ACTH secreting pituitary adenomas within the sphenoid sinus: an overview

Philip C Johnston¹, Laurence Kennedy¹, Amir H Hamrahan¹ Robert J Well²
Department of Endocrinology and Diabetes¹, Department of Neurosurgery², Cleveland Clinic, Ohio, USA

BACKGROUND

• In ACTH-dependent Cushing’s syndrome, ACTH can originate from a pituitary adenoma or ectopic ACTH syndrome, or rarely from an ectopic ACTH secreting pituitary adenoma (EAPA).

• We present a case of an EAPA within the sphenoid sinus and provide an overview of all previously reported cases in the literature.

CASE

• A 38-year old woman with a previous history of gastric bypass reported weight gain, hirsutism and bruising for 2 years.

• She appeared ‘classically’ cushingoid.

• 24 hr UFC 278 μg (0-50).

• ACTH 121 μg/ml (6-48).

• 8am serum cortisols of 17 μg/dL and 14 μg/dL after 1 mg overnight and 2 day low dose dexamethasone suppression test (DST), respectively.

• A >50% decrease in serum cortisol occurred after an overnight high dose (8mg) DST [pre:28.4 μg/dL, post:13.9 μg/dL].

• Inferior petrosal sinus sampling (IPSS) did not demonstrate a significant central to peripheral (C/P) ACTH ratio, of either 2:1 before, or 3:1 after administration of CRH. The test was interpreted as non-diagnostic due to the lack of proper IPS catheterization based on a C/P ratio of <1.3 for prolactin. ACTH response to CRH in peripheral blood during IPSS was >50%. CT C/A/P was unremarkable.

• CT of the head [Fig.1,arrow] revealed an incidental lesion in the right sphenoid sinus which was also present on MR [Fig.2,arrow] imaging and felt to be an incidental polyp, the pituitary gland appeared normal.

DISCUSSION

• Ectopic corticotroph adenomas are a rare but important cause of ACTH-dependent hypercortisolism.

• 21 EAPA cases in sphenoid sinus, reported to date.

• Macroadenoma:15, micro; 6, range 3-55 mm.

• Majority (20/21) demonstrated clinical evidence of hypercortisolism.

• In 12/21 cases, pre-operative imaging identified a sphenoid mass or ‘polyp’.

• IPSS performed in 9/21 cases, a C/P ACTH gradient was present in 8/9.

• 16/21 reported to be in remission after surgery.

CONCLUSION

Ectopic corticotroph adenomas can be a diagnostic challenge in patients with Cushing’s disease; failure to recognize one may lead to a sellar exploration that fails to find an adenoma and induce disease remission. The presence of a sphenoid ‘polyp’ as an etiology of ACTH dependent hypercortisolism should prompt the clinician not to overlook this often regarded ‘incidental’ radiological finding.