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Pediatric Giant Prolactinoma Presenting With Acute Obstructive Hydrocephalus and Intracranial Hypertension

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Image Article Case Report

Pediatric prolactinomas (PP) are rare but represent 50% of all pediatric pituitary adenomas [1-3]. Girls are affected more frequently than boys, although PP tend to be larger and more aggressive in boys, occurring at an earlier age, with larger mass and higher prolactin levels [1,4-6]. Thus, microadenomas (tumors <10 mm in diameter) are typical in females and macroadenomas (10-40 mm in diameter) are typical in males [1,2,4-6]. Giant prolactinomas (>40 mm in maximum diameter) are also more commonly found in boys [1]. Giant prolactinomas vary in mass characteristics and resulting hormone deficiencies [1,2]. In a large case series of PP patients, the largest tumor volume reported was 93.5 cm^3 [1]. Here we report a giant prolactinoma, with tumor volume of 105 cm^3 and hypopituitarism, in a teenage girl requiring ventriculoperitoneal shunt for decompression.

A 16-year-old female presented with 2 weeks of intractable headache, nausea and vomiting, vision impairment, and changes in balance. Historical review revealed primary amenorrhea and short stature. Initial exam demonstrated Tanner stage III breasts with no galactorrhea at rest or with stimulation and stage V pubic hair. Ophthalmologic exam revealed right eye afferent pupillary defect and decreased visual acuity (20/200), normal left eye visual acuity (20/20), concern for loss of color vision, and bilateral optic nerve edema with blurred disc margins.

Brain magnetic resonance imaging showed a large lobulated mass centered in the suprasellar cistern, measuring approximately 6.4 × 5.8 × 5.7 cm with a tumor volume of 105 cm^3. There was extension superiorly, anteriorly, and laterally, with homogeneously enhancing and cystic components, and mass effect resulting in obstructive hydrocephalus. There was also involvement of the cavernous sinuses, Meckel's caves bilaterally, and orbital apices through the right foramen ovale. No definite tumoral calcifications were seen (Fig. 1). Differential diagnoses included craniopharyngioma, germinoma, and adenoma. Initial tests demonstrated prolactin of >2000 ng/mL, with diluted result of 17 811 ng/mL.

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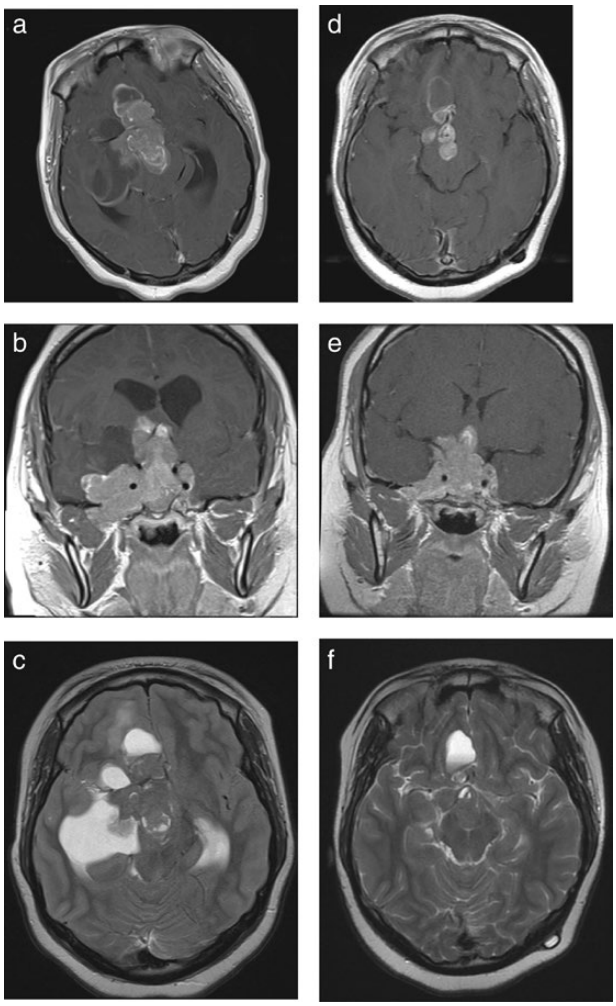

Figure 1. (A-C) Tumor prior to treatment. (D-F) Tumor at 3- month follow-up imaging.

Morning fasting labs confirmed central hypothyroidism, adrenocorticotropin deficiency, growth hormone deficiency, and hypogonadotropic hypogonadism. She was started on hydrocortisone and levothyroxine (Table 1).

Due to massive tumor size with marked obstructive hydrocephalus and concern for permanent vision loss, she underwent ventriculoperitoneal shunt placement for decompression. She was started on cabergoline and did not require surgical resection. Our patient's response to cabergoline is consistent with previous studies, with decrease in prolactin level from 17 811 ng/mL to 2350 ng/mL, 824 ng/mL, and 152 ng/mL at 1-week, 1-month, and 2-month follow-up, respectively, and a ~70% reduction in tumor volume from 105 cm^3 (6.4 × 5.8 × 5.7 cm) to 31 cm^3 (4.1 × 4 × 3.8 cm) [1,2]. She had improvement of central vision and resolution of papilledema and headaches.

PP presenting with hydrocephalus and intracranial hypertension are very rare in pediatrics, especially in girls. Our patient presented a unique case, with a rare giant prolactinoma and multiple pituitary hormone deficiencies.

Table 1. Morning fasting laboratory studies at the time of diagnosis

Laboratory study	Level at time of diagnosis	Reference range
Prolactin, ng/mL	>2000 (17 811 diluted)	3.34-26.76
Thyroid studies		
TSH, µIU/mL	1.11	0.34- 5.66
Free T4, ng/dL	0.41	0.52-1.21
Hypothalamic-pituitary-adrenal axis		
ACTH, pg/mL	62	15-66
Cortisol, µg/dL	3.1	5.0-25.0
Growth factors		
GH, ng/mL	0.4	<8.2
IGF1, ng/mL	85	20-200
IGF-BP3, µg/L	2696	2682-6470
Gonadotropins		
FSH, mIU/mL	0.034	1.5-12.8
LH, mIU/mL	<0.005	0.10-12.0

Abbreviations: ACTH, adrenocorticotropin; FSH, follicle-stimulating hormone; GH, growth hormone; IGF1, Insulin-like growth factor 1; IGF-BP3, insulin-like growth factor binding protein 3; LH, luteinizing hormone; T4, thyroxine; TSH, thyroid-stimulating hormone.

Given the high potential of aggressive behavior in giant prolactinomas, strict follow-up with repeat prolactin levels and serial magnetic resonance images are required for follow up of medical responsiveness to treatment.

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Statement of Ethics: The subject and her parent have given their written informed consent to publish their case (including publication of images).

Additional Information

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