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Pseudotumor cerebri resulting in empty sella syndrome and multiple pituitary hormone deficiencies

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BACKGROUND

Pseudotumor cerebri (PTC) is frequently associated with an empty sella turcica. While commonly asymptomatic, it may present with non-specific neurologic symptoms or more rarely with pituitary dysfunction. This case highlights a patient with PTC and secondary empty sella syndrome whose symptoms of hypopituitarism were wrongly attributed to complications in the management of his PTC resulting in delayed diagnosis and management.

CASE PRESENTATION

A 17 year old male was referred to pediatric endocrinology with concerns for stalled puberty in the setting of known PTC. He was diagnosed with PTC at 10 years old and was medically managed until 13 years old when a VP shunt was placed. He required multiple shunt revisions and endured chronic headaches and back pain requiring pain specialty management with prolonged opiate and lidocaine patch use. He had decreased energy, poor stamina, and frequent nausea and vomiting that was attributed to his underlying PTC, complications from his surgical interventions, and resulting chronic narcotic use. This resulted in withdrawal from school and bedridden status for four years.

An abnormal genital exam with small phallic and testicular size led to a preliminary laboratory evaluation. This resulted in a referral to pediatric endocrinology for significantly low testosterone and an inappropriately low LH/FSH consistent with hypogonadotropic hypogonadism.

In addition to signs and symptoms of hypogonadism, he continued to complain of chronic headaches, back pain, decreased energy, and frequent nausea and vomiting. His growth velocity had slowed over the previous 3 years. On exam, he had a eunuchoid body habitus without gynecomastia. He had sparse axillary hair, Tanner II pubic hair, and a phallus smaller than expected for age. Testicular exam was concerning for testes measuring approximately 4-5 mL (normal 15-20mL) bilaterally with normal consistency.

EVALUATION

Following preliminary laboratory studies that were consistent with hypogonadotropic hypogonadism, further evaluation of the pituitary demonstrated other abnormalities (Table 1). On cosyntropin stimulation testing, the cortisol peak was 10 mcg/dL, meeting criteria for adrenal insufficiency (normal ≥18 mcg/dL).

On radiographic evaluation, his bone age was 14 years at a chronological age of 17 years and 4 months. The patient's previous brain imaging studies were reviewed and a partially empty sella was appreciated by a pediatric radiologist on retrospective evaluation (Image 1). There were no masses or calcifications appreciated on the MRI and CT completed a year prior to endocrine evaluation.

Pituitary hormones were serially evaluated. At 16 months after initial referral, TSH was inappropriately normal for a low fT4, consistent with central hypothyroidism.

Table 1. Serial laboratory evaluations

Lab	Initial evaluation	6 weeks later	16 months later	Normal range for age
Total testosterone (ng/dL)	12.26	646.9	200.2	84-350
LH (mIU/mL)	1.6	<0.1	0.4	0.4-7
FSH (mIU/mL)	3	0.4	0.6	2.6-11
DHT (ng/dL)	6.8			24-65
Prolactin (ng/mL)	12.5			3-18
TSH (mcIU/mL)	2.47		0.62	0.5-4.8
Free T4 (ng/dL)	1.33		0.9	0.93-1.6
8am Cortisol (mcg/dL)	8.9	1.9		8-19
Sodium (mmol/L)	140			133-145

TREATMENT

Testosterone was started with gradual titration to adult replacement dosing. He noticed almost immediate improvement in energy with notable progression of puberty and linear growth acceleration. Subsequently, physiologic hydrocortisone replacement therapy resulted in resolution of his nausea, a dramatic increase in energy level, and a much anticipated return to public high school from his home education program. Finally, thyroid hormone replacement was initiated 16 months later following biochemical evidence of hypothyroidism.

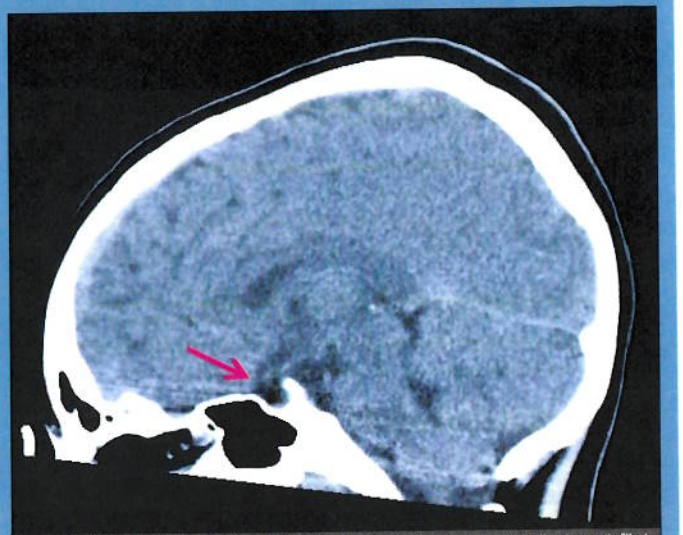

Image 1. Sagittal, midline CT image, 13 years old. The arrow shows the partially empty sella that is mostly filled with CSF and some pituitary tissue is seen flattened along the floor of the sella.

DISCUSSION

We present a case of PTC with empty sella syndrome and multiple pituitary hormone deficiencies that was undiagnosed for several years. Serial pituitary screening is necessary if there are concerns for empty sella syndrome, especially in the setting of abnormal linear growth, pubertal development, or poor energy. Early recognition and management of pituitary dysfunction is essential for normal growth and development. This patient had symptoms of pituitary deficiency for several years leading to bedridden status and significantly poor quality of life. This case stresses the importance of expanding a differential diagnosis when appropriate medical and surgical management does not result in improved symptom control. Avoidance of anchoring in a diagnostic evaluation may help to avoid unnecessary treatments and significantly increase quality of life.

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Pseudotumor cerebri resulting in empty sella syndrome and multiple pituitary hormone deficiencies

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Background: Pseudotumor cerebri is frequently associated with an empty sella turcica in neurologic literature. This radiologic abnormality is commonly asymptomatic, but it may present with non-specific neurological symptoms or more rarely with pituitary dysfunction. This case highlights a patient with pseudotumor cerebri and secondary empty sella syndrome whose symptoms of hypopituitarism were attributed to the pseudotumor cerebri, chronic pain, and medication side effects and resulted in delayed diagnosis and management.

Case presentation: A 17 year old male presented with concerns for delayed puberty and hypogonadism in the setting of known pseudotumor cerebri. The patient was diagnosed with pseudotumor cerebri seven years prior and had received medical and surgical management including a ventriculoperitoneal (VP) shunt requiring multiple revisions. The patient continued to suffer from chronic headaches and back pain despite prolonged opiate and lidocaine patch use. In addition to chronic pain, he had decreased energy, poor stamina, and frequent nausea and vomiting that was attributed to his underlying pseudotumor cerebri, surgical interventions, and chronic narcotic use. This resulted in withdrawal from school and bedridden status for four years. Upon evaluation by pediatric endocrinology, previous CT and MRI images of the brain demonstrated a partially empty sella. Testing of pituitary function revealed multiple pituitary dysfunctions including hypogonadotropic hypogonadism, secondary adrenal insufficiency, and central hypothyroidism. He had a rapid increase in energy level following treatment with testosterone, hydrocortisone, and levothyroxine. He was subsequently able to wean off his narcotic pain management and graduate from high school.

Conclusions: Pseudotumor cerebri may cause secondary empty sella syndrome and thus potential for pituitary dysfunction. Serial pituitary function screening is necessary, especially in the setting of abnormal growth and development or poor energy. This patient had symptoms of pituitary deficiency for several years, which were attributed to chronic pain medication use. Early recognition and management of pituitary dysfunction is essential for normal growth and development. Avoidance of anchoring in a diagnostic evaluation may help to avoid unnecessary treatments and significantly increase quality of life.