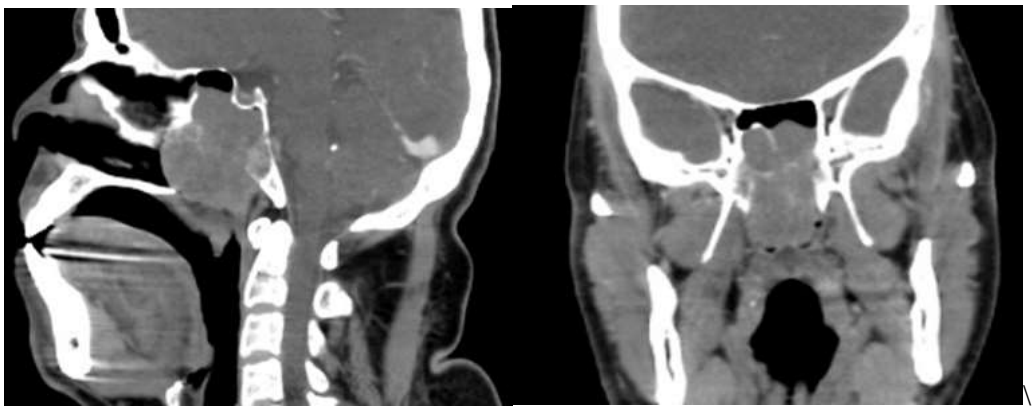

Figure 1: Contrast-enhanced CT scan: Sagittal and coronal views showing a well defined heterogenous lesion measuring 4.8x3.5x4.8 cm in the nasopharynx eroding into the sphenoidal sinuses, body of sphenoid bone and clivus, obliterating bilateral fossa of Rosenmuller and Eustachian opening. No obvious intracranial extension. Sella appears normal.

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To rule out intracranial extention and dural invasion. A contrast enhanced MRI was also done which showed a heterogeneously enhancing well defined lobulated mass lesion involving the nasopharynx, extending into the sphenoid sinuses, posterior ethmoid sinuses and posterior nasal cavity. Superiorly, the lesion was totally obliterating the sphenoid sinus and extending to abut the pituitary fossa without obvious infiltration into it.

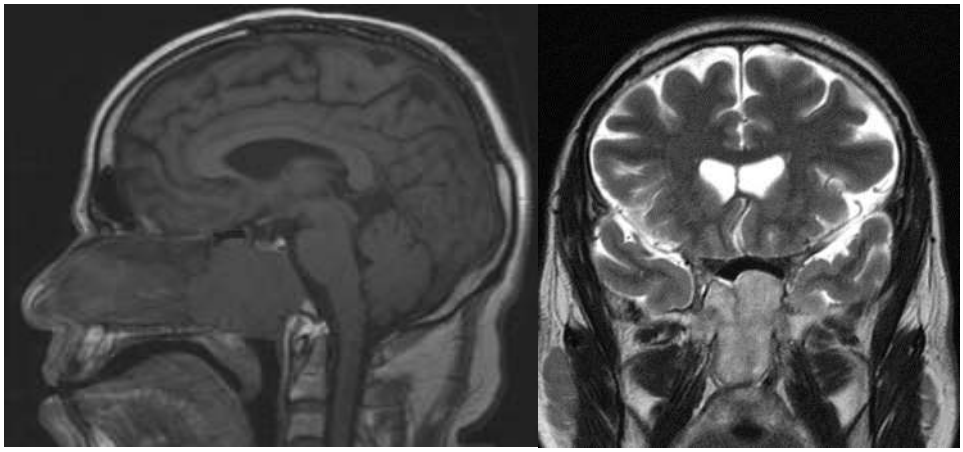

Figure 2: Contrast enhanced MRI: A heterogeneously enhancing well defined lobulated mass lesion appearing T2W hyperintense, T1W isointense lesion, showing mild diffusion restriction, measuring ~4.6 x 3.3 x 4.4 cm noted involving the nasopharynx, extending into the sphenoid sinuses, posterior ethmoid sinuses and posterior nasal cavity. Superiorly, the lesion is totally obliterating the sphenoid sinus and extending to abut the pituitary fossa without obvious infiltration into it

The patient subsequently underwent a diagnostic nasal endoscopy with biopsy of the lesion as an outpatient procedure. The histopathological examination from the biopsy specimen was reported to have no malignant cells, with no definitive discernible features. In view of clinical and radiologically suspicion of nasopharyngeal malignancy, patient underwent transnasal trans-sphenoid KTP Laser assisted excision of the lesion under general anaesthesia. Frozen section was sent intra-op which was inconclusive.

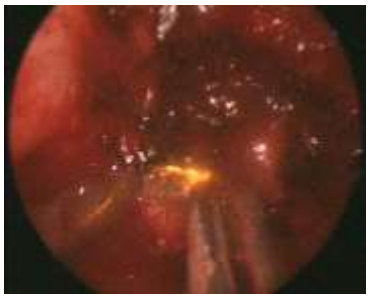

Figure 3: KTP laser assisted excision of the nasopharyngeal lesion

Patient recovered well from the procedure. The final histopathological report was suggestive of pituitary adenoma. Immunohistochemical analysis was positive for synaptophysin and negative for prolactin. Hormone assays for Adrenocorticotropic hormone (ACTH), Lutenising Hormone (LH), Follicle Stimuating Hormone (FSH), Thyroid Stimulating Hormone TSH and cortisol levels were done post-operatively in view of the histopathological diagnosis, which were found to be normal.

DISCUSSION

Although discovery of pituitary tissue in the nasopharynx is very rare in clinical practice, this case emphasizes the importance of keeping in mind the differential diagnosis of pituitary adenoma while evaluating adult patients presenting with a nasopharyngeal mass. Usually, such a pituitary mass in the nasopharynx is due to infrasellar extension of the pituitary adenoma invading into the sphenoid sinus, nasopharynx or clivus region. When such a lesion is not in continuity with the intrasellar normal pituitary gland and the bony sella turcica can be visualized radiologically to be intact, a diagnosis of Ectopic

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Pituitary Adenoma can be considered. They are believed to arise from cell rests that have arrested during the anterior pituitary primordium migration through the craniopharyngeal canal and then potentially undergo neoplastic transformation. (Riccio, L. *et al*., 2020)

Ectopic pituitary adenoma is very rare with less than 50 cases reported in literature, among which those occurring in the nasopharynx are extraordinarily rare. To the best of our knowledge, this is only the fifth report of such a case in literature.

While most pituitary adenomas tend to be non-functioning, it is observed that more than 60% of ectopic pituitary adenomas are hormone-secreting tumours. (Riccio, L. *et al*., 2020) However, in our case report we found that the patient had no signs or symptoms of hormone secreting tumour and also the hormone assays which were subsequently done were normal. Like in our case report, most of these patients present with symptoms of mass effect including nose block, eustachian tube dysfunction, epistaxis, headache etc.

Modern day imaging techniques with high spatial definition power have been able to pick up these ectopics at an early stage. These lesions should display no connection with pituitary fossa and stalk. They usually appear isodense on CT scans, showing adjacent bone remodelling, sclerosis, or erosion, and only rarely calcification. Additionally, they typically look isointense to grey matter on T1- and T2-weighted sequences, with moderate, heterogeneous enhancement, characterized by a rapid enhancing and slow washout pattern. (Ferraz-Filho, J.R. *et al*., 2014) However, a pre-existing clinical suspicion is key in making accurate diagnosis. Nevertheless, the final diagnosis can only be made by histopathological evaluation.

The primary modality of treatment in such cases is surgical- transphenoidal gross tumour resection in this case. Other modalities of treatment like radiotherapy, particularly stereotactic radiosurgery can be considered as an adjuvant treatment. Medical therapy can be employed in hormone-secreting tumours as an adjunct but the final definitive management still remains surgical excision. (Katznelson, L. *et al*., 2014)

To conclude, ectopic pituitary adenomas are an extremely rare entity, probably also under-reported due to frequent misdiagnoses but they should always be kept in mind when encountering a nasopharyngeal mass in an adult patient. The ability to accurately diagnose and provide timely management depends on the clinicians suspicion, the radiologist's acumen to pick it up on imaging and the pathologist's meticulous pursuit of the histopathological diagnosis.

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