



Rare Presentation of Precocious Puberty Secondary to LH-Secreting Adenoma

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Patient Presentation

History:

- 8-year-old previously healthy male
- Facial hair x 1.5 years
- Phallic growth, body odor and acne x 9 months

Physical Exam:

- HEENT: **sparse facial hair above upper lip**, normal fundoscopic exam
- Neck: no thyromegaly
- GU: **phallic enlargement, testes 4cc bilaterally** and w/o palpable masses, Tanner stage I pubic hair

Initial Work-up:

- Labs notable for elevated LH and testosterone (**Table 1**)
- Evidence of pituitary mass on MRI (**Figure 1**)

Initial Labs

LH	9	0.0-0.3 mIU/mL
Testosterone	519	2-8 ng/dL
Estradiol		0-13 pg/mL
17-OHP	155	<63 ng/dL
Androstenedione	0.363	0.03-0.3 ng/mL
FSH	<0.1	0.0-2.8 mIU/mL
DHEA-S	79	5-115 ug/dL
Alpha Subunit	9	<0.55 ng/mL

Table 1: Initial laboratory studies most notable for elevated LH & testosterone

Imaging

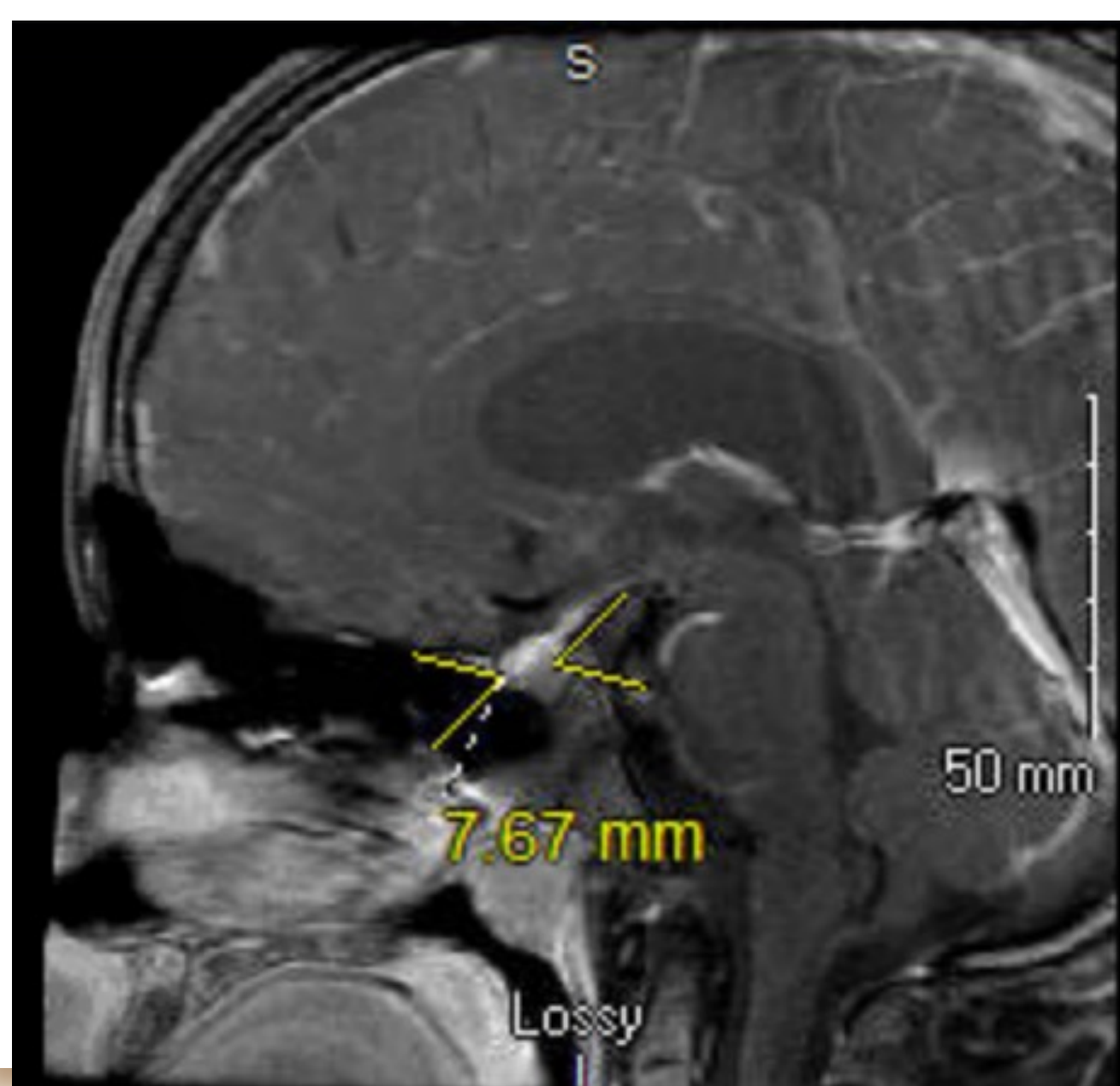


Figure 1 (Left): 8 x 12 x 10 mm hypo- enhancing mass consistent with anterior pituitary adenoma.

Central precocious puberty is more often associated with CNS lesions in males and requires further evaluation with head imaging

Departures from the typical sequence of development should raise concern for nonconcurrent secretion of gonadotropins

LH & Testosterone Levels Throughout Patient Course

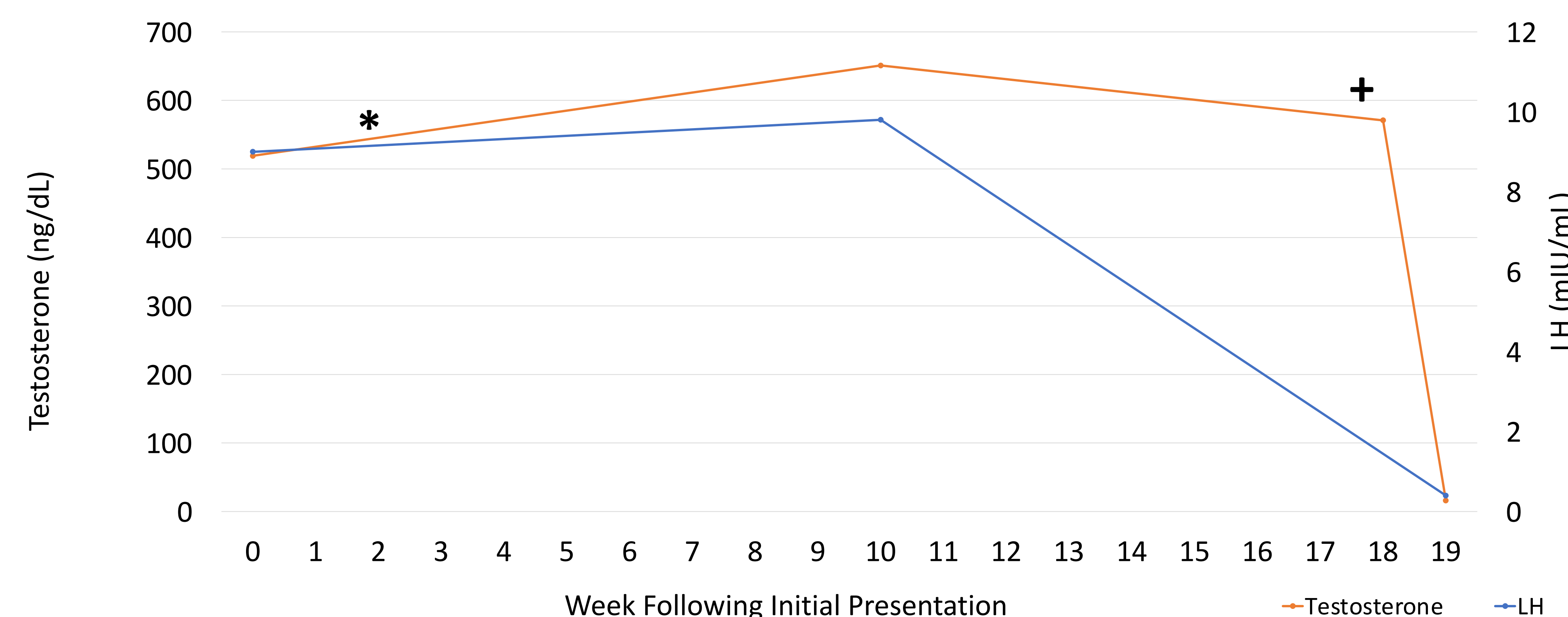


Figure 2: Testosterone and LH levels remained elevated despite treatment with leuprolide (*), although declined to near-normal levels following adenomectomy (+).

Discussion

- Preferential LH secretion led to high testosterone, phallic enlargement and other secondary sex characteristics, while absent FSH led to minimal testicular enlargement. This atypical pubertal progression raised suspicion for a pathologic cause.
- Functioning gonadotroph adenomas are rare, with most secreting FSH or co-secreting FSH and LH. Though surgical pathology was negative for LH, drastic reduction of LH and testosterone following surgery and steroidogenic factor-1 (SF1)+ pathology are consistent with an LH-secreting adenoma.
- Prior to surgery, LH and testosterone levels continued to increase despite treatment with Lupron, indicating failure of typical negative feedback signaling and necessitating surgical intervention.

Patient Course

Surgical Pathology:

- + SF1
- -LH, FSH & TSH

Diagnosis:

- LH-secreting pituitary adenoma
- Started leuprolide without symptomatic or biochemical improvement
- Started spironolactone and anastrozole to preserve height, with slowing of progression in secondary sex characteristics
- Transsphenoidal adenomectomy resulted in reduction of LH & testosterone (**Figure 2**)
- After surgery, patient noted stalling of phallic enlargement with further improvement in acne, facial hair, and growth acceleration
- Patient started central puberty at age 9