Pediatric Giant Prolactinoma Presenting With Acute Obstructive Hydrocephalus and Intracranial Hypertension

Grace Hendrix,1 Robert Benjamin,2 Nancie Maclver,3 Daniel P. Barboriak,4 Herbert Fuchs,5 and Pinar Gumus Balikcioglu6

1Division of Pediatric Endocrinology and Diabetes, Duke University Medical Center, Durham, NC, USA; 2Division of Pediatric Endocrinology and Diabetes, Duke University Medical Center, Durham, NC, USA; 3Division of Pediatric Endocrinology and Diabetes, University of North Carolina Medical Center, Chapel Hill, NC, USA; 4Division of Neuroradiology, Duke University Medical Center, Durham, NC, USA; 5Division of Pediatric Neurosurgery, Duke University Medical Center, Durham, NC, USA; 6Division of Pediatric Endocrinology and Diabetes, and Duke Molecular Physiology Institute, Duke University Medical Center, Durham, NC, USA

ORCiD numbers: 0000-0001-8682-872X (G. Hendrix); 0000-0003-3676-9391 (N. Maclver); 0000-0002-2536-6266 (D. P. Barboriak); 0000-0002-5259-9196 (P. G. Balikcioglu).

Received: 9 August 2021; Editorial Decision: 18 October 2021; First Published Online: 28 October 2021; Corrected and Typeset: 10 November 2021.

Key Words: Pediatric prolactinoma, hydrocephalus, intracranial hypertension

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 cm³ [1]. Here we report a giant prolactinoma, with tumor volume of 105 cm³ 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 cm³. 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.
Morning fasting labs confirmed central hypothyroidism, adrenocorticotropic 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 cm³ (6.4 × 5.8 × 5.7 cm) to 31 cm³ (4.1 × 4 × 3.8 cm) [1,2]. She had improvement of central vision and resolution of papilledema and headaches.

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.

Acknowledgments

**Funding:** P.G.B. receives support from NIDDK under the award number K23DK117067, Duke Strong Start Award Program, Duke University Pediatric Departmental Support and Diabetes Research Connection. The content of this paper is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health.

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

**Additional Information**

**Correspondence:** Pinar Gumus Balikcioglu, MD, MHSc, Division of Pediatric Endocrinology and Diabetes, Duke University Medical Center, 3000 Erwin Rd, Durham, NC 27705, USA. E-mail: pinar.gumus@duke.edu.

**Disclosure:** The authors have no conflicts of interest to declare.

**Data Availability:** Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

**References**

1. Hoffmann A, Adelmann S, Lohle K, Claviez A, Muller HL. Pediatric prolactinoma: initial presentation, treatment, and long-term prognosis. *Eur J Pediatr*. 2018;177(1):125-132.
2. Breil T, Lorz C, Choukair D, et al. Clinical Features and Response to Treatment of Prolactinomas in Children and Adolescents: A Retrospective Single-Centre Analysis and Review of the Literature. *Horm Res Paediatr.* 2018;89(3):157-165.

3. Colao A, Loche S. Prolactinomas in children and adolescents. *Endocr Dev.* 2010;17:146-159.

4. Acharya SV, Gopal RA, Bandgar TR, Joshi SR, Menon PS, Shah NS. Clinical profile and long term follow up of children and adolescents with prolactinomas. *Pituitary.* 2009;12(3):186-189.

5. Fideleff HL, Boquete HR, Suárez MG, Azaretzky M. Prolactinoma in children and adolescents. *Horm Res.* 2009;72(4):197-205.

6. Guaraldi F, Storr HL, Ghizzoni L, Ghigo E, Savage MO. Paediatric pituitary adenomas: a decade of change. *Horm Res Paediatr.* 2014;81(3):145-155.