Clinical Case Reports International

Case Study
Published: 26 May, 2020

Precocious Puberty Induced by a Pilocytic Astrocytoma in a Boy

Rosa Finozzi[1], Beatriz Mendoza[1], Camila Keosseian[1], Mariana Risso[1] and Alvaro Ronco[2]*

[1]*Department of Endocrinology and Metabolism, University of the Republic, Uruguay*

[2]*Department of Oncology and Radiotherapy, Pereira Rossell Hospital, Uruguay*

Abstract

The incidence of central isosexual Precocious Puberty (PP) in males is low. Its most frequent cause is an organic one. We herewith present the case of an 8-year-old male patient with a tumor-induced PP caused by a pilocytic astrocytoma. Very few cases of this tumor type have been reported in the medical literature as a cause of PP. The Magnetic Resonance Imaging showed a mass displacing and compressing the III ventricle and the optic chiasm. The mass extended to the middle cranial fossa causing a dilation of the lateral ventricles. A male patient bearing a central PP should lead us to think of a Central Nervous System (CNS) tumor. Tumors affecting the CNS particularly those affecting the hypothalamic region- can express themselves as puberty disorders. In this case we highlight the remarkable reversal of the secondary sexual characteristics following the excision of the tumor, as well as the patient's seven year survival.

Keywords: Precocious puberty; CNS; FSH; Pilocytic astrocytoma

Introduction

Central isosexual Precocious Puberty (PP) in males is defined as the onset of secondary sexual characteristics before the age of 9 [1-6], associated with an accelerated growth rate. The incidence of this medical entity is low and the cause is frequently organic, due to the existence of a localized tumor in the hypothalamus-pituitary region [2]. A limited number of cases reported in literature in which this type of tumor causes precocious puberty, reflects its rarity and motivates this communication. The patient's mother signed giving her consent for the publication of the case, and the document is at disposal.

Case Presentation

The patient is an 8-year-old male with no pertinent perinatological or personal history, who is brought to the office because he had started six months before showing secondary sexual characteristics, including: a) Appearance of pubic hair; b) increased testicular volume; c) penis enlargement; and d) changes in tone of voice, associated with a reduced vision and headaches, with no signs suggesting organicity.

Physical examination

The patient denied any episodes of orthostatic hypotension. The confrontation visual field test revealed a bitemporal hemianopsia. The patient showed: An adolescent stance and his voice was already masculine; Tanner stage IV genitalia (pigmented, coarse pubic hair, as in Tanner stage III, 12-cm penis, clearly visible dorsal vein, pigmented scrotum, testicles in scrotum- 15 cc at right and 20 cc at left) (Figure 1); Bone age [7] advanced by 5 years; his height was forecasted as 174 cm according to the Bayley-Pinneau method, while his genetic target height would be 180 cm ± 10 cm (Table 1).

Biochemistry

Pubertal pattern was inconsistent with chronological age, with clearly elevated Luteinizing Hormone (LH) and testosterone levels performed with Fluorescence immunochemical tests (ICMA); LH was 6.3 mIU/mL, (normal range 0.5 to 1.0); Follicle Stimulating Hormone (FSH) was 2.2 mIU/mL, (normal range 0.5 to 3.5); testosterone levels were also clearly elevated: 5.51 nmol/mL (normal range 0.03 to 0.68). All other pituitary hormone levels were within normal range.

Neuroimaging

A Magnetic Resonance Imaging (MRI) scan was requested with the presumptive diagnosis of

OPEN ACCESS

Correspondence:
Rosa Finozzi, Department of Endocrinology and Metabolism, University of the Republic, Montevideo, 11600, Uruguay,
E-mail: rofinozzi@gmail.com

Received Date: 07 *May 2020*
Accepted Date: *21 May 2020*
Published Date: *26 May 2020*

Citation:
Finozzi R, Mendoza B, Keosseian C, Risso M, Ronco A. Precocious Puberty Induced by a Pilocytic Astrocytoma in a Boy. Clin Case Rep Int. 2020; 4: 1160.

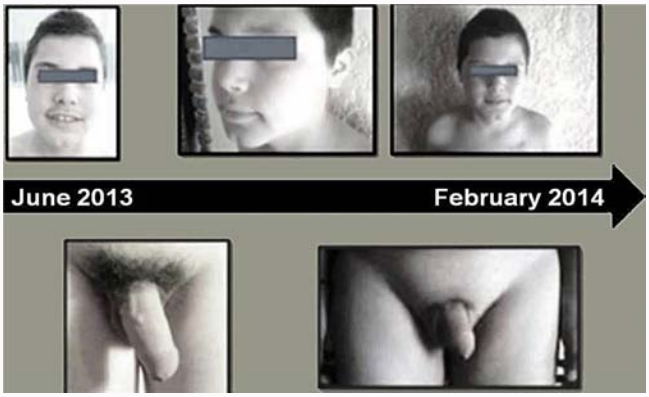

Figure 1: Preoperative presentation and postoperative outcomes.

Table 1: General physical parameters at baseline.

Weight	47 kg	Percentile >97
Height	146 cm	Z score >3.5
Arm span	143 cm	
Growth rate	14 cm/year	Percentile >97
Blood pressure	100/60 mmHg	

a Central PP (CPP) secondary to a CNS tumor (Figure 2). The MRI scan showed a 48 mm × 25 mm × 29 mm midline expansive mass displacing and compressing the III ventricle and the optic chiasm. The mass extended to the middle cranial fossa causing a dilation of the lateral ventricles. The conventional visual field revealed a severe global reduction of the left eye visual field, while the right eye showed a depression in the lower-upper temporal quadrant.

Pathology

The patient underwent a partial surgical excision of the mass, and the pathology was reported as low-grade pilocytic astrocytoma as per the international classification (ICD-O 9421/1 code of International Agency for Research on Cancer, World Health Organization). Chemotherapy was then added to complement the surgical treatment.

Postoperative outcomes

Following surgery, the patient resumed an infantile attitude; there was a reduction of the growth rate and a partial regression of the genital secondary sexual characteristics, i.e., shrinkage of the testicular volume going from Tanner stage IV to II, with regression of the pubic hair and testicular volume.

In the immediate postoperative period the patient presented a clinical picture consistent with Central Diabetes Insipidus (CDI), which was initially handled with intranasal desmopressin at variable doses; currently he is asymptomatic on 10 µg/day, delivered in a night puff. He has a hypopituitarism expressed by secondary hypothyroidism and adrenal insufficiency, both managed with replacement therapy. The former is being successfully treated with sodium levothyroxine at 1.8 µg/kg/day, achieving a clinical and biochemical normalization of the thyroid function. The adrenal insufficiency was treated with hydrocortisone according to body surface (10 mg/m^2). At present, the patient presents no symptoms of adrenal insufficiency or Cushing syndrome.

The visual field performed one month after surgery showed the persistence of a severe global reduction of the vision in the left eye, while the right eye presented a depression in the lower-upper temporal quadrants. It is worth noting that the patient has not lost the school year despite his visual impairment.

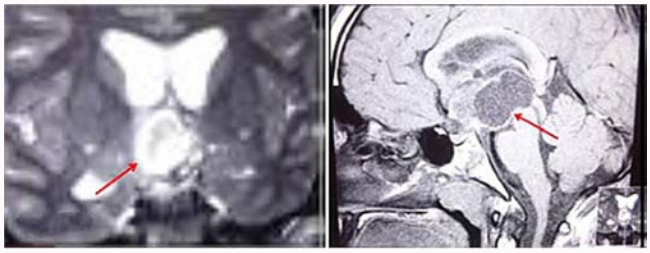

Figure 2: Magnetic Resonance Imaging of the pilocytic astrocitoma, a midline expansive mass displacing and compressing the III ventricle and the optic chiasma.

Discussion

The CPP involves the premature activation of the hypothalamic neurons in charge of producing the Gonadotrophin-Releasing Hormone (GnRH) and it is associated with glial tumors and hypothalamic hamartomas [3]. Astrocytomas typically occur in the early childhood, being their incidence peak between 2 and 6 years of age [2,5]; most of such tumors show a pilocytic histology (as was the case of this patient), and they tend to be located in the cerebellum [8]. This glioma is poorly differentiated; its natural history varies, and the extent of the surgical resection achieved is its main prognostic predictor. In his study, Burkhard et al. [5] claims that these tumors have a benign biological behavior, which-together with the advances in neurosurgery-provide excellent survival rates. Fernandez et al. [4] reports a 100% 5-year survival and 92% when the excision is only partial. Ana plastic or malignant transformation is rare [9].

It is to be highlighted that all the secondary sexual characteristics subsided remarkably after surgery in this patient, not requiring any GnRH-analog therapy. On the other hand, he presented complications that are usual of midline tumor surgery, including hypopituitarism and CDI. We believe that the absence of the seizures reported by other authors as a complication of the tumor is due to the location of the mass in the sella turcica [4,9].

Given the side effects associated with radiation therapy, postoperative chemotherapy may be recommended initially, as it may objectively reduce the tumor bulk and postpone the need for radiation therapy in most patients [10,11]. Due to the tumor site and its partial persistence, the medical team decided to complement the treatment with chemotherapy, using vincristine and carboplatin, therefore avoiding the significant complications of radiation therapy. After 7 years into the surgery there is no evidence of any imaging changes.

Conclusion

A CPP in a male patient should suggest the presence of a tumor, being the hypothalamus the most common location; hence, detecting pubertal signs is a valuable tool to guide the diagnosis.

References

1. Vargas F, Fuentes MA, Lorenzo L, Marco MV, Martínez-Aedo MJ, Ruiz R. Pubertad precoz. Protoc Diagn Ter Pediatr. Asoc Española de Pediatria (AEP). 2011;1:193-204.

2. Rivero M, Alcázar MJ, Villar MJ, Nieto N, Sanz M, Cueto M, et al. Pubertad precoz y patología tumoral. Presentación de dos casos clínicos en niñas.

Rev Esp Endocrinol Pediatr. 2012;3(1):81-6.

3. Di Palma ML, Guercio G, Costanzo M. Alteraciones de la pubertad como forma de presentación de patología orgánica del sistema nervioso central. Presentación de casos clinicos del Servicio de Endocrinología del Hospital de Pediatría Garrahan, Buenos Aires, Argentina, Mayo 2011. Endocr Ped online. 2011;33.
4. Fernandez C, Figarella-Branger D, Girard N, Bouvier-Labit C, Gouvernet J, Paz Paredes A, et al. Pilocytic astrocytomas in children: Prognostic factors-- a retrospective study of 80 cases. Neurosurgery. 2003;53(3):544-55.
5. Burkhard C, Di Patre PL, Schüler D, Schüler G, Yaşargil MG, Yonekawa Y, et al. A population-based study of the incidence and survival rates in patients with pilocytic astrocytoma. J Neurosurg. 2003;98(6):1170-4.
6. Soriano-Guillén L, Argente J. Pubertad precoz central: Aspectos epidemiológicos, etiológicos y diagnóstico-terapéuticos. An Pediatr (Barc). 2011;74(5):336e1-13.
7. Dahlberg PS, Mosdøl A, Ding Y, Bleka O, Rolseth V, Straumann GH, et al. A systematic review of the agreement between chronological age and skeletal age based on the Greulich and Pyle atlas. Eur Radiol. 2019;29(6):2936-48.
8. Khan MA, Godil SS, Tabani H, Panju SA, Enam SA. Clinical review of pediatric pilocytic astrocytomas treated at a tertiary care hospital in Pakistan. Surg Neurol Int. 2012;3:90.
9. Otero-Rodriguez A, Sarabia-Herrero R, García-Tejeiro M, Zamora-Martinez T. Spontaneus malignant transformation of a supratentorial pilocitic astrocytoma. Neurocirugia (Astur). 2010;21(3):245-52.
10. Gnekow AK, Falkenstein F, von Hornstein S, Zwiener I, Berkefeld S, Bison B, et al. Long-term follow-up of the multicenter, multidisciplinary treatment study HIT-LGG- 1996 for low-grade glioma in children and adolescents of the German Speaking Society of Pediatric Oncology and Hematology. Neuro Oncol. 2012;14(10):1265-84.
11. Packer RJ, Ater J, Allen J, Phillips P, Geyer R, Nicholson HS, et al. Carboplatin and vincristine chemotherapy for children with newly diagnosed progressive low-grade gliomas. J Neurosurg. 1997;86(5):747-54.