International Journal of Otorhinolaryngology and Head and Neck Surgery
Singh VP et al. Int J Otorhinolaryngol Head Neck Surg. 2020 Nov;6(11):2146-2149
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20204651

Case Report

Ectopic pituitary adenoma: a rare and unexpected diagnosis, clinical presentation and review of the literature

V. P. Singh[1], Pankaj Arora[2], Alessia Rubini[3], Chetan Bansal[4]*, Arvind Varma[5]

[1]Department of ENT, Max Hospital, Dehradun, Uttarakhand, India
[2]Department of Neurosurgery, [5]Department of ENT, SGRRIM&HS, Dehradun, Uttarakhand, India
[3]Department of ENT, University of Verona, Verona, Italy
[4]Department of ENT, ONGC Hospital, Dehradun, Uttarakhand, India

Received: 07 September 2020
Revised: 14 October 2020
Accepted: 16 October 2020

***Correspondence:**
Dr. Chetan Bansal,
E-mail: drchetanent@yahoo.in

ABSTRACT

Nasal obstruction can be caused by many causes. Most common cause of nasal obstruction is deviated nasal septum followed by nasal masses. Nasal masses can be polyps, tumors or foreign bodies. We are presenting a case of ectopic pituitary tumor as a as cause of nasal obstruction along. Ectopioc pituitary tumors are rare tumors and can present in nasal cavity causing nasal symptoms. They can cause unilateral or even bilateral nasal obstruction depending on their spread in nasopharynx and nose and should be kept in mind while treating a case of nasal masses.

Keywords: Pituitary tumors, Nasal masses, Ectopic, Macroadenoma

INTRODUCTION

Ectopic pituitary tumors are rare tumors exclusively found outside the sella turcica.[1] Embryologically, the anterior pituitary is derived from an outpouching of the roof of the primordial oral cavity, Rathke's pouch.[1] These rare tumors seem to derive from cells of the Rathke pouch during its migration in embryogenesis.[1] In the literature are present only 8 cases of ectopic pituitary tumor located in the nasopharynx. Current report is a case of ectopic pituitary adenoma found in the nasopharynx with extension to the clivus, parasellar regions and infratemporal fossa, bilaterally. The description of the pathological findings, radiologic studies and the surgical management are described.

CASE REPORT

History and examination

A 45 year old woman presented with an occasional nasal bleeding for 10 years and progressive bilateral nasal obstruction for 2 years. Regarding the past medical history, she underwent a percutaneous transvenous mitral commissurotomy in 2006, so she was taking anticoagulant therapy (nicoumalone).

She was evaluated at the ENT department, and a nasopharynx mass was rule out during nasal endoscopy. The mass was covered by pinkish smooth mucosa covered and filled the rinopharinx, with posterior choana obstruction bilaterally (Figure 1). Therefore, the patient was taken for biopsy of the mass, under general

anesthesia, following a radiological study with a non-contrast computed tomography (NCCT) of paranasal sinuses and a brain MRI with gadolinium contrast.

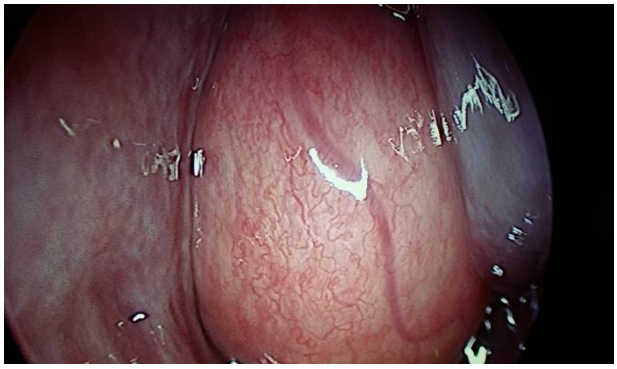

Figure 1: Endoscopic transnasal view with a zero degree endoscope: a pinkish mass was detected in the nose extending to nasopharynx, obliterating completely the choana.

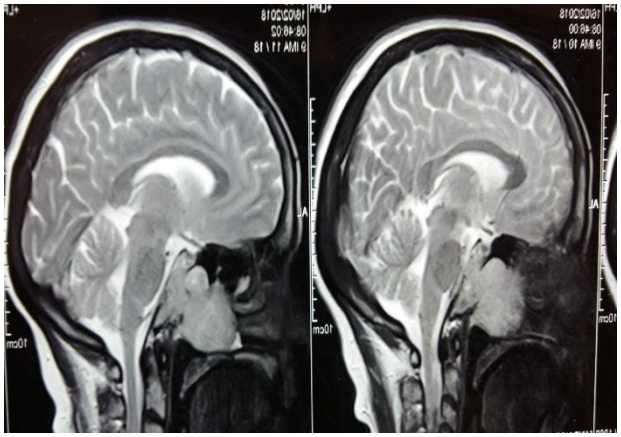

Figure 2: Contrast enhanced magnetic resonance imaging of the paranasal sinuses sagittal view. The ectopic pituitary adenoma is located in the posterior nasal fossae, nasopharynx with extension in the sphenoidal sinuses and clivus with internal carotid artery involvement.

Radiology

Contrast enhanced magnetic resonance imaging (Figure 2 and Figure 3) and a non-contrast computed tomography (NCCT) scan (Figure 4) of the paranasal sinuses demonstrated the presence of large lobulated well defined homogeneous soft tissue mass lesion measuring 4.6x5.5x3.4 cm (APxMLxCC) in the base of skull extending superiorly involving bilaterally the sphenoid sinus, posteriorly causing destruction of the clivus, anteriorly entering into choanae bilaterally and inferiorly abutting the hard palate, entering bilaterally parasellar regions eroding pterygoid roots and medial part of basi sphenoid bone (Figure 2, Figure 3, and Figure 4). Thinning of the sella was seen, however pituitary glands appeared normal. At the MRI the mass lesion appeared almost isointense to gray matter on T1/FLAIR/T2 weighted images and on post contrast scans, it showed diffuse homogeneous enhancement.

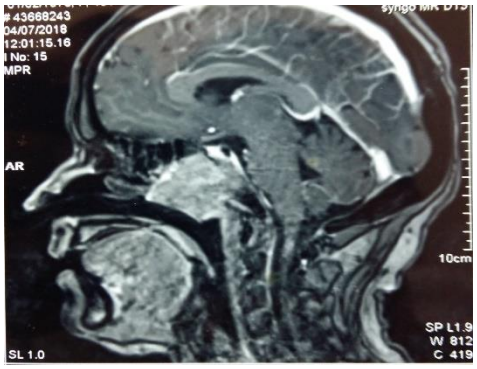

Figure 3: Non contrast computed tomography (NCCT) scan of the paranasal sinuses, the tumor is showed in a coronal view, the pterygoid plates erosion is seen bilaterally.

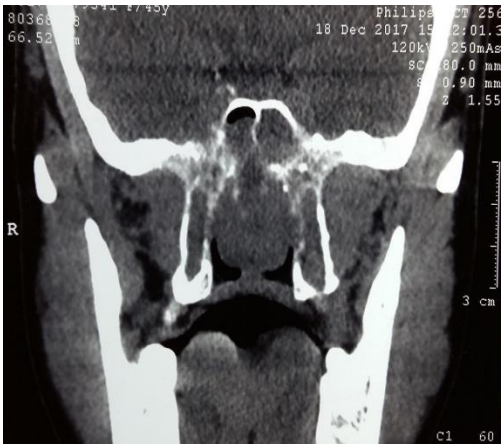

Figure 4: Contrast-enhanced magnetic resonance imaging of the paranasal sinuses, saggittal view of the ectopic pituitary adenoma.

Pathological findings

Unexpectedly, the histological result of the biopsy tissue showed presence of tumor comprising monomorphic tumor cells arranged in organoid, solid and trabecular growth patterns. The individual tumor cells were plasmocytoid and are relatively monomorphic having round to oval nuclei with dispersed chromatin, inconspicuous nucleoli and abundant eosinophilic granular plasmacytoid cytoplasm. Mitosis is infrequent. No necrosis was noted. Morphologic features were pituitary adenoma. Tumor cells were positive for ACTH expression on immunohistochemistry, while negative for GH and PRL (Figure 5).

Surgery and post-operative course

The patient was scheduled for surgical excision of the nasopharynx ectopic pituitary adenoma, through a

transnasal transethmoidal endoscopic approach. Using a zero degree endoscope an antero-posterior ethmoidectomy was performed and the mucosa of the posterior portion of the septum was removed. The tumor arised from the posterior nasal septum and extended towards the nasopharynx and the pterygopalatine fossa bilaterally. The tumor was removed endoscopically along with the posterior portion of the nasal septum. The rostrum and the intersphenoid sinus septum were drilled, opening the sphenoid sinus, which was free from disease. The medial pterygoid plates were also removed in order to obtain a good surgical removal laterally, then the tumor was dissected posteriorly. Intraoperatively, erosion of the clivus posteriorly was noted, without any interruption of the bone.

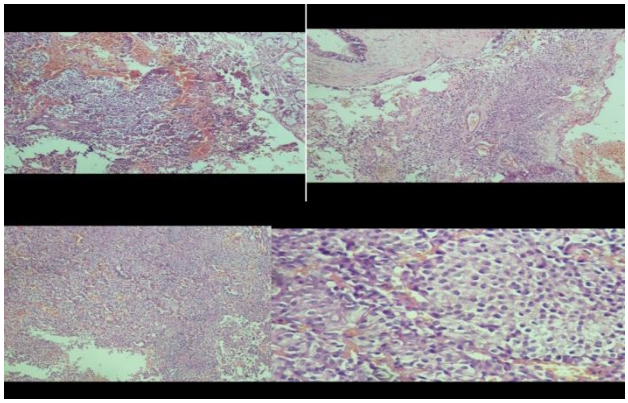

Figure 5: Histopathology slides, morphologic features suggestive of pituitary adenoma.

The post-operative course was regular and no intra- and post-operative complications were observed. The final histologic exam confirms the diagnosis of pituitary adenoma. The patient after 8 months of follow up presents regular result of surgery, no recurrence of disease at the endoscopic examination. An MRI is planned to rule out any residual disease.

DISCUSSION

Ectopic pituitary adenoma is defined as a pituitary adenoma occurring outside of the sella turcica, without any direct connection to either the intrasellar gland or the pituitary stalk.[1] The most common locations are the sphenoid sinus, clivus, suprasellar space, nasopharynx, and cavernous sinus.[2]

The majority of ectopic pituitary tumors are reported as being functional secretory tumors. Nowadays, in total 85 cases of ectopic pituitary tumors are described in literature.[2] Out of these 85, 6 were classified as malignant or as having undergone a malignant transformation. In the majority of the cases (59% of total) are functional tumors (featured hormonal expression on immunohistochemistry staining and were found to be clinically active (caused hormonal symptoms in the patient), and ACTH producing tumors were the most common subtype (46%).[1]

The origin of ectopic pituitary tumors is not clear; nowadays, the accepted hypothesis suggests that the tumors are derived from cells of the Rathke's pouch during its migration in embryogenesis.[3,4]

The anterior pituitary is derived from an outpouching of the roof of the primordial oral cavity, Rathke's pouch. The pouch during embryogenesis, ascends from the pharyngeal roof to join the neurohypophysis. It then loses its attachment to pharyngeal roof when its stalk ruptures. While the posterior pituitary is derived from an outpouching of the diencephalon. The deposition of residual adenohypophysis cells in the infrasellar region, occurs by partial persistence of the pouch in the wall of the buccal cavity, craniopharyngeal canal, or sphenold sinus.

On the other hand, aberrant suprasellar adenohypophysis may originate later when the pars tuberalis develops from the pars anterior of the pituitary. The adenohypophysis originates extracranially and only later becomes attached to the neurohypophysis, so the pituitary cells may easily be displaced in the leptomeninges of the peri-infundibular region. Hori analyzed the routine autopsy examination of all fetal brains older than the 16th gestational and "normal" adult brains and he found that pharyngeal location of pituitary cells is a constant finding and almost always present in the leptomeninges of the suprasellar region. Therefore, it seems that the origin of extracranial ectopic tumor, might arise from extracranial pre-existent normal aberrant pituitary tissue while intracranial ectopic tumor might arise from suprasellar normal aberrant pituitary tissue.[5] Anyway, the criteria for diagnosing ectopic adenoma require detailed histopathological examinations including immunocytochemical and ultrastructural studies.[6]

We presented a case of ectopic giant pituitary adenoma with positive staining for ACTH, located into the nasopharynx extending to the clivus, the sphenoid sinuses and parasellar regions. Nasopharinx and clivus are the rarest locations for an ectopic pituitary adenoma, only 14 cases in the clivus are described in literature and 8 cases in the nasopharynx. Moreover, in our case, because of the dimensions of the mass, it may be considered as a giant ectopic pituitary adenoma. Giant pituitary adenoma is defined as tumors 4 cm or greater in maximum diameter, are very rare, account for 5%-14% of adenomas in surgical series.[7-9]

The clinical presentation of this lesion was related to the location and size of the mass, since the patient presented with bilateral nasal obstruction and occasionally epistaxis, without hormonal signs and/or symptoms. Radiologically, ectopic pituitary adenomas can mimic other skull base lesions. MRI is useful in evaluating the

extension of the extrasellar mass, the integrity of the sellar dura and the presence of normal pituitary gland.[10]

Radiologically and intraoperatively, in the reviewed case, the mass presented bony erosion, as described in the literature that the common ectopic pituitary adenoma locations with bone involvement are the clivus and nasopharynx. Because of these radiological characteristics, in our case the differential diagnosis at the MRI and the CT scan were chordoma, chondrosarcoma, plasmocytoma, nasopharyngeal carcinoma or methastasis. The mass was isointense to gray matter on T1/FLAIR/T2 weighted images and on post contrast scans, it showed diffuse homogeneous enhancement. Chondrosarcoma typically shows at the MRI heterogeneously enhanced mass with high signal intensity on T2 and at the CT scan show a chondroid mineralization.

Plasmacytoma is a lytic lesion iso-hypointense to gray matter on T1, homogeneous, isointense on T2, homogeneous, iso or hyperintense to gray matter on FLAIR. While chordoma is a midline mass originating from the clivus hyperintense on T2WI and intermediate to low signal on T1 with a mixed density with areas of cystic necrosis mixed with enhancing soft tissue at the contrasted enhanced CT scan.

The main management of the ectopic pituitary adenomas are the surgical removal, because in the literature malignant ectopic pituitary adenoma or as having undergone a malignant transformation are described, although in very rare cases. The surgical procedures varied according to the location and the extensions. We choose to remove the ectopic pituitary adenoma with a gross total resection, through an endoscopic transnasal transethmoidal approach because of the location and extension of the tumor that no contraindicated the exclusive endoscopic procedure. In our case, since the giant adenoma was ectopic, the main goal of the surgery was a maximal safe tumor resection to relief the symptoms avoiding residual disease.

CONCLUSION

Ectopic pituitary tumors are rare tumors that can present as nasal mass. These should also be kept as differential diagnosis while evaluating nasal masses.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Seltzer J, Lucas J, Commins D, Lerner O, Lerner A, Carmichael JD, et al. Ectopic ACTH-secreting pituitary adenoma of the sphenoid sinus: case report of endoscopic endonasal resection and systematic review of the literature. Neurosurg Focus. 2015;38 (2):E10.
2. Shuman W, Loewenstern J, Pai A, Bederson J, Shrivastava R. Variability in clinical presentation and pathologic implications of ectopic pituitary tumors: critical review of literature. World Neurosurg. 2019;122:397-403.
3. Appel JG, Bergsneider M, Vinters H, Salamon N, Wang MB, Heaney AP. Acromegaly due to an ectopic pituitary adenoma in the clivus: case report and review of literature. Pituitary. 2012;15(Suppl 1):S53-6.
4. Hong JF, Ding XH, Wang SS. Coexistence of ectopic pituitary adenoma and empty sella in a patient with acromegaly: a case report and review of literature. Neurol India. 2012;60(3):304-6.
5. Hori A. Suprasellar peri-infundibular ectopic adenohypophysis in fetal and adult brains. J Neurosurg. 1985;63(1):113-5.
6. Matsuno A, Katayama H, Okazaki R, Toriumi M, Tanaka H, Akashi M, et al. Ectopic pituitary adenoma in the sphenoid sinus causing acromegaly associated with empty sella. ANZ J Surg. 2001; 71(8):495-8.
7. Goel A, Nadkarni T, Muzumdar D, Desai K, Phalke U, Sharma P. Giant pituitary tumors: a study based on surgical treatment of 118 cases. Surg Neurol. 2004;61(5):436-45;
8. de Paiva Neto MA, Vandergrift A, Fatemi N, Gorgulho AA, Desalles AA, Cohan P, Wang C, Swerdloff R, Kelly DF. Endonasal transsphenoidal surgery and multimodality treatment for giant pituitary adenomas. Clin Endocrinol (Oxf). 2010;72 (4):512-9.
9. Mortini P, Barzaghi R, Losa M, Boari N, Giovanelli M. Surgical treatment of giant pituitary adenomas: strategies and results in a series of 95 consecutive patients. Neurosurgery. 2007;60(6):993-1002
10. Ali R, Noma U, Jansen M, Smyth D. Ectopic pituitary adenoma presenting as midline nasopharyngeal mass. Ir J Med Sci. 2010;179(4): 593-5.

Cite this article as: Singh VP, Arora P, Rubini A, Bansal C, Varma A. Ectopic pituitary adenoma: a rare and unexpected diagnosis, clinical presentation and review of the literature. Int J Otorhinolaryngol Head Neck Surg 2020;6:2146-9.