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Brazilian Journal of
OTORHINOLARYNGOLOGY

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CASE REPORT

Right ectopic sphenoid sinus pituitary adenoma[☆]

Adenoma hipofisário ectópico de seio esfenoidal direito

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Received 24 November 2012; accepted 21 April 2013

Available online 3 July 2014

Introduction

Adenomas are the most common pituitary tumors, corresponding from 10% to 20% of all brain tumors.¹ Occasionally, these tumors can extend out of the pituitary fossa, and, on rarer occasions, they can also be found in ectopic sites, having uncertain origin and diverse location.²

Ectopic pituitary adenomas are clinically detected because of their local mass effect and/or hormone hypersecretion.² Paranasal sinus computed tomography and magnetic resonance imaging are used to study these conditions preoperatively, with diagnosis confirmed by histopathology and immunohistochemistry.³

Treatment includes surgical resection, which may or may not be associated with radiation therapy, and the prognosis is good.³

Clinical case

E.S., an 82-year-old female had a history of headache and nasal congestion for one year. She had previously undergone treatment for rhinosinusitis with no clinical improvement.

Upon presentation, a nasal mass filling the right nostril and the right aspect of the cavum was detected on endoscopic examination.

Computed tomography of the paranasal sinuses revealed a tumor occupying the right nostril and the right aspect of the cavum (Fig. 1).

Endonasal endoscopic surgery was conducted, and the entire tumor mass was resected from the right sphenoid sinus. The specimen was sent for histopathological examination, with inconclusive results. Immunohistochemical study confirmed that it was an ectopic pituitary adenoma. On follow-up, the patient has noted improvement of her symptoms.

Discussion

An ectopic pituitary adenoma is defined as a pituitary gland tumor located out of the sella turcica and having no connection with the intrasellar gland.⁴ They were described by Erdhelm in 1909, and may be found in the sphenoid sinus region, clivus, parapharyngeal space, nasal cavity and nasopharynx, hypothalamus, third ventricle, and in the suprasellar locations.^{3,4}

They are considered rare neoplasms originating from embryonic remnants of Rathke's pouch. Since they were first described, approximately 50 cases of ectopic pituitary adenoma have been reported, 62% of which occurred in women at a mean age of 50 years, most commonly located in the sphenoid sinus (40%) and in suprasellar sites (33%).⁴

* Please cite this article as: Brito LB, Tinoco P, Tinoco T, Ferreira FR, Carrara VL. Right ectopic sphenoid sinus pituitary adenoma. Braz J Otorhinolaryngol. 2014;80:451–2.

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Figure 1 Paranasal sinus tomography showing a sphenoid sinus tumor extending into the cavum.

According to medical literature, about one-third are endocrine-inactive tumors diagnosed as an examination finding or from their local effect. The remaining two-thirds have hormonal activity and usually secrete ACTH, eliciting a Cushing's disease picture, although they can also be associated with acromegaly and high blood prolactin.⁵

Diagnosis should be made through history, physical examination, paranasal sinus computed tomography and magnetic resonance imaging showing a soft-tissue density mass viewed in a paranasal cavity, as in the clinical case described, with no sellar abnormalities.⁵ In cases of suspected tumor endocrine activity, the patient should have additional tests, such as salivary cortisol estimation, ACTH, and CRH when Cushing's syndrome is suspected; serum random GH and IGF-1 tests when acromegaly is suspected; when considering hyperprolactinemia, serum prolactin and TSH should be performed.^{3,5}

Differential diagnosis includes chordomas, nasopharyngeal carcinoma, or a tumor derived from a minor salivary gland. However, a clivus lesion should be distinguished from a meningioma, an epidermoid cyst, fibrous dysplasia, and pituitary tumors.⁶

Management includes surgical resection via either transsphenoidal or transsphenoethmoidal approach to reach the sphenoid sinus in addition to transfacial and transnasal transmaxillary approaches to the clivus.^{5,6} Malignant transformation is unusual; in such cases, postoperative radiation therapy may be added if resection is incomplete.⁶

Final comments

The ectopic pituitary adenoma is of great importance, as it is a rare condition. Once diagnosis is made, complete clinical improvement can be achieved by surgical management, whether or not this is associated with radiation therapy.

Conflicts of interest

The authors declare no conflicts of interest.

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