# ACTH secreting pituitary adenomas within the sphenoid sinus: an overview

Philip C Johnston[1], Laurence Kennedy[1], Amir H Hamrahian[1] Robert J Weil[2]

Department of Endocrinology and Diabetes[1], Department of Neurosurgery[2],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 pg/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 catherization 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 pitutary gland appeared normal.

## CASE

- At surgery, two small (3-4mm) pseudo-encapsulated lesions on the right side of the pituitary were removed which were negative for a corticotroph adenoma.
- A soft polypoid 5 mm lesion in the inferior aspect of sphenoid sinus was identified, removed and revealed pituitary adenoma tissue and demonstrated ACTH positivity on immuno-histochemical staining.
- The patient developed hypocortisolism post-operatively (nadir cortisol of 1.3 µg/dL) at 48 hours after surgery.
- At 44 months post-surgery, she remains in remission.

Fig.1

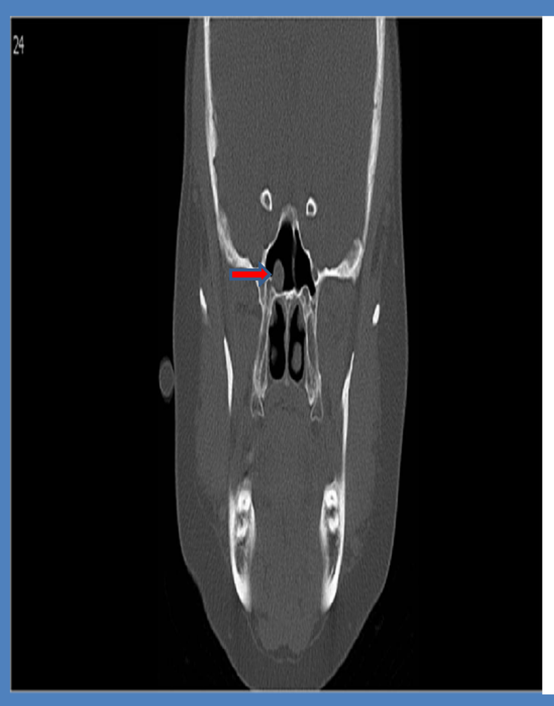
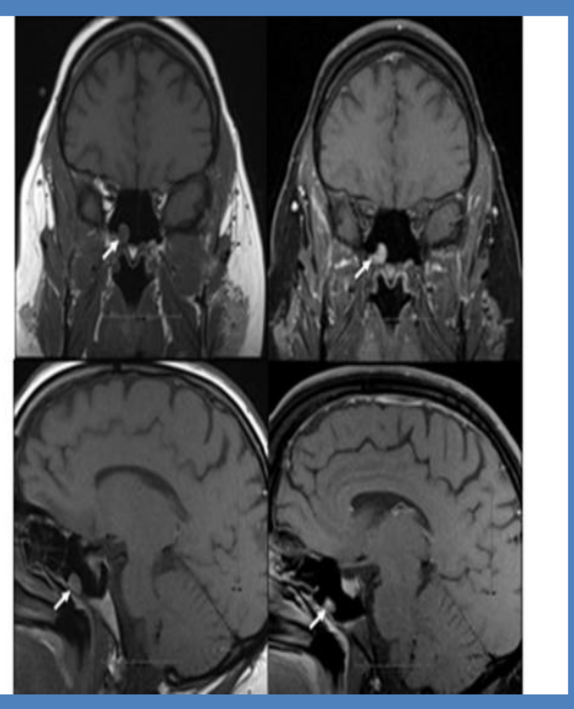


Fig.2



## 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. Johnston PC, et al Endocrine 2015

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Clinical Cases Pituitary/Adrenal

Philip Johnston

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