Case Report

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Extrasellar Sphenoid Sinus Pituitary Adenoma: A Case Report

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ABSTRACT

Pituitary tumors are usually benign adenomas presenting as space occupying lesions in the sella turcica. Rarely these tumors may present beyond the sella either as an extension of the primary tumor or tumor arising from an aberrant pituitary location. One such extra sellar extension or origin of these tumors is in the olfactory spaces, comprising of 2% of all the nasal tumor. Because of the rarity of lesion and sometimes nonfunctional nature of the tumor, these may present diagnostic problems to the otolaryngologists and are diagnosed as polyps ornasopharyngeal malignant tumors. Measurement of pituitary hormones may provide a clue to the diagnosis in cases of paranasal polyps or rhinorrhea as it may change the management in such cases. We present here a case of a patient with an invasive pituitary adenoma who had presented in the Departmentof Otorhinolaryngology with epistaxis.

Keywords: *Chromophobe Adenoma, Invasive Pituitary Adenoma,Ectopic Sphenoid Sinus Pituitary Adenoma*

Introduction

Pituitary adenomas comprise of 10 -20% of all intracranial tumors with majority of them located in the sella turcica [1]. Sometimes they present outside the sella in the nasopharynx, sphenoid sinus and nasal cavity with an incidence of 2% of all adenomas [2]. These extrasellar tumors may result from invasion of floor of the sella by an aggressive pituitary adenoma or may arise from a remnant of Rathke's pouch.

Nasopharyngeal masses form a small fraction of cases presenting with nasal obstruction. Commonest cause of nasopharyngeal mass 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 location.

Case Report

A, 59 year old female presented to the department of Otolaryngology with complaint of right side nose bleeds for the past 4 days. The examination of the nasal cavity by nasal endoscopy revealed a deviated nasal septum toward the left, a pale mucosa with a normal external frame work. Systemic examination was normal. Radiological examination using a CT scan for the paranasal sinuses showed an intra sphenoidal mass extending to the right posterior ethmoid sinuses and right middle meatus. Posteriorly it is seen extending into the clivus and superiorly reaching up to the floor of sella. The posterior part of the floor of sella appears deficient, however the outlines of the pituitary gland appear fairly well defined and separate from the mass. Further signal intensity of the pituitary is slightly more hyperintense than the mass.

Radiological possibilities included chordoma, sphenoidal malignancy probably a nasopharyngeal carcinoma or an invasive pituitary tumor. Based on the above radiological report the patient underwent a transnasal endoscopic debulking of the sphenoidal mass under general anesthesia. Post-operative period was uneventful.

A, single globular grey brown soft tissue mass measuring 1.8x1.2x1 cm was submitted for histopathological examination. External surface of the tissue appeared grey brown with a grey white homogenous cut surface. Microscopical examination of the mass showed nests and trabeculae of relatively monomorphic cells with their nucleus showing a salt and pepper type of chromatin admixed with vascularized fibrous tissue. There was no increase in mitosis / necrosis. Structures resembling pseudo rosettes were seen focally (Fig.1). Reticulin stain showed the lesion to be reticulin poor. A differential diagnosis of a pituitary adenoma, peripheral neuroectodermal tumor, olfactory neuroblastoma, lymphoma and nasopharyngeal carcinoma were considered.

Immunohistochemistry was done for pan cytokeratin, synaptophysin, chromogranin, NSE, CD99, and LCA.

The tumor cells showed positive staining for vimentin, synaptophysin, chromogranin and focally positive for S100 (Fig. 2). Negative staining for pan cytokeratin, EMA, CD45, CD99 and LCA. Further IHC testing for pituitary panel showed positive staining for FSH and ACTH. Other markers, namely prolactin, TSH, LH were tested negative. A diagnosis of extra sellar pituitary adenoma was made. Activity of the tumor was assessed using proliferative index marker Ki67 which showed 1% positivity. Serum hormonal levels were not available as the earlier clinical diagnosis was not a pituitary adenoma. Post surgical values of pituitary hormones were within normal limits.

The final diagnosis was as Aggressive chromophobe pituitary adenoma arising from the extrasellar region.

Discussion

This case highlights an unusual presentation of pituitary adenoma. Up to 2% of pituitary tumors have an intrasellar extension with extension into the sphenoid sinus, nasopharynx and nasal cavity and present as a nasal polyp

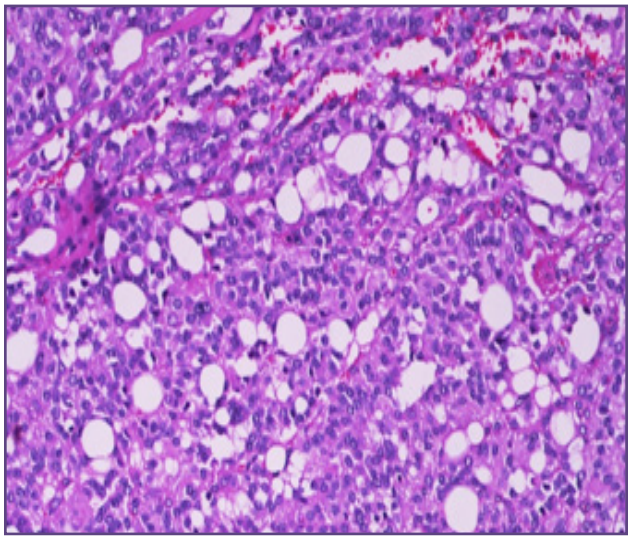

Fig. 1: Tumor showing sheets of tumor cells arranged in trabecular pattern with monomorphic cells with nucleus showing a salt and pepper type of chromatin admixed with vascularized fibrous tissue (20X) H & E.

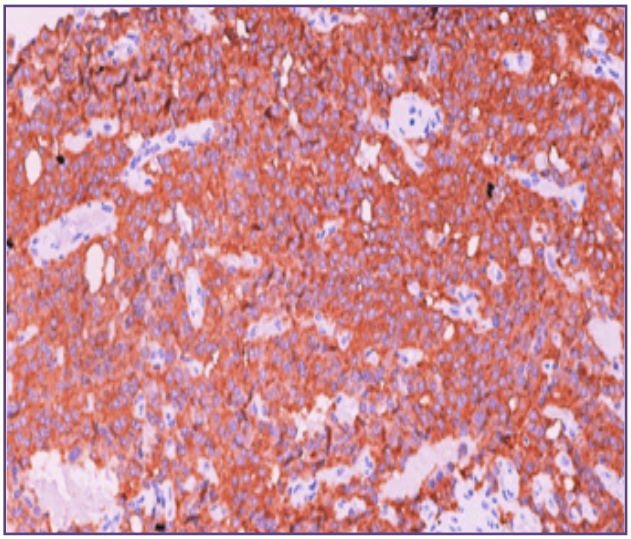

Fig. 2: Immunohistochemistry of tumor with synaptophysin (20X) IHC.

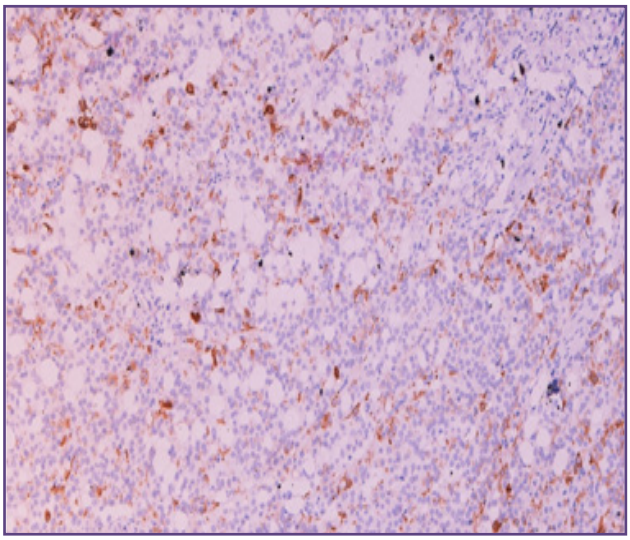

Fig. 3: Immunohistochemistry of tumor with FSH (20X) IHC

[2]. Rarely, they may arise in the sphenoid sinus with no continuity to the pituitary gland as Ectopic Sphenoid Sinus Pituitary Adenoma (ESSPA) [3,4]. Approximately 35 cases of nasopharyngeal invasion of pituitary adenomas have been reported. The initial cases of pituitary adenoma presenting as nasal polyp was identified by Sir Harvey Cushing in 1912[5]. Most of the cases reported are chromophobe adenomas followed by few macroprolactinomas [6] and one case of somatotroph and corticotroph adenoma.

Diagnosis in case of the extrasellar pituitary adenomas is always a challenge as most of these tumors are clinically silent and present as mass lesions with presentation as polyps, local destructive effects resulting in epistaxis, nerve palsies or hypopituitarism [7, 8]. Functional pituitary adenoma are usually rare in these locations and when present are not difficult as signs of PRL, GH TSH [9] or ACTH excess would be obvious. Diagnosis becomes especially challenging in cases where a pituitary adenoma is nonfunctional and has an exclusive intrasellar spread when it would mimic a nasopharyngeal mass [10].

Rarely, an ectopic pituitary tissue is located in extracranial sites like sphenoid sinus, nasal cavity, and nasopharynx [11]. In 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

Histological examination is usually helpful in providing the first clue 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. Differential diagnosis on histology includes small cell carcinoma, non-Hodgkin′s lymphoma, and plasmacytoma. Immunohistochemically positivity with neuroendocrine markers, and pituitary hormones in the tumor cells helps in confirming the diagnosis.

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 trans sphenoidal or endoscopic transnasal surgery may be used to treat with choice of therapy guided by surgeon′s experience.

Conclusion

In conclusion, pituitary adenomas presenting as nasopharyngeal masses either as an extension from an aggressive sellar pituitary tumor or ectopic ESPPA should be a part of differential diagnosis of Sino nasal masses. The diagnosis needs a coordinated effort from the otolaryngologists, radiologists and pathologists. Detailed endocrine evaluation with special emphasis on features of hypopituitarism, pituitary hormonal excess effects, space occupying pressure effect should be part of an evaluation of tumors in the sphenoid sinus. Proper accurate preoperative diagnosis shall have a profound effect on management and follow up of such cases in view of the difference in approach to management of pituitary adenomas and nasopharyngeal tumors.

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