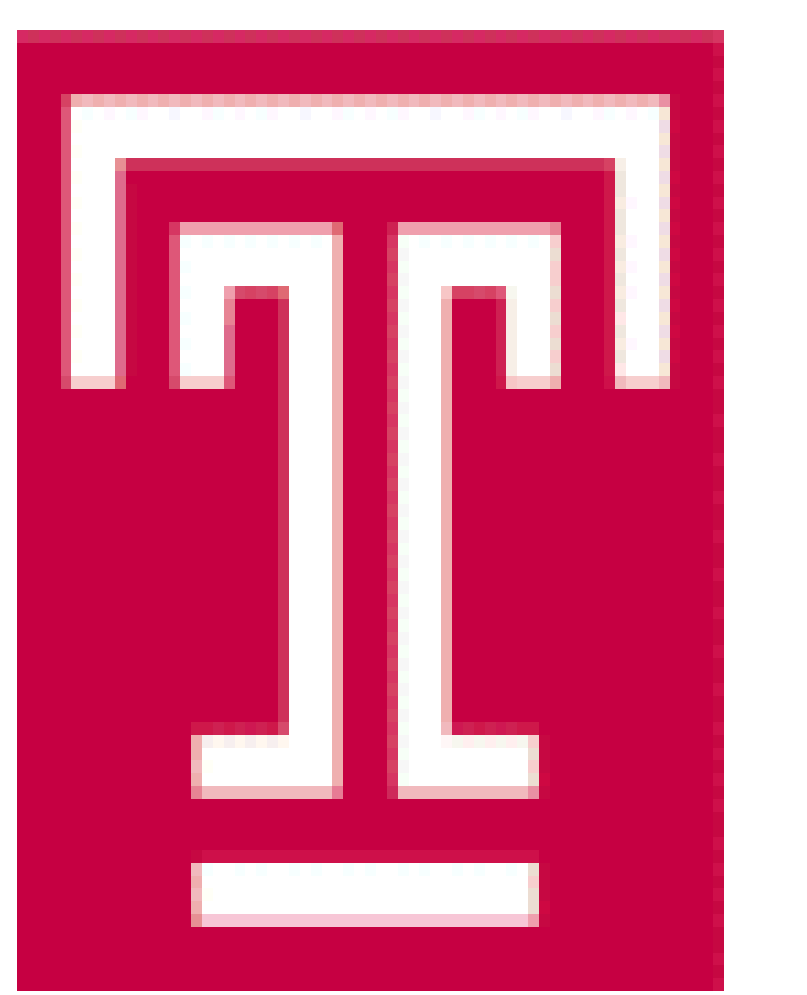


# CUSHING'S DISEASE WITH TWO ACTH-PRODUCING PITUITARY TUMORS

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## CASE DESCRIPTION

A 36 year-old female found to have left supraclavicular fossa swelling was screened for Cushing's syndrome

- Midnight salivary cortisol levels elevated at 0.636 ug/dL and 0.316 ug/dL (<0.010-0.090 ug/dL)
- 24-hour urine cortisol 162 ug/24 hr (0-50 ug/24 hr)
- 1-mg dexamethasone suppression test 14.0 ug/dL
- 8 AM cortisol 26.4 ug/dL, ACTH 66.7 pg/mL (7.2-63.3 pg/mL)
- MRI brain with and without contrast showed a 7-mm relatively hypoenhancing lesion of the anterior pituitary gland
- 8-mg dexamethasone suppression test 2.7 ug/dL

She underwent transsphenoidal surgery (TSS) and pathology was consistent with a pituitary adenoma staining positive for ACTH.

No residual tumor was seen

- Postoperative 8 AM cortisol 17.0 ug/dL and ACTH 79 pg/mL (9-46 pg/mL)

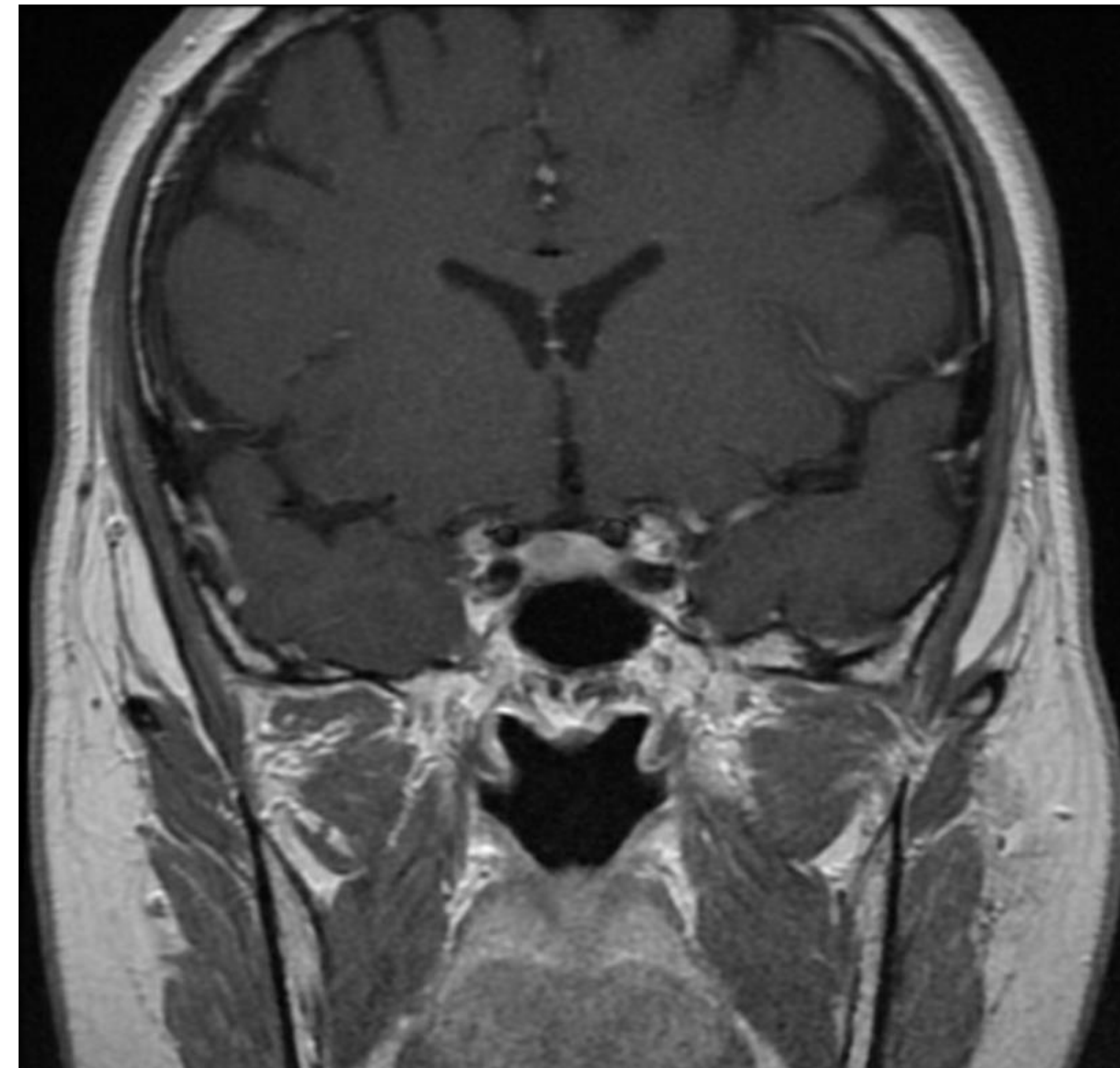
She had repeat TSS and the prior area of resection was clean with no residual tumor but a second adenoma was found that was not visualized on MRI and was distinct from the initial lesion

- Postoperative 8 AM cortisol 0.7 ug/dL and ACTH <9 pg/mL (9-46 pg/mL)
- Pathology consistent with pituitary adenoma staining positive for ACTH
- Now on steroids for central adrenal insufficiency

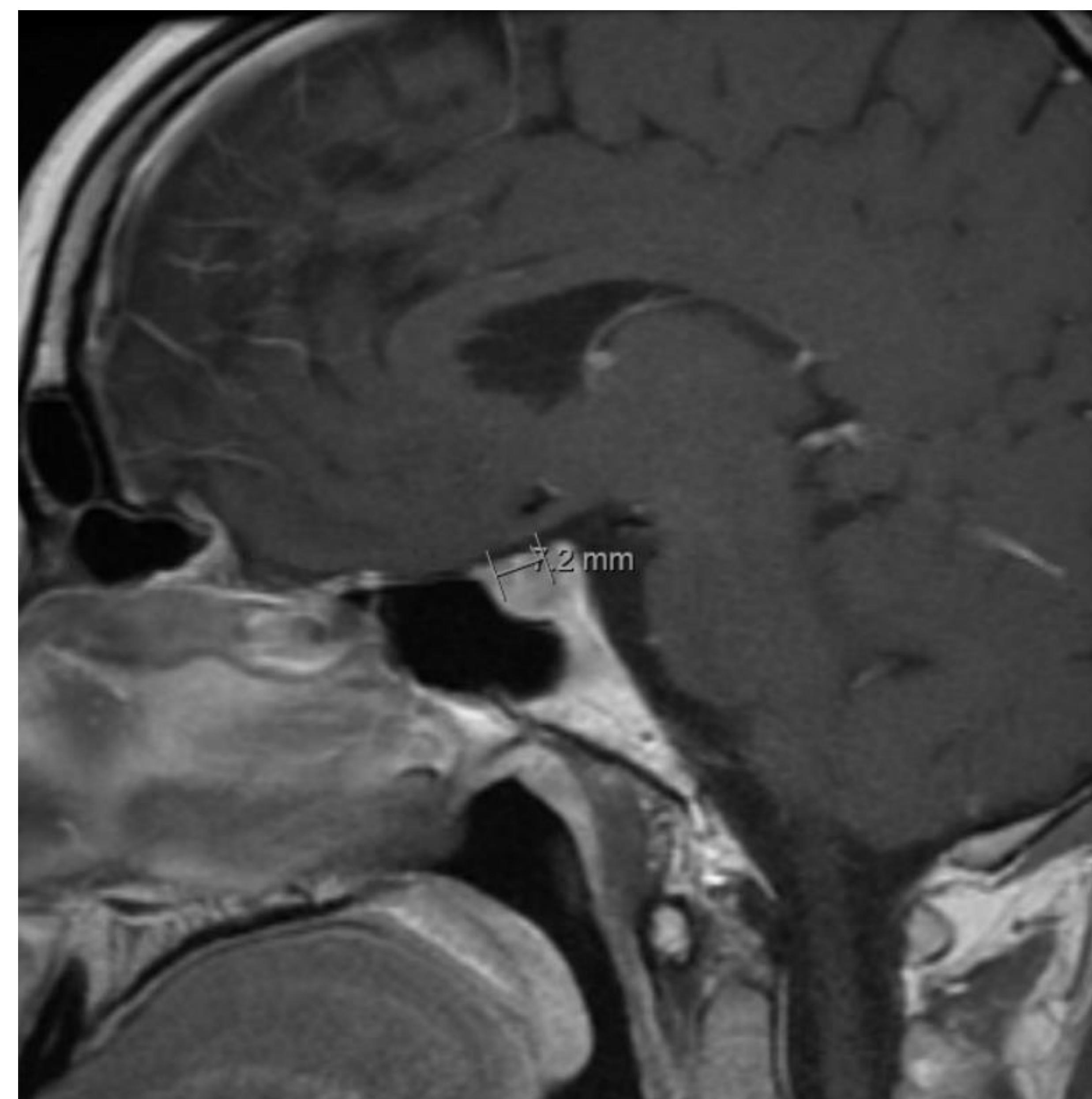
### Preoperative Hypercortisolism Evaluation

Midnight Salivary Cortisols (normal range <0.010-0.090 ug/dL)	0.636 ug/dL	0.316 ug/dL
24-hour Urine Cortisol (normal range 0-50 ug/24 hr)	162 ug/24 hr	
1-mg Dexamethasone Suppression Test (ug/dL)	14 ug/dL	

**Table 1:** Preoperative testing for hypercortisolism including 2 midnight salivary cortisol levels on 2 separate nights, 24-hour urine cortisol, 1-mg dexamethasone suppression test



(a)



(b)

**Figure 1:** MRI Brain with and without contrast showing a microadenoma. It measures 6 x 4 mm in coronal dimension and 7 mm in AP dimension. (a) coronal view and (b) sagittal view

### ACTH and Cortisol Response Post Transsphenoidal Surgery

	Preoperative	After 1st TSS	After 2nd TSS
ACTH (pg/mL)	66.7 pg/mL	79 pg/mL	<9 pg/mL
Cortisol (ug/dL)	26.4 ug/dL	17 ug/dL	0.7 ug/dL

**Table 2:** ACTH and 8 AM cortisol tests before surgery and after first TSS when they did not decrease as expected. And finally ACTH and cortisol after second TSS when they decreased appropriately. ACTH (pg/mL, normal range 7.2-63.3 pg/mL), cortisol (ug/dL)

## DISCUSSION

First-line treatment for Cushing's disease is surgical resection of the primary lesion<sup>1</sup>. Remission rates are 73-76% for selectively resected microadenomas but 43% for macroadenomas<sup>1</sup>. For patients who undergo a noncurative surgery, second-line therapies include repeat TSS, radiotherapy, medical therapy, and bilateral adrenalectomy. Repeat TSS is recommended particularly in patients who had evidence of incomplete resection or a pituitary lesion on imaging although this was not the case with our patient. Repeat TSS is cited to be successful in about 50-60% of cases<sup>2</sup> but carries an increased risk of hypopituitarism and lower likelihood of remission compared to initial surgery. Remission can be achieved more rapidly compared to other second-line treatments.

## CONCLUSION

In Cushing's disease with unsuccessful initial surgery, repeat TSS may be considered when there is access to an expert pituitary surgeon.

## REFERENCES

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- 2 Patil CG, Veeravagu A, et al. Outcomes after repeat transsphenoidal surgery for recurrent Cushing's disease. *Neurosurgery.* 2008;63(2):266-270.