Brief Communication

An interesting case of pituitary adenoma presenting as an invasive nasopharyngeal tumor

Roopal Panchani, Tarun Varma, Ashutosh Goyal, Sudhir Tripathi

Department of Endocrinology, Sir Ganga Ram Hospital, New Delhi, India

ABSTRACT

Pituitary masses usually present as sellar masses with suprasellar or parasellar extension. However, in unusual cases pituitary tumors especially macroprolactinomas and nonfunctional adenomas can present with intranasal extension which can be misdiagnosed as nasal polyp or a primary invasive nasopharyngeal malignant tumor. The otolaryngologists should be familiar with this rare presentation of pituitary masses. Measurement of prolactin (PRL) is essential in cases of recalcitrant nasal polyps or rhinorrhea as it may change the management in such cases. Here we describe case of a patient with an invasive pituitary adenoma who had presented in the otorhinolaryngology department with a nasal obstruction and epistaxis. We have also reviewed 30 cases of pituitary adenoma with nasopharyngeal invasion published in past.

Key words: Chromophobe adenoma, invasive pituitary adenoma, macroprolactinomas, nasopharyngeal malignancy

Introduction

Nasopharyngeal masses form a small fraction of cases presenting with nasal obstruction. Commonest cause of nasopharyngeal malignancy is classical and undifferentiated variety of nasopharyngeal carcinoma. Discovery of pituitary tissue in the form of a nasopharyngeal mass is an extremely rare occurrence for the otolaryngologist. A careful history focusing on subtle features of hypopituitarism, imaging of sella, and histopathological examination for neuroendocrine features plays key role in identifying such cases. Here we discuss a case of a pituitary adenoma with an unusual nasopharyngeal invasion.

Case Report

A 47-years-old man presented in the otorhinolaryngology department with a 4 month history of chronic left sided nasal obstruction, periodic epistaxis along with pain and difficulty in opening left eye. He also complained of frontal headache. Patient suffered from diabetes mellitus since last 10 yrs and had proliferative diabetic retinopathy in right eye.

On examination, patient had frontal sinus tenderness. There was complete third nerve palsy on left side [Figure 1]. Other cranial nerves were not affected. There was no periorbital edema. Ocular examination on right side was normal. There was no blackish discoloration of nasal or palatal mucosa. There was no conductive deafness. Patient was provisionally suspected of having an invasive nasopharyngeal malignancy or rhino-ocular mucormycosis of paranasal sinuses. However, nasal endoscopy revealed sphenoid sinus filled with expansive firm tumor. Biopsy from the mass showed nests and cords of synaptophysin positive cells with uniformly looking nuclei accompanied with necrotic fibro connective tissue consistent with possibility of pituitary adenoma with extensive necrosis. In view of the histopathological diagnosis, magnetic resonance imaging (MRI) of sella was planned and endocrine consultation was done.

MRI revealed a heterogenous mass lesion in sphenoid sinus extending posteriorly in pituitary fossa with stalk

Access this article online	
Quick Response Code:	Website: www.ijem.in
	DOI: 10.4103/2230-8210.119533

Corresponding Author: Dr. Roopal Panchani, Department of Endocrinology, Sir Ganga Ram Hospital, New Delhi - 110 060, India.
E-mail: drroopalpanchani@yahoo.in

displaced posteriorly and pituitary gland not separately discernible from the lesion. The fat plane between the pituitary gland and lesion was effaced [Figures 2-5]. Upon endocrine evaluation, the patient had evidence of anterior pituitary hypofunction in the form of weakness, easy fatigability, reduced libido, nausea, vomiting, and increased episodes of hypoglycemia. He denied of having polyuria. Thyroid stimulating hormone (TSH) was 0.213 μIU/ml (0.2-5.1μIU/ml) with a free thyroxine (T4) of 0.6 ng/dl (0.95-2.25 ng/dl). Cortisol measured at 8:00 am was 4.93 μg/dl (5-23 μg/dl). His prolactin (PRL) level measured in serial dilutions was 2.0 ng/ml (2.7-17.0 ng/ml). Luteinizing hormone (LH) and Follicle-stimulating hormone (FSH) levels were normal. Transnasal transsphenoidal surgery was planned after optimization of thyroid and adrenal status. Postoperatively hormone supplementation was continued. Histopathological examination confirmed the diagnosis of pituitary adenoma. On immunohistochemistry, the

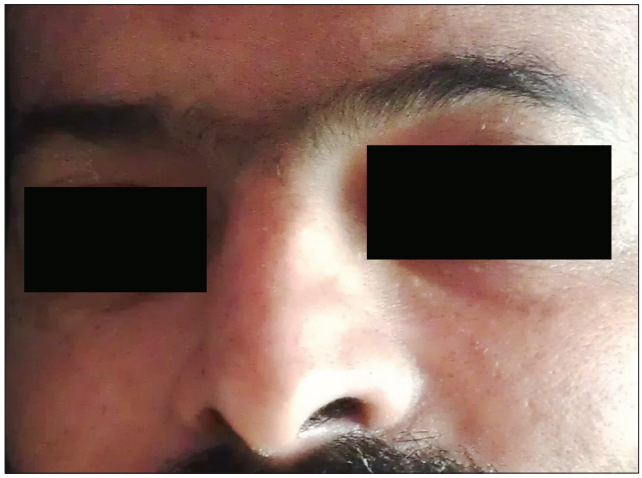

Figure 1: Patient with left sided ptosis

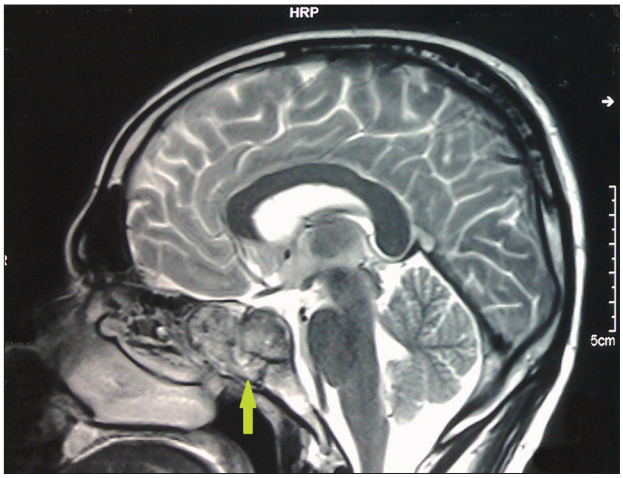

Figure 2: Sagittal T2 weighted MRI sella view showing heterogenous mass infiltrating sella completely. Pituitary tissue is not separately discernible from the mass (green arrow)

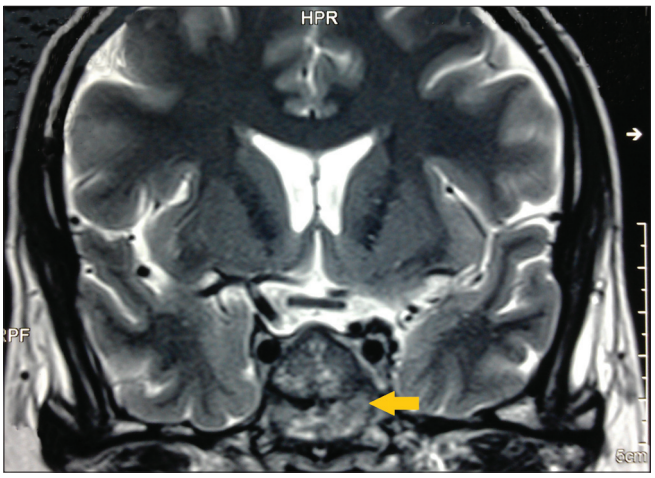

Figure 3: Coronal view T2 weighted image showing mass infiltrating the sphenoid sinus completely (yellow arrow) with minimal suprasellar bulging. Optic chiasm is intact. Stalk is slightly shifted to right with normal thickness and enhancement. Parasellar spread is minimal

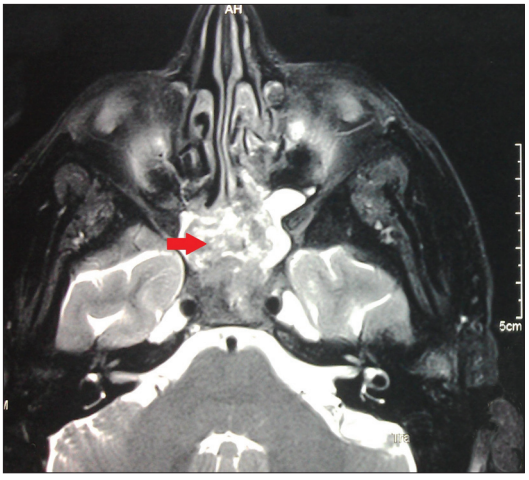

Figure 4: Axial view showing infiltration by the invasive pituitary adenoma into sphenoid sinus and posterior ethmoidal air cells bilaterally (red arrow)

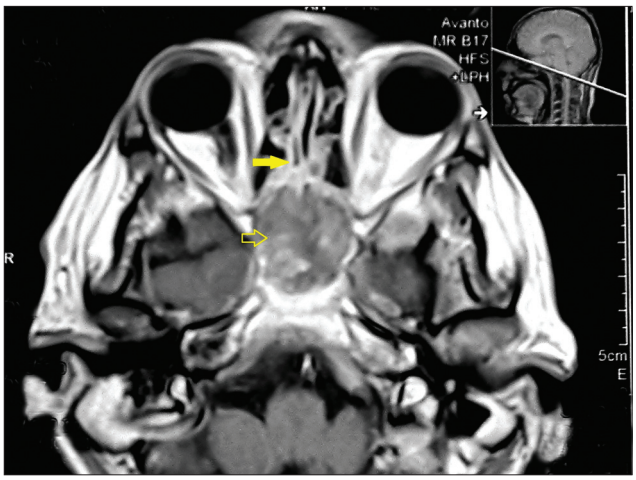

Figure 5: Axial T1 weighted view with contrast showing heterogenous enhancement by the mass and some areas of non-enhancement suggestive of necrosis. There is no evidence of hemorrhage

cells were positive for synaptophysin and negative for prolactin, Adrenocorticotropic hormone (ACTH), and growth hormone (GH) markers. So the diagnosis was consistent with chromophobe adenoma of pituitary.

Table 1: Cases of pituitary adenoma invading nasopharynx published in the past[$]

Author	Year of publication	Cases	Type of adenoma
Boyd	1910	1	NA
Cushing HW	1912	3	Chromophobe adenoma
Bailey OT	1940	1	Chromophobe adenoma
Shea JJ	1941	1	Chromophobe adenoma
White JC	1945	1	Chromophobe adenoma
Kay S	1950	3	Chromophobe adenoma
Aquino JA	1957	1	Chromophobe adenoma
Borsanyi S	1960	1	Chromophobe adenoma
Davis JM	1980	1	Chromophobe adenoma
Lessard ML	1985	1	Chromophobe adenoma
Dent JA	1989	2	Chromophobe adenoma
Sancho M	1992	1	Prolactinoma
Van der Lely AJ	1992	4	Prolactinoma
Iwai Y	1992	1	Prolactinoma
Sharma K	1995	1	Somatotroph adenoma
Luk IS	1996	2	1 Prolactinoma, 1 Corticotroph adenoma
Giannou P	2001	1	Chromophobe adenoma
Jankiewicz WJ	2001	1	Prolactinoma
Johnston PC	2012	3	Prolactinoma

[$]Cases of ectopic pituitary adenoma arising in nasopharynx are excluded.
NA: Data not available

Discussion

This case highlights an unusual nasal presentation of pituitary adenoma. Up to 2% of pituitary tumors have an infrasellar extension[1] but very few of them erode the floor of sphenoid sinus to enter in nasopharynx and nasal cavity to present as a nasal polyp. Approximately 35 cases of nasopharyngeal invasion of pituitary adenomas have been reported. The initial cases of pituitary adenoma presenting as nasal polyp were identified by Boyd[1] and Sir Harvey Cushing in 1910 and 1912.[2] Most of the cases reported are chromophobe adenomas followed by few macroprolactinomas and one case of somatotroph and corticotroph adenoma [Table 1].[3,4]

Diagnosis in case of a functional pituitary adenoma is not difficult as signs of PRL, GH or ACTH excess would be obvious. Clinical features of hypopituitarism, although more common with pituitary tumors, can be occasionally seen in cases of meningiomas, craniopharyngiomas, and chordomas infiltrating sella. Diagnosis becomes especially challenging in cases where a pituitary adenoma is nonfunctional and has an exclusive infrasellar spread when it would mimic a nasopharyngeal mass.

Rarely, an ectopic pituitary tissue is located in extracranial sites like sphenoid sinus, nasal cavity, and nasopharynx. In

Table 2: Differential diagnosis of invasive nasopharyngeal masses involving base of the skull[#]

Diagnosis	Clinical features	Special features
Nasopharyngeal carcinoma	Most common tumor in this space, common in Asian, African, and Chinese males present usually with cervical lymphadenopathy, trismus, otitis, nasal twang, and regurgitation	Enlargement of retropharyngeal lymph nodes is typical. High dose radiotherapy is treatment of choice
Nasal angiofibroma	Relatively common in young males, frequent profuse epistaxis, recurrent nasal blockade, otomastoiditis	Aggressive vascular tumor, biopsy leads to extensive bleeding, widening of sphenopalatine foramen on CT, salt pepper appearance on MRI with multiple flow voids, marked contrast enhancement
Sphenoid sinus mucocele	Rare, can have pleiotropic manifestations, nasal blockage, visual symptoms and cranial nerve palsy. Pituitary involvement rare	Homogenous mass expanding sphenoid sinus with bone erosions and bony defect, no contrast enhancement, isodense on CT
Sinonasal meningiomas	Very rare, nasal symptoms predominant	Bright contrast enhancement on T1, dural tail sign, bone erosion, hyperostosis
Chordoma (primary or extension of clival lesion)	Peak in the fourth decade, male predilection, midline nasopharyngeal mass	CT-Bony lytic changes along the anterior surface of the clivus, midline sinus tract, intratumoral septa. MRI-heterogeneous hyperintense T2 signal intensity
Pharyngeal craniopharyngiomas	More frequent in >15 yr age, similar sex distribution, frontal headache, nasal symptoms, cavernous sinus symptoms	CT-Heterogeneous nature of the tumor with its solid and cystic components, calcification, multicysts, lytic lesions, irregular enhancement. MRI-shows cystic portions, mixed intensity signal, inhomogeneous or heterogeneous enhancement
Non Hodgkins lymphoma	Most common site of extranodal non-Hodgkin lymphoma in this region. Occurs in the sixth decade, Predominant cervical lymphnode enlargement	Mainly large B cell and follicular. homogeneous tumor, diffusely involve all walls of the nasopharynx, exophytic spread filling the airway, propensity to extend down into the tonsil, rather than up into the skull base
Extramedullary plasmacytoma	Common in males in 6th and 7th decades Nasal obstruction without bone destruction	Well-demarcated homogeneous soft-tissue density, bone erosion, significant contrast enhancement with central inhomogeneity
Nasopharyngeal tuberculosis	Mimics NPC, especially in Asian patients. Discrete polypoid mass in the adenoids or diffuse soft-tissue thickening. Extension outside nasopharynx not usually a major feature	Discrete polypoid masses in the center of the roof and upper posterior wall of the nasopharynx, arising from the site of the adenoids. Characteristic septations on T2-weighted images and contrast-enhanced T1-weighted images

[#]In these entire differential diagnoses pituitary or sellar involvement is less frequent and, hence, invasive pituitary adenoma tops the list in case of nasopharyngeal masses with concomitant hypopituitarism

such cases, presence of a normal pituitary gland in sella can help to differentiate it from an adenoma arising from ectopic pituitary tissue.

Other intracranial tumors which have nasal invasion are meningiomas, chordomas, and rarely germinomas. The differential diagnosis of nasopharyngeal tumors with intracranial invasion includes nasal angiofibroma, sphenoid sinus mucocele, pharyngeal craniopharyngioma, olfactory neuroblastoma, giant cell tumor, and adenoma arising from ectopic pituitary tissue [Table 2].[5]

Histological examination is utmost essential for differentiating pituitary tissue from other tissues. Histologically, pituitary tissue can be identified by a typical endocrine growth pattern which consists of tumor cells arranged in packets, ribbons, or rosettes, with prominent delicate vascularized stroma and immunohistochemical showing of neuroendocrine markers, and pituitary hormones in the tumor cells. Differential diagnosis on histology includes small cell cancer, non-Hodgkin's lymphoma, and plasmacytoma.[6]

Due to low frequency of occurrence, there are no specific guidelines on management of pituitary adenomas with intranasal extension. Medical treatment is justified in cases of prolactinomas without cerebrospinal fluid (CSF) leak or other complications. However, in presence of complications like pituitary apoplexy, neurological deficit or intolerance to medications or in case of nonfunctional adenomas, the best approach is surgery as per review of existing literature. Either microscopic transsphenoidal or endoscopic transnasal may be used to treat with choice of therapy guided by surgeon's experience.

To conclude, pituitary adenomas with nasopharyngeal invasion even for being very rare occurrence should be a part of differential diagnosis of sinonasal masses. The diagnosis needs a collaborative work by an expert otolaryngologist, an experienced endocrinologist, a sound radiologist, and an astute pathologist. Detailed endocrine evaluation with special emphasis on features of hypopituitarism, PRL, GH or ACTH excess, cranial nerve involvement is mandatory. The impact of accurate preoperative diagnosis on management and follow up of such cases cannot be underestimated considering the vast difference between management of pituitary adenomas and nasopharyngeal tumors.

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Cite this article as: Panchani R, Varma T, Goyal A, Tripathi S. An interesting case of pituitary adenoma presenting as an invasive nasopharyngeal tumor. Indian J Endocr Metab 2013;17:S135-8.

Source of Support: Nil, **Conflict of Interest:** None declared.