Clinical Medicine Insights: Case Reports

CASE REPORT

Obstructive Hydrocephalus, Fifth Nerve and Hypothalamus Involvement: Acute Presentation of a Giant Prolactinoma

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Abstract

Introduction: Pituitary tumors from lactotrope cells account for about 40% of all functioning pituitary cancers. Men tend to present with a larger, more invasive and rapid growth prolactinomas than women, possibly because hypogonadism features are less evident.

Case report: A 27-year-old, previously asymptomatic Saudi man presented with a 3-day history of emesis with severe left-sided frontal headache, left face and right upper limb numbness, with signs of obstructive hydrocephalus. Brain Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) revealed a giant pituitary mass occupying several regions (sellar, infra-sellar, and supra-sellar) measuring 6.5 × 5.7 × 5.9 cm, and invading the sphenoid sinus as well as the cavernous sinuses bilaterally, with intra-pituitary hemorrhage compressing the third ventricle causing obstructive hydrocephalus. Prolactin levels were >200,000 mIU/L, consistent with invasive giant prolactinoma (IGP). He was treated with Cabergoline which eventually normalized the prolactin level and significantly reduced the size of IGP.

Conclusion: This is a rare case of obstructive hydrocephalus with super-imposed intra-pituitary hemorrhage secondary to IGP, highlighting the importance of a full hormonal assessment for proper diagnosis and management.

Keywords: (IGP) invasive giant prolactinoma, obstructive hydrocephalus

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Introduction

Pituitary tumors from lactotrope cells account for about 40% of all functioning pituitary cancers.[1] Men tend to present with a larger, more invasive and rapid growth prolactinomas than women, possibly because hypogonadism features are less evident.[2] Non-specific manifestations include menstrual irregularities for women, sexual dysfunction, galactorrhea,[3] and osteopenia.[4] Other signs and symptoms are due to compression and obstruction effects, most common of which is the loss of vision and bitemporal hemianopia.[5] Rarely, patients with prolactinoma will present with obstructive hydrocephalus or seizures.[6] Current recommendations suggest Dopamine agonists as the treatment of choice for prolactinomas, including giant and invasive types as these drug suppresses PRL secretion and synthesis as well as lactotrope cell proliferation.[7]

Case Presentation

A previously asymptomatic, 27-year-old Saudi man, presented with a 3-day severe unilateral headache involving the left eye, not relieved with medications, accompanied with emesis and prick-like sensation at the left part of his face and right upper limb. Patient had a history of blurred vision, no fever. On examination, patient was fully conscious and oriented. Vital signs revealed hypotension (94/68 mmHg) and bradycardia (50 bpm, regular). Temperature was 37 °C. The rest of the routine physical and laboratory exams were unremarkable. ECG showed sinus bradycardia. CT scan of brain without contrast revealed hyper-dense sellar, supra-sellar mass extending to the left pontine area obstructing third ventricle with mild subarachnoid hemorrhage. Few days later, patient became drowsy, with no significant changes in physical signs. Patient was kept head up 30°. Dexamethazone and Tegretol were given, and emergent ventricle-portal shunt has been placed. Patient thereafter experienced improvement in headache and vomiting. CSF revealed WBC: 2 c/cumm; RBC: 2.196; glucose: 67 mg/dL and protein 17 mg/dL. CSF culture was negative. He was subsequently referred to our department.

Upon referral, patient denied history of syncope, fever, night sweats, and skin dryness. He noted a decrease in his libido, impotence, decreased facial and chest hair one year before his presentation but did not seek medical advice. On examination, he was

Table 1. Hormonal assessment of the patient from baseline to 18 months.

Hormones	Admission	2 weeks	1 month	2 months	5 months	6 months	10 months	12 months	18 months	24 months
Prolactin	273.86	12420	2918	2017	961.3	579.4	383	589	148	137.5
TSH	0.16	–	5.89	6.04	1.2	–	5.26	7.8	3.2	–
FT4	–	–	11.41	11.34	17.84	–	11.86	10.95	15.1	11.6
Testosterone	0.07	–	2.29	3.66	6.72	1.81	2.94	12.06	3.44	2.76
SHBG	16.3	–	16.55	–	–	16.04	13.61	14.77	–	–
Free testosterone index	<0.423	–	13.8	–	–	–	–	81.7	–	8.31
LH	4.33	–	2.14	3.72	–	–	<0.1	–	–	–
FSH	3.53	–	2.18	3.78	–	–	0.21	–	–	–

conscious, coherent and ambulatory. He was obese with a BMI 35.6 kg/m², blood pressure 115/70 mmHg, heart rate 64 bpm regular, respiratory rate 22 bpm, oxygen saturation 96% in room air and afebrile. Secondary sexual characteristic features revealed decreased facial and trunk hair, neck and cranial nerve examinations were unremarkable. Further investigations and endocrine work up is shown in Table 1. Prolactin level ≥ 100,000 mIU/L (normal 86–390 mIU/L); repeat measurement revealed ≥ 200,000 mIU/L. Other hormones IGF-I, GH, ACTH were normal.

Patient was referred to an ophthalmologist and assessment revealed no evidence of visual field abnormalities; optic disc and macula were normal. The patient was started on Cabergoline 0.25 mg po twice per week with gradual increase in the dose to 1 mg twice a week. Table 1 shows normalization of the prolactin level. MRI 1 year post-treatment showed significant regression in the overall size and extension of the pituitary adenoma with subsequent interval resolution of the mass effect on the suprasellar optic pathway and in the dilatation of third ventricle compared with the previous MRI as shown Figure 1.

Discussion

In this case report we present an adult Saudi patient with IGP, who had significant history and signs of hypogonadism for one year without seeking medical consultation, and finally presenting with mass effect symptoms of IGP.

Remarkable progress has been made with regards to the management of giant prolactinoma, the ultimate goal of which is to achieve eugonadism, euprolactinemia status, and reduction of tumor size.[8] Other modalities include radiotherapy, which is not the usual preferred treatment because of high complication rates.[9] On the other hand, therapy with dopamine agonist drugs (Bromocriptine, Cabergoline), are consistently promising in hypogonadal status reversal, prolactin level correction and regression size of pituitary adenomas.[10–15]

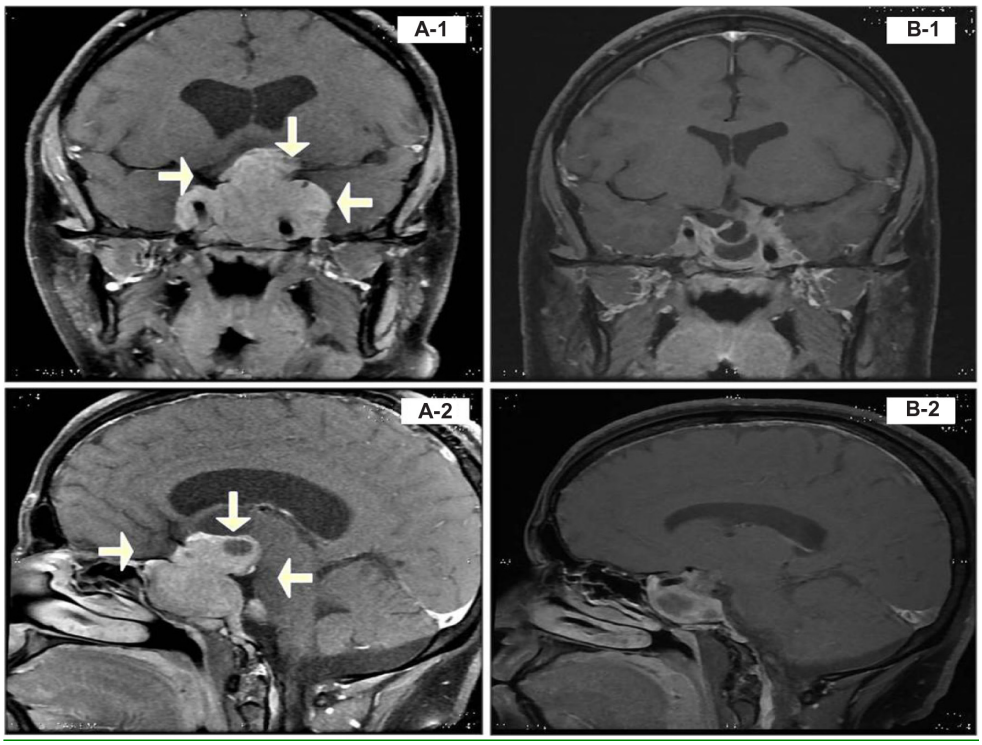

Figure 1. Sagittal and frontal T1 MRI before (**A-1** and **A-2**), and after treatment with Cabergoline (**B-1** and **B-2**).
Notes: MRI of the brain and pituitary gland showed enormous invasive tumor mass lesion is mainly solid with two cystic components at sellar, infra-sellar, and supra-sellar area measures 6.5 × 5.7 × 5.9 cm in its anterior-posterior, transverse, craniocaudal dimensions respectively. The mass displaced the optic chiasm superiorly. Anterior portion shows retrochiasmatic component which is lobulated and shows a small to medium size lobule extending posteriorly to the left CP angle (compressing the the left trigeminal nerve), with another small lobule extending posteriorly to the left orbit through the superior orbit fissure. The two cystic components are noted at the hypothalmic and third ventricle with fluid level (evidence of intra-lesional hemorrhage of the posterior extension of this mass along the left tentorial leaflet and adjacent left lateral border of the brainstem), likely representing pituitary apoplexy.

Conclusion

This case highlights a rare presentation of IGP in the form of obstructive hydrocephalus, and the importance of accurately diagnosing prolactinomas. As was revealed in this case, our patient improved significantly medical therapy, avoiding unnecessary invasive treatments, and preventing short and long term complications.

Consent

Written informed consent was obtained from the patient for publication purposes and accompanying images. A copy is available upon request.

Author's Contributions

SA and NJ diagnosed, investigated and managed the patient. SA and NJ contributed equally in the writing of this case report. Both authors read and approved the final version of the manuscript.

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Competing Interests

The authors have no conflict of interest.

Disclosures and Ethics

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