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Pituitary Apoplexy Revealing A Pediatric Prolactinoma: A Case Report

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Abstract
Pituitary apoplexy (PA) is an extremely uncommon condition in children and adolescents. It's an acute state involving hemorrhage or infarction of the pituitary gland.
This article presents a case report of an 11-year-old pubertal female who presented intense cranial headaches with a sudden deterioration in her visual acuity especially in her right eye. Laboratory testing revealed very high prolactin levels exceeding 2000 ng/ml. Pituitary MRI (Magnetic Resonance Imaging) showed an intra-sellar pituitary lesion described as hyperintense on T1 and T2 and characteristics of hemorrhage consistent with a pituitary macroadenoma in apoplexy. The patient underwent endoscopic endonasal transsphenoidal surgery of the tumor. Postoperative evolution was marked by amelioration of right eye vision and a decreased level of prolactin. The biological profile was marked by persistence of panhypopituitarism, hormonal replacement therapy was prescribed.
As far as we are aware, since 1980, only 30 cases of clinical pituitary apoplexy have been reported in patients under 20 years of age. The present case is the 31st and involves the youngest patient reported to date.
This case highlights the importance of early recognition, prompt surgical intervention, and the critical role of multidisciplinary care in managing pediatric pituitary disorders.

Keywords: Pituitary Apoplexy, Prolactinoma, Magnetic Resonance Imaging, Pubertal Girl.

1. Introduction

Pituitary adenomas are uncommon during childhood and adolescence, accounting for an estimated 2–6% of all pituitary adenomas that require surgical treatment [1]. Fewer than 10% of all pituitary tumors are diagnosed in children and adolescents. The majority of these cases about 80 to 90% are

craniopharyngiomas, while adenomas represent a smaller fraction, accounting for approximately 3% of all intracranial tumors. Macroprolactinomas, which are large prolactin-secreting tumors account for a small proportion of pituitary tumors in children are known to cause significant morbidity. Among adenomas occurring in individuals under 20 years of age, only 16% are associated with pituitary apoplexy (PA) [2].

In children, pituitary adenomas differ in their characteristics compared to adults. In adult patients, about a third of these tumors are functional and typically small, measuring less than 10 mm. However, in pediatric cases, the vast majority 95 to 97% are functional, and are generally much larger suprasellar extension at the time of diagnosis [3].

It is an infrequent yet potentially life-threatening condition, marked by the abrupt onset of headache, nausea, vomiting, vision impairment, cranial nerve issues, and, in some cases, loss of consciousness, often resulting from the loss of hormonal support from the pituitary gland [4].

We report the case of an 11-year-old girl who presented with sudden onset of headache accompanied by a decline in vision. The diagnosis of macroprolactinoma was confirmed after MRI imaging and very high prolactin levels, her clinical course from diagnosis to postoperative management was detailed. This article examines the incidence and characteristics of clinical pituitary apoplexy in pediatric and adolescent patients, aiming to improve awareness of this rare condition and inform treatment approaches.

2. Case Report

An 11-year-old prepubescent girl presented to the emergency department with acute onset of severe headache, nausea, and vomiting. Her symptoms were accompanied by acute deterioration in her visual acuity especially in her right eye. The patient had no significant past medical history or familial endocrine disorders. During the clinical assessment, her vital parameters were within normal range, and she had a full Glasgow Coma Scale (GCS) score of 15/15. Her pubertal development, including stature and BMI, was within normal limits. She had no evidence of spontaneous galactorrhea, acrofacial dysmorphic syndrome or Cushing's syndrome signs.

Contrast-enhanced CT showed a pituitary mass exhibiting peripheral enhancement along with evidence of hemorrhage, whereas MRI images showed an intra-sellar pituitary lesion measuring 20 x 23 x 33 mm in diameter (AP x T x H) , appearing as a T1 hyperintensity, T2 hyperintensity, and FLAIR hyperintensity, with a fluid-fluid level, containing a tissue portion with mild post-contrast enhancement in the inferolateral region, consistent with a hemorrhagic pituitary macroadenoma in apoplexy with compression of optic chiasm and no cavernous sinus invasion (Figure 1). Visual field analysis revealed a partial deficit in the upper right temporal field and a complete deficit in the left temporal side.

Her hormonal evaluation showed a prolactin level greater than 2000 ng/mL and a lowered concentration of free T4 (9,72 pmol/L, normal range 12,6–21 pmol/L) and TSH (3,17 mUI/L normal range 0.46–4,6 mIU/L) levels. The LH level was measured at 0.10 mIU/mL (normal 1.1–12,1 mIU/mL) and FSH (0,18, normal 6,3-24 mIU/mL) were decreased, In contrast, the levels of her other hormones remained within normal limits IGF1: 139,7 ng/mL (normal 76 – 349), and morning 8 am cortisol was 18,6 ug/dl (normal 7 – 25 ug/dl) with normal ACTH levels. These results pointed toward a diagnosis of prolactinoma with hypopituitarism, primarily impacting thyroid and gonadotropic hormone function.

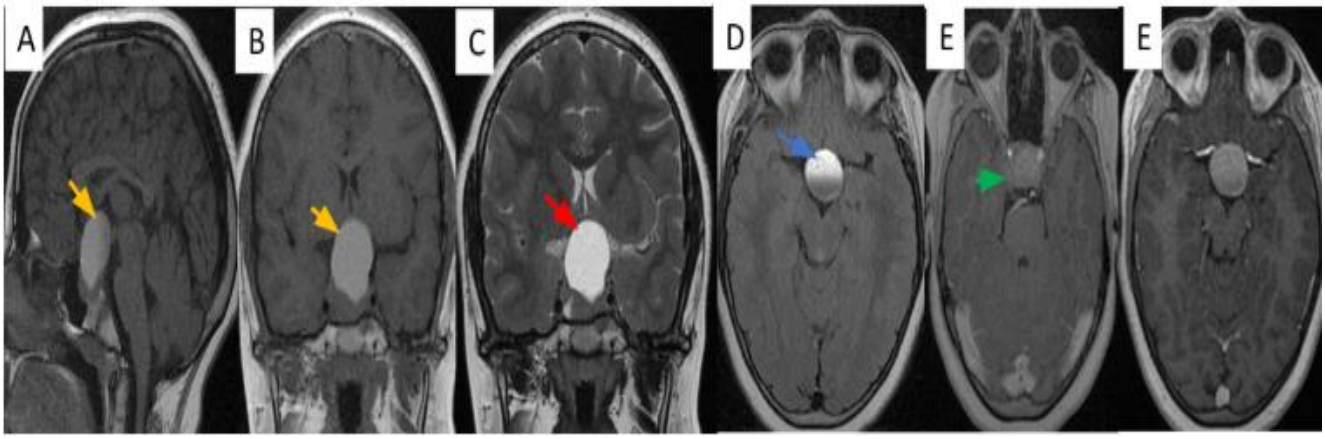

Figure 1: Pituitary magnetic resonance imaging: in T1-weighted sagittal (A) and coronal (B) sequences, coronal T2 (C), FLAIR (D), and axial T1 after gadolinium injection (E): showing an intra-sellar pituitary lesion, appearing as a T1 hyperintensity (yellow arrow), T2 hyperintensity (red arrow), and FLAIR hyperintensity, with a fluid-fluid level (blue arrow), containing a tissue portion with mild post-contrast enhancement in the inferolateral region (green arrow), consistent with a hemorrhagic pituitary macroadenoma (pituitary apoplexy).

Based on clinical, biochemical, and radiological findings, the patient was diagnosed with pituitary apoplexy secondary to a macroprolactinoma. Given the acute presentation and evidence of optic chiasm compression, urgent surgical intervention was planned.

Decompression of the pituitary macroadenoma was achieved through an endoscopic endonasal transsphenoidal surgical approach. The procedure involved a minimally invasive endonasal approach, allowing direct access to the sellar region. Intraoperative findings included a hemorrhagic pituitary tumor with necrotic macroscopic lesions, which was carefully debulked to relieve pressure on the optic chiasm. No complications were encountered during the surgery. After the surgery, her recovery was noted by a gradual enhancement in the vision of her right eye. Histopathological examination confirmed the diagnosis of a prolactinoma. (Figure 2 and 3).

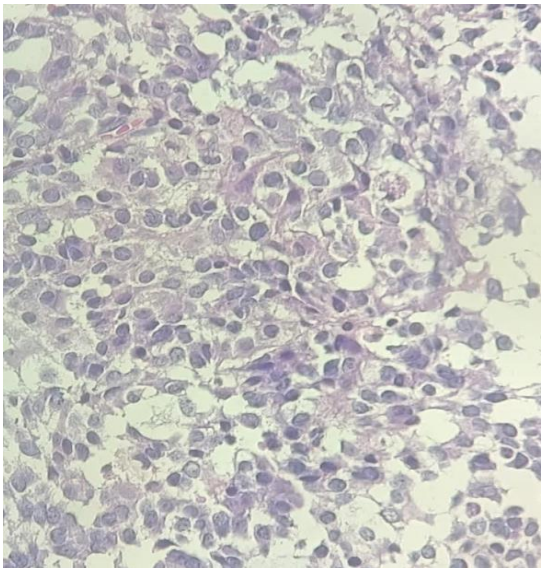

Figure 2: Histological images supporting a diagnosis of pituitary adenoma.

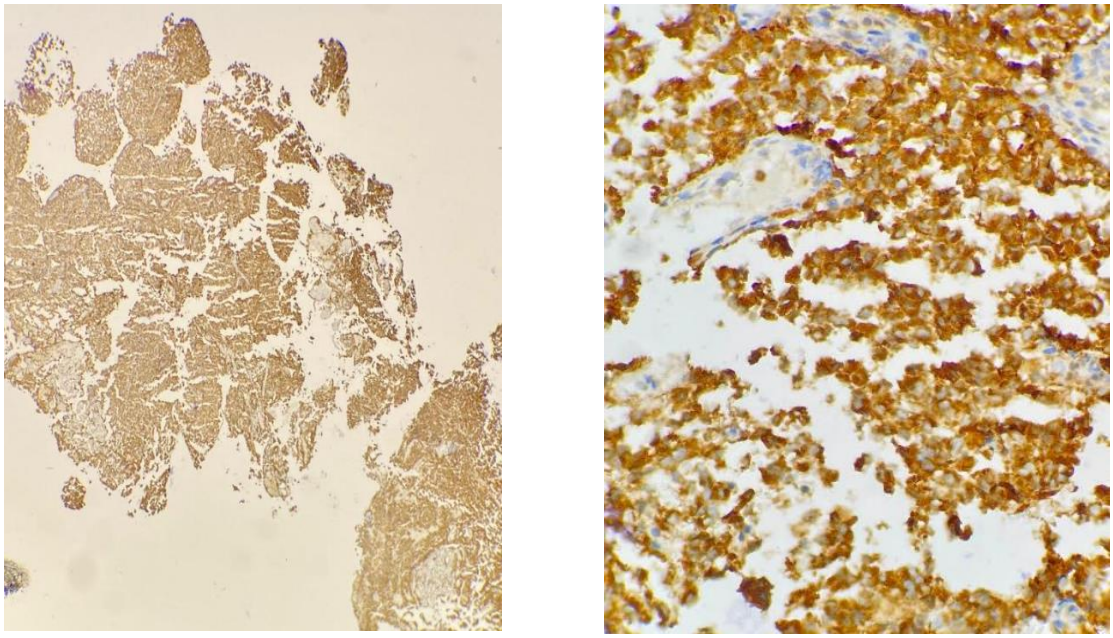

Figure 3: Immunohistochemical images supporting a diagnosis of prolactin-secreting pituitary adenoma with positive staining for anti-prolactin antibodies.

The patient's immediate postoperative recovery was uneventful. Visual acuity improved significantly within 48 hours, and follow-up visual field testing showed resolution of bitemporal defect. Hormonal postoperative evaluation after 1 week of surgery revealed decreased prolactin level 35,8 ng/mL without dopamine receptor agonist treatment. Persistent deficiencies in multiple pituitary axes, consistent with panhypopituitarism were present. The patient was treated on a regimen of hormone replacement therapy, including hydrocortisone, levothyroxine and growth hormone. After two weeks, her hormone profile was retested, showing a prolactin level of 37.6 ng/mL. She was prescribed cabergoline 0.5 mg once a week and will have routine follow-up appointments for endocrine monitoring, visual evaluations, and MRI scans. Estrogen replacement therapy, will be introduced at a later stage to induce puberty. A pituitary MRI is scheduled three months after surgery.

3. Discussion

Pituitary apoplexy (PA) is a medical emergency resulting from sudden hemorrhage or infarction in the pituitary gland, most often occurring within a pre-existing pituitary adenoma. Pituitary adenomas in children and adolescents account for an estimated 2–6% of all surgically treated cases. Due to their rarity, there is limited consensus regarding their clinical characteristics [5].

Non-functioning pituitary adenomas account for only 3% of all pituitary adenomas in patients under 20, likely due to the slow growth of these tumors [6] [7]. To date, only four detailed cases of pituitary apoplexy (PA) originating from non-functioning pituitary adenomas in this age group have been reported [8] [9] [10] [11].

Headache (70%) and visual disturbances (27%) are the most common symptoms, though the headache can vary in presentation. This condition is typically triggered by meningeal irritation due to blood entering the basal cisterns or by the stretching of the dura. Additionally, the presence of blood in the suprasellar cistern can lead to chemical meningitis, as noted by Wen-Yi et al. [12]. It is commonly linked with nausea and vomiting, and may be mistaken for other neurological emergencies such as subarachnoid hemorrhage,

cavernous sinus thrombosis, or migraine [13] [14]. In our young patient, headaches were the first symptom revealing the PA.

The most frequent and life-threatening hormonal complication of pituitary apoplexy is corticotropic deficiency, which causes severe hypotension and hyponatremia due to insufficient glucocorticoid levels. Treatment typically involves steroid replacement therapy followed by urgent surgery. There are limited case studies focusing on the treatment and long-term recovery both hormonal and neurological of pituitary apoplexy in children. Consequently, the differences in treatment outcomes between pediatric and adult patients are not well established [2]. In our patient, thyrotrope and gonadotropic axes were the first pituitary axes that showed a hormonal deficiency, whereas corticotropic axe remained intact.

Restricted visual field, often resulting in bitemporal hemianopia, is caused by the stretching of the optic nerves and chiasm. Lateral hemorrhage and necrosis can lead to deficits in the third, fourth, fifth (first and second branches), and sixth cranial nerves. A lowered level of consciousness may be due to endocrine imbalances from hypopituitarism, pressure on the brainstem, or compression of the hypothalamus [15]. Our patient had a restricted visual field in both sides probably due to compression of optic chiasm.

A pituitary mass with peripheral enhancement is visible on contrast-enhanced CT, with or without evidence of hemorrhage. Non-enhanced CT may show patchy or confluent hyperdensity. In some cases, a subarachnoid hemorrhage is also present. Early-stage T1 MRI imaging reveals an enlarged pituitary gland that appears iso- or hypointense compared to the brain, while in later acute or sub-acute stages, the gland appears hyperintense. Hyperintensity may be observed, as acute compression of the hypothalamus and optic chiasm can cause hyperintensity along the optic tracts on T2 MRI. Restricted diffusion within the adenoma may also suggest early apoplexy [15]. The MRI imaging of our patient showed a T1, T2 and FLAIR hyperintensity with a fluid-fluid level consistent with a hemorrhagic pituitary macroadenoma in apoplexy.

Histological findings in patients under 20 years of age, as reported in the largest series of pituitary adenomas in children and adolescents by Mindermann et al., revealed that 52.9% had prolactinomas, 30.9% had ACTH-secreting adenomas (Cushing's disease), 8.8% had GH-secreting adenomas, and 2.9% had non-functioning adenomas.[6]. Before, during, and after puberty, the types of adenomas varied. ACTH-releasing adenomas were most commonly diagnosed in prepubescent children (ages 0–11), while prolactinomas were more frequently observed in older children and adolescents. Similar findings have been reported in other large studies on pediatric pituitary adenomas, where prolactin and ACTH-secreting tumors are the most common, and non-functioning adenomas are comparatively rare. [16] [17] [18]. Histopathological findings in our patient revealed an adenomatous tissue with no evidence of malignant lesions, however immunohistochemical complement found a positivity of anti-prolactin antibodies confirming the diagnosis of prolactinoma.

Singh et al. [19] reviewed the outcomes of various treatment approaches for pituitary apoplexy (PA) and concluded that most patients had excellent results, with no statistically significant differences between those treated surgically and those treated conservatively. All patients with endocrine deficiencies were managed acutely with hormone replacement therapy. The endoscopic endonasal approach is emerging as a key procedure for treating large pituitary adenomas with apoplexy in children and adolescents, as shown in the current case, with advancements in technology making it a less invasive option. Our patient underwent a transsphenoidal debulking surgery that went without complication and hormonal replacement therapy was prescribed with regular clinical, hormonal, ophthalmological and radiological checkups.

4. Conclusion

This case illustrates the complexities of diagnosing and managing pituitary apoplexy in children. Early recognition of symptoms, prompt imaging, and multidisciplinary care are essential to achieving optimal outcomes. The successful surgical treatment of this 11-year-old girl underscores the efficacy of the transsphenoidal approach, while her postoperative course highlights the need for vigilant endocrinological follow-up.

As awareness of pediatric pituitary disorders grows, clinicians must remain attuned to the unique challenges posed by these rare but impactful conditions. Continued research and collaboration will be key to improving diagnostic accuracy and therapeutic strategies for young patients with pituitary apoplexy and related disorders.

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