Ectopic non functional pituitary adenoma – A case report

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ABSTRACT

Ectopic pituitary adenoma comprises 1–2% of all pituitary adenomas with a slight female predominance. Cases have been reported in the sphenoid sinus, cavernous sinus, parasellar region, nasal cavity, clivus, nasopharynx, and third ventricle. Here, we report the case of a 48-year-old female patient who presented with a 2 years history of nasal obstruction, epistaxis, and headache. The patient underwent necessary hematological, biochemical, and radiological investigations. Magnetic resonance imaging brain with angiography study showed a large soft-tissue mass in nasopharynx eroding the clivus and extending to involve bilateral sphenoid sinuses and parasellar region. The airway was also compromised. The pituitary gland appeared normal. All hematological and biochemical investigations were within normal limits. An endoscopic biopsy was also done. Tissue was submitted for histopathological examination and a diagnosis of pituitary adenoma was given. The patient had uneventful post-operative recovery and regularly came for follow-up.

Key words: *Ectopic, Nasopharynx, Pituitary adenoma, Pituitary gland*

Pituitary adenomas are common tumors present in the sella turcica. The location of this tumor outside this region is a rare condition. Ectopic pituitary adenoma comprises 1–2% of all pituitary adenomas with a slightly higher female predilection. The first case of ectopic pituitary adenoma was reported in 1909 by Austrian pathologist, Jacob Erdheim [1]. The location of ectopic pituitary adenoma includes suprasellar region, sphenoid sinus, petrous temporal bone, clivus, hypothalamus, parapharyngeal area, parasellar, and third ventricle [2].

We present the case of a 48-year-old female, who presented with epistaxis and nasal obstruction for 2 years. Imaging revealed lesion in nasopharynx extending to the sphenoid sinus and parasellar region. The correct diagnosis of ectopic pituitary adenoma can be made by histopathological examination. It is important to accurately diagnose this entity, especially when non-functional and located in a nasopharyngeal location for appropriate management.

CASE REPORT

A female patient, aged 48 years, was apparently asymptomatic 2 years back when she came to the outpatient department with chief complaints of nasal obstruction, difficulty in breathing, snoring, and sneezing associated with nasal discharge. She had epistaxis 2–3 times a week and the left-sided frontal headache. There was no history of excessive sweating, increased facial or body hair, stretch marks, easy bruising, weight gain, insomnia, or swelling in extremities. The patient gave a negative history of any trauma or hormonal disturbance.

On examination, the vitals were stable. The thyroid profile, serum insulin-like growth factor-1, serum 8 am cortisol levels, serum luteinizing hormone, serum follicular-stimulating hormone, and serum prolactin assays were within normal limits for age. Serum potassium level was 3.9 mEq/L (3.5–5.0).

Non-contrast computerized topography paranasal sinuses showed the presence of a large ill-defined soft-tissue mass in the base of the skull extending to sphenoid sinuses, bilateral choanal, abutting the hard palate, causing erosion of the clivus, dorsum sellae, bilateral pterygoid plates, and posterior surface of the vomer. Contrast-enhanced magnetic resonance (MR) imaging brain and neck revealed a large, lobulated, soft-tissue signal intensity mass of size ~ 4.6 (anteroposterior) × 5.5 (transverse) × 3.4 (craniocaudal) cm almost isointense to T1/T2/fluid-attenuated inversion recovery weighted images arising from the clivus, extending into nasopharynx significantly occluding the airway (Fig. 1), causing erosion of pterygoid plates, basisphenoid bone with thinning of floor of the sella, and extending in bilateral sphenoid sinuses. However, the pituitary gland appeared normal. MR angiography of cerebral vessels showed normal flow-related enhancement of major intracranial arteries (Fig. 2).

Multiple brownish soft-tissue pieces were received for histopathological examination altogether measuring 4 cm × 3 cm × 1 cm. Whole tissue was processed, embedded and blocks were made. H and E stained sections examined revealed

fibrocollagenous and bony tissue bits. A few bits were lined by respiratory epithelium. There was a cellular infiltrating neoplasm comprising monomorphic tumor cells arranged in organoid, solid, and trabecular patterns in the submucosal region and in intertrabecular space (Fig. 3). The tumor cells had round to oval nuclei with dispersed chromatin, inconspicuous nucleoli, and granular eosinophilic cytoplasm. Some cells had vacuolated cytoplasm. Minimal nuclear pleomorphism was seen. Mitosis/necrosis was not observed. In focal areas, the morphology was obscured. Chronic inflammatory infiltrates, a few seromucinous glands and dilated congested blood vessels were seen in the stroma (Fig. 4). Hence, a diagnosis of ectopic pituitary adenoma was given. The immunohistochemical study showed a few tumor cells to be adrenocorticotropic hormone (ACTH) positive.

Endoscopic transseptal excision of pituitary adenoma was done. The patient had uneventful post-operative recovery and comes for regular follow-up.

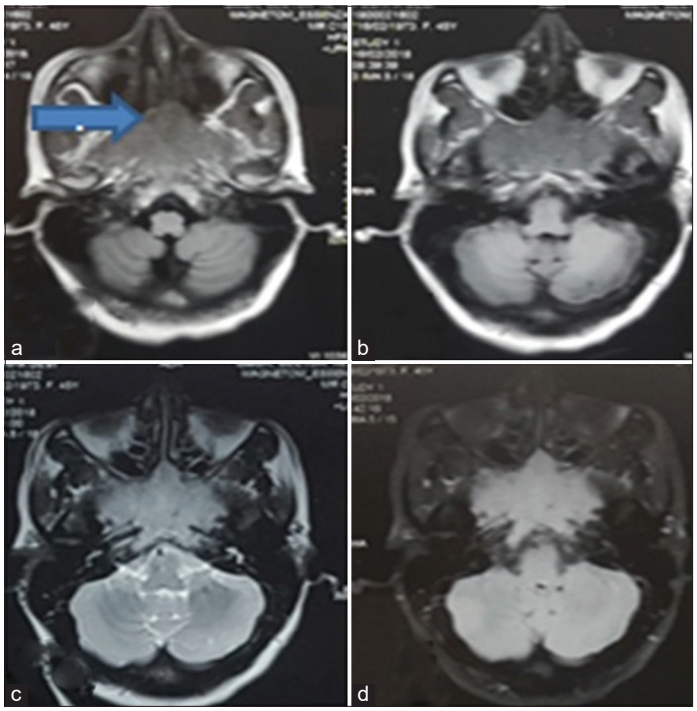

Figure 1: Magnetic resonance imaging axial T1 (a, b), T2 (c), and fluid-attenuated inversion recovery (d) images showing large soft-tissue isointense mass lesion arising from the clivus and extending into nasopharynx

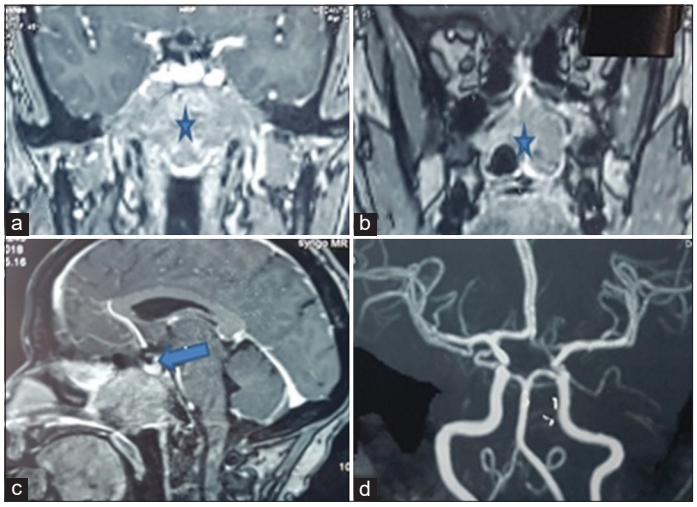

Figure 2: Contrast-enhanced MR coronal (a, b) and sagittal (c) images showing enhancing mass in nasopharynx (asterisk in a), nasal cavity (asterisk in b), abutting sellar floor with intact pituitary (arrow in c) with maintained flow in major intracranial arteries (d)

DISCUSSION

The World Health Organization defines ectopic pituitary adenoma as a benign pituitary neoplasm occurring separately from and without the involvement of sella turcica [3]. An ectopic pituitary adenoma is often located in nasopharynx, where it arises after neoplastic transformation in the pharyngeal pituitary. Pharyngeal pituitary arises from embryonic craniopharyngeal duct and remnants of Rathke's pouch [4].

The first case of ectopic pituitary adenoma was reported by Austrian pathologist, Jakob Erdheim [1]. Hori, on the autopsy of adult and fetal meninges, found the evidence of ectopic adenohypophysis. They have even described the entity of the pharyngeal pituitary [5]. Fuller and Batsakis found that it results from incomplete obliteration of the craniopharyngeal duct after the migration of Rathke's pouch [6]. Thompson *et al*. studied 32 cases of sphenoid sinus ectopic pituitary adenomas and found that certain ectopic pituitary adenoma has some

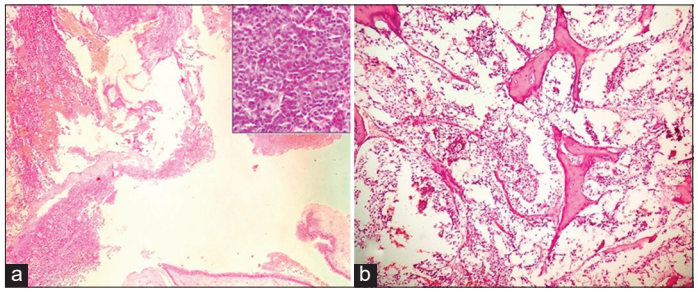

Figure 3: Photomicrograph showing (a) tumor cells in sheets along with respiratory epithelium; (H and E, ×100). Inset shows monomorphic tumor cells (H and E. ×400); (b) tumor cells scattered in intertrabecular area (H and E, ×100)

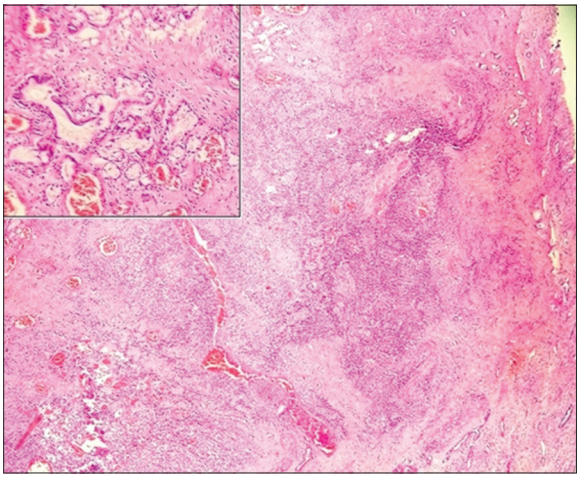

Figure 4: Photomicrograph showing infiltrating tumor cells (H and E, ×100). Inset shows seromucinous glands and dilated congested blood vessels (H and E, ×400)

atypical features such as oncocytic changes and prominent eosinophilic cytoplasm that are different from intrasellar pituitary adenoma [7].

Diagnosis of ectopic pituitary adenoma is difficult and sometimes it is impossible before surgery. Moreover, the differential diagnosis of nasopharyngeal and sphenoid sinus mass (sinonasal tract) is broad and includes typical carcinoids, atypical carcinoids, paraganglioma, small cell carcinoma, Ewing sarcoma, olfactory neuroblastoma, mucosal melanoma, and ectopic pituitary adenoma. Careful microscopic examination of these tumors along with immunohistochemical analysis is important to make an accurate diagnosis of ectopic pituitary adenoma.

Nasopharynx and sphenoid sinus pituitary can be fully functional since the pharyngeal pituitary begins to produce hormones around the 17–18th weeks of gestation, about 8 weeks late than intrasellar pituitary adenoma. Knowledge of the development of pituitary helps us to concede the diagnosis of ectopic pituitary adenoma at an abnormal site [8,9]. Approximately 58% of the patients present clinically with abnormal hormone production resulting in Cushing syndrome, acromegaly, or amenorrhea/galactorrhea [10]. In the present case, the patient did not have any obvious hormone-related symptoms and all the hormonal assays were within normal limits, and hence, the presenting case was diagnosed as a non-functional ectopic pituitary adenoma. However, the immunohistochemical study revealed positive staining for ACTH. This could be due to the early detection and removal of the tumor before the hormone could have its clinical manifestations.

CONCLUSION

Ectopic pituitary adenoma is an exceedingly rare neoplasm. Only a few cases have been reported in the literature. They are a diagnostic challenge when presenting with atypical clinical features along with the abnormal locations. It should be considered as one of the differential diagnoses while evaluating the nasopharyngeal masses. Clinicians and pathologists should be aware of this rare entity and its atypical clinical presentation, location, and pathological features for appropriate management.

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