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Aggressive Super-Giant Prolactinoma in a Male Adolescent: A Case Report

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Abstract **Case Report**

Giant prolactinoma (GP) in childhood and adolescence is a rare entity with scarce litterature. We report a case of A 17-year-old adolescent Moroccan boy presented with a Rapidly progressive vision loss complicated with an incomplete Intracranial hypertension syndrome. Magnetic resonance imaging (MRI) showed a very large intrasellar mass with suprasellar extension and evidence of recent necrotic and hemorrhagic remodeling. Patient underwent an urgent transsphenoidal surgery for tumor resection. Post operative labroratory findings were in favor of a hyperprolactinemia a panhypopituitarism and transient Diabetes insipidus. Histological assessment confirmed diagnosis of prolactinoma. Pituitary MRI control revealed an intrasellar residue. Patient was then started on cabergoline.This case highlights the importance of early identification and the need for a multidisciplinary approach in managing Giant aggressive prolactinomas in young patients.

Keywords: Giant Prolactinoma, Aggressive Prolactinoma, Adolescent, Case Report.

INTRODUCTION

Pituitary adenomas in pediatric patients are uncommon, representing less than 3% of all intracranial tumors in children, with prolactinomas accounting for the majority.

Due to this, they are frequently misdiagnosed as other suprasellar masses, such as craniopharyngiomas. Prolactinoma is a rare benign tumor that results from the monoclonal proliferation of lactotrophic cells in the adenohypophysis, likely caused by somatic mutations. In contrast to adults, macroprolactinomas in children (defined as masses ≥1 cm in diameter) are more prevalent, occurring in approximately 60–80% of cases. Giant prolactinomas are a subset of macroprolactinomas, characterized by a diameter of ≥4 cm or a suprasellar extension of ≥2 cm.

CASE REPORT

A 17-year-old boy who was previously healthy presented first to an ophthalmologist with an acute frontal headache and visual loss, without vomiting following a one month of rapidly progressive history of blurred vision, no galactorrhea or gynectomastia. He was not taking any medication. His family history was unremarkable for endocrinopathies.

An urgent brain MRI was performed, revealing an expansive sellar and suprasellar mass measuring 125 × 97 × 60 mm exerting compressive effects and displacing the cavernous sinuses and the optic chiasm, consistent with a pituitary macroadenoma exhibiting necrotic and hemorrhagic remodeling **(Figure 1).** Goldmann Visual field testing revealed total left eye vision loss and a profound deficit in the entire visual right field with an inferonasal crescent of vision.

Taking into account the surgical urgency and the patient's limited financial resources, a minimal workup was requested revealing a corticotropic deficiency with a morning cortisol of 2.1 ug/dl, associated with thyrotropic deficiency : TSH= 1.87Ui/L, T41= 6.32 pmol/L.

Blood electrolyte panel, Liver and renal function were normal. Preoperative prolactin levels were not assessed.

The Patient underwent an urgent transsphenoidal surgery for tumor resection under intravenous hydrocortisone coverage, then subtituted three days later with levothyroxine.

The postoperative course was marked by transient diabetes insipidus, managed for one week with oral sublingual Desmopressin.

Immunostaining revealed neoplastic cells with diffuse strong positively for prolactin and negative for ACTH, FSH, LH, TSH and GH, with a Ki 67 index at 2%.

Three months after surgery, he was adressed to our Endocrinology department. Blood pressure 119/70 mmHg, heart rate 84/min, anthropometric measurements showed a height of 155 cm (-2.74 Standard deviation) weight of 85.4kg (-0.36 Standard deviation), and body mass index of 26.2 kg/m2 (+1.42 Standard deviation). Physical examination showed a Tanner stage 5 axillary hair and pubic hair, testicular volume of 20 ml bilaterally.

Prolactin level was at 43.75 ng/ml. Pituitary MRI control 3 months after surgery revealed an intrasellar residue lateralized to the left, measuring 10 × 8 mm, communicating with the sphenoidal sinus. Integrity of the optic chiasm and the opto-chiasmatic cistern preserved **(Figure 2).**

Hydrocortisone and Levothyroxine replacement were continued and the patient was started on Cabergoline (dopamine agonist), 0.5 mg twice a week and increased gradually in 4 weeks to reach 1 mg twice a week. A possible dose increase is planned based on further evaluation, Aiming to lower prolactin levels, induce tumor shrinkage, improve visual function, and achieve reversal of panhypopituitarism.

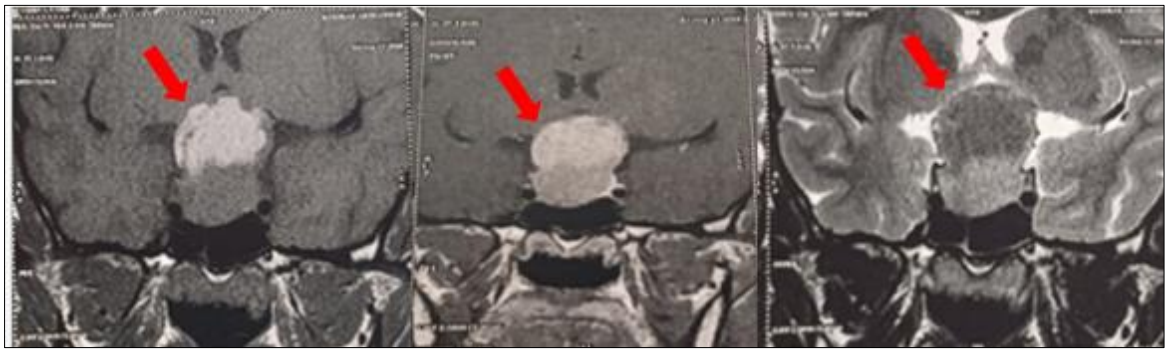

Figure 1 : Preoperative coronal T1- and T2 weighted magnetic resonance imaging pituitary revealing an expansive sellar and suprasellar mass measuring 125 × 97 × 60 mm exerting compressive effects and displacing the cavernous sinuses and the optic chiasm, consistent with a pituitary macroadenoma exhibiting necrotic and hemorrhagic remodeling

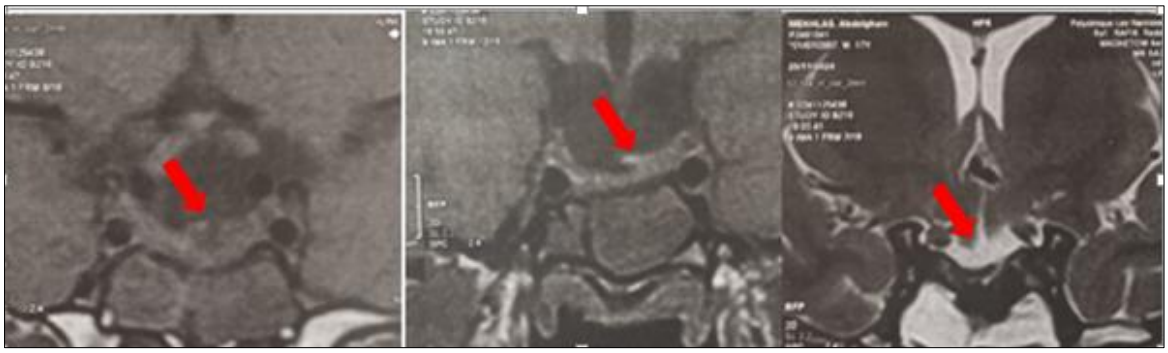

Figure 2 : 3 months Postoperative T1- and T2 weighted magnetic resonance imaging pituitary revealing an intrasellar residue lateralized to the left, measuring 10 × 8 mm, communicating with the sphenoidal sinus. Integrity of the optic chiasm and the opto-chiasmatic cistern preserved

DISCUSSION

Giant prolactinoma is exceptionally rare in childhood with an estimated frequency of 0.5–4.4% for all pituitary tumors. They present with symptoms of hyperprolactinemia and tumor mass effect, such as neuro - ophthalmologic symptoms. Rarely pituitary apoplexy may occur when acute bleeding inside the tumor occurs causing a sudden headache, visual loss, double vision and pituitary failure [1, 2].

Consensus guideline for the diagnosis and management of pituitary adenomas in childhood and adolescence recommends [3].

Offering serum prolactin measurement in case of one or more of the following signs and symptoms: delayed puberty; galactorrhoea; visual field loss; growth or pubertal arrest; or girls with menstrual disturbance.

That The diagnosis of hyperprolactinaemia requires age-specific and sex-specific prolactin reference ranges and the exclusion of confounding conditions such as hypothyroidism, renal and/or hepatic impairment, and use of medications that cause hyperprolactinaemia.

Ruling out mixed prolactin and GH hypersecretion. Age-dependent and sex-dependent insulin-like growth factor 1 (IGF1) evaluation should always accompany prolactin assessment in children and young people with prolactinomas.

Offering a dopamine agonist as first-line therapy to reduce serum prolactin concentrations and induce tumour shrinkage.

In studies of children and young people with prolactinomas, dopamine agonists lower prolactin concentrations in 60–70% of patients, reduce tumour size by 80–88%, improve visual deficits, resolve pubertal delay , and eliminate headache [4, 5].

Cabergoline was recommanded as the dopamine agonist of choice given its superior effectiveness as it has a longer half-life and greater affinity for the dopamine receptor with lower adverse effect profile. It has been recommanded even in the presence of visual disturbance and pituitary apoplexy, while carefully monitoring for any deterioration in vision, pituitary function or general status.

In an observational study of 28 paediatric patients, prolactinomas smaller than 13.5 mm in diameter (13 patients) achieved normalization of prolactin levels without surgery, using conventional cabergoline doses (up to 2 mg/week) [6].

At the start of treatment with a dopamine agonist It's necessary to offer an echocardiogram; then yearly for patients receiving >2 mg per week cabergoline and every 5 years if on ≤2 mg per week.

Moreover, another series of 22 Children and young people with prolactinomas reported that all tumours of >20 mm diameter required surgery. Although successful dopamine agonist discontinuation has been achieved in, younger patients and those with high serum prolactin concentrations at diagnosis (a marker of adenoma size) are less likely to achieve complete remission and euprolactinaemia [4-7].

If the serum level of prolactin has been normalized for at least 2 years on medical therapy and there is no visible residual prolactinoma on MRI, consider gradual cabergoline dose reduction to maintain normoprolactinaemia and eventual treatment discontinuation, with continued serum prolactin monitoring for at least 2 more years.

In cases of Resistance to standard doses of cabergoline, It has being recommanded to offer graduated dose increments of up to 3.5 mg per week. High-dose cabergoline is reportedly well tolerated and doses of up to 7 mg per week have been used to successfully treat children and young people with prolactinoma [8].

A multidisciplinary discussion is crucial in the following cases :

- Offering surgery when the patient is unable to tolerate or is resistant to high-dose cabergoline, Or when he develops deteriorating vision on cabergoline
- Offering radiotherapy can be discussed in some cases when surgery is not an option

CONCLUSION

Aggressive giant prolactinoma represents a particularly challenging clinical scenario, necessitating a multidisciplinary approach to ensure comprehensive patient care and optimal outcomes. Despite significant efforts to codify and standardize management protocols, these guidelines may not fully address the unique complexities of each case. Therefore, a personalized, holistic approach that integrates the expertise of endocrinologists, neurosurgeons, radiologists, and other specialists remains essential.

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