

# Ectopic growth hormone-secreting pituitary adenoma involving the clivus treated with octreotide: role of magnetic resonance imaging in the diagnosis and clinical follow-up

Adenoma hipofisário ectópico secretor de hormônio de crescimento envolvendo o clivus tratado com octreotida: papel da ressonância magnética no diagnóstico e seguimento clínico

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Ectopic pituitary adenomas (EPAs) are rare neoplasms, mainly described in the sphenoid sinus (SS) and suprasellar region; hormone-secreting EPAs with clival involvement have been rarely reported. We describe a case of a pharmacologically treated GH-secreting EPA involving the clivus and SS, with satisfactory clinical outcome and tumor size reduction. As transsphenoidal resection is the standard approach, post-treatment imaging findings in the nonsurgical management of this entity, to our knowledge, have not been reported before.

## CASE REPORT

A 30-year-old woman presented with clinical signs and symptoms of acromegaly for the past one year. A hormonal profile confirmed acromegaly [plasma IGF-1 level >500 ng/mL (94–309 ng/mL), GH=218 ng/mL (<5 ng/mL) and prolactin=7 ng/mL (3–29 ng/mL)]; an oral glucose tolerance test revealed no suppression of GH values. With suspicion of a pituitary adenoma, a brain MRI was performed, which revealed, however, a 3.0 x 2.0 cm enhancing mass lesion on both T1- and T2-weighted images involving the clivus and SS, well demarcated from the normal pituitary, which was cranially displaced (Fig 1). After a careful exclusion of other rare causes of acromegaly, association of clinical, laboratorial and imaging findings were consistent with a GH-secreting EPA.

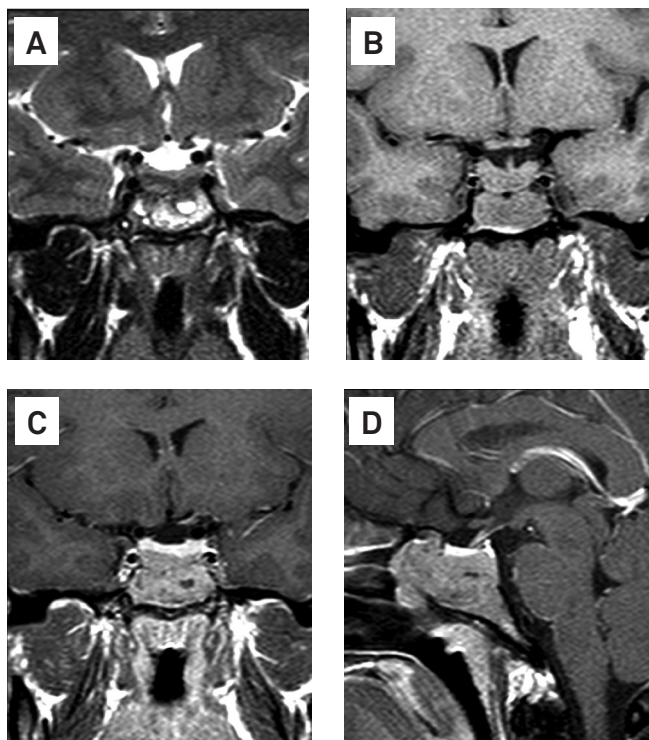
Despite appropriate medical counseling, the patient refused surgical treatment. Therefore, pharmacological

treatment with octreotide, a somatostatin analogue, was indicated in association with a close follow-up. The patient presented stabilization of IGF-1 levels and random measurements of GH showed safe levels <2.0 µg/L, confirming an adequate response<sup>1,2</sup>. Annual control MRI scans demonstrated a progressive tumor shrinkage with heterogeneous enhancement (probably indicating areas of necrosis) (Fig 2). The patient remains well and asymptomatic after three years of follow-up, still under pharmacological treatment.

## DISCUSSION

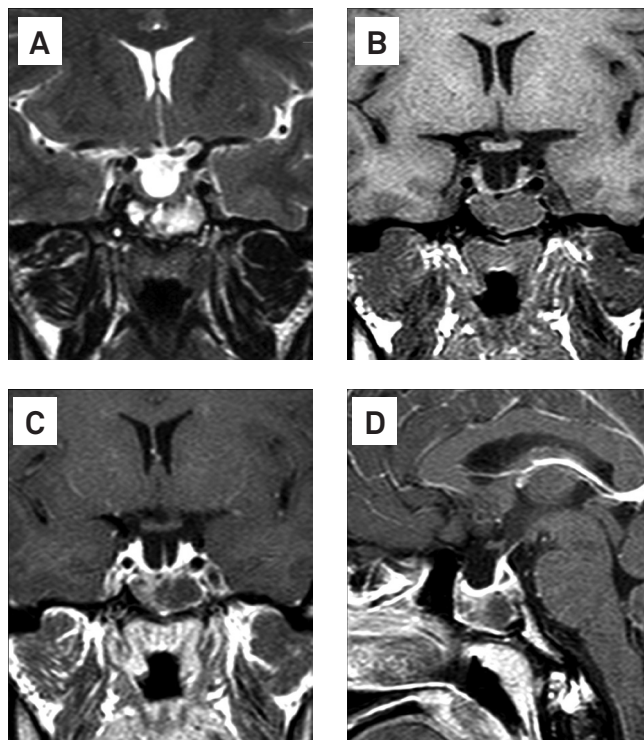
EPAs arise from the extrasellar region without any involvement of the normal pituitary gland in the sella turcica<sup>3</sup>. It has been suggested that EPAs could arise along the cranial migration route of Rathke's pouch toward the hypothalamus, after its development as an invagination of the primitive oral cavity during the third week of gestation; remnants of this pharyngopituitary canal in bone areas, for example, could explain EPAs in the clivus or sphenoid bone<sup>4,5</sup>.

Surgical resection of the adenoma causing acromegaly has been the traditional approach, although normalization of IGF-1 and GH levels is observed in only 40–70% of patients<sup>1</sup>. Although octreotide is not currently recommended as a primary therapy (except when surgical treatment cannot be performed)<sup>6</sup>, in several trials it has demonstrated to be as effective as surgery regarding the control of IGF-1 and GH levels, also inducing tumor shrinkage<sup>6</sup>.



**Fig 1.** T2- and T1-W pre-contrast coronal images (A and B) demonstrate a heterogeneous mass lesion containing cystic areas involving the clivus and sphenoidal sinus; post-contrast T1-W coronal (C) and sagittal images (D) show a homogeneous enhancement of the lesion, without evidence of involvement of the normal hypophyseal parenchyma.

In our case, cross-sectional imaging played a pivotal role in the diagnosis, despite the absence of a confirmatory histopathological study. With effect, pituitary adenomas can be classified on the basis of excessive hormonal secretion<sup>7</sup>. Subsequently, MRI contributed in the clinical



**Fig 2.** T2- and T1-W pre-contrast coronal images (A and B) obtained at the third year of follow-up demonstrate tumor size reduction; post-contrast T1-W coronal (C) and sagittal images (D) show a heterogeneous enhancement of the lesion, probably secondary to areas of necrosis, with reduction of the hypophyseal parenchymal thickness and partial filling of the sella turcica with cerebrospinal fluid (empty sella).

follow-up by evaluating tumoral response to pharmacological treatment. Finally, the good clinical response fortunately obtained and principally the tumor shrinkage after specific treatment with a somatostatin analogue favored our initial diagnosis.

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