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Case report

Prolactinoma with apoplexy in the context of thrombocytopaenia: A case report

Oliver Chow [a], Therese Pham [b], Brandon He [c,*], Amy Hort [d], Damien Gibson [e], Gemma Olsson [f]

[a] Westmead Hospital Plastics and Reconstructive Department, Australia
[b] Liverpool Hospital Plastics and Reconstructive Department, Australia
[c] University of New South Wales, Australia
[d] Westmead Hospital General Surgical Department, Australia
[e] Westmead Hospital, Australia
[f] Westmead Hospital Neurosurgery Department, Australia

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ABSTRACT

Introduction and importance: Pituitary apoplexy is a clinical syndrome that occurs secondary to abrupt haemorrhage or infarction and can cause a presentation with headache, visual disturbances, and various neurological deficits. Pituitary apoplexy is a rare clinical syndrome that is often misdiagnosed or discovered late in its clinical course due to its relatively nonspecific clinical presentation.

Case presentation: The case describes patient with a giant prolactinoma which developed haemorrhagic progression in the context of previously undiagnosed coagulopathy from idiopathic thrombocytopaenia. Transcranial (rather than transnasal) surgery was performed for evacuation and debulking.

Conclusion: The case highlights the management of an exceedingly large adenoma in the context of coagulopathy contributing to rapid clinical progression, as well as a positive clinical outcome in a patient with restored vision and pituitary function.

1. Introduction

Pituitary apoplexy (PA) was first described by Bailey after he treated a patient with acromegaly presenting with a sudden headache, visual loss, occulomotor nerve palsy and vomiting [1]. Despite there being a 10–40% identification rate of a precipitating factor in PA, their exact role in pathophysiology is still not fully understood [3]. The presentation of the syndrome is highly variable, making it difficult to diagnose and discriminate from other neurological conditions such as meningitis or stroke. Pituitary apoplexy is a rare clinical syndrome that involves an acute haemorrhage or infarction of the pituitary gland. It occurs in up to 12% of patients with pituitary adenomas and is reported in approximately 21% of non-functional pituitary adenomas [2,3]. The typical presenting features of PA include severe headache (typically retro-orbital–present in 80% of cases) and visual disturbances (in 50% of cases) with bitemporal hemi-anopia and oculomotor-nerve palsies the most frequently observed. Other neurological signs include nausea, vomiting, photophobia, fever, and stroke-like deficits, such as hemiparesis or dysphasia [4]. This case is reported according to the SCARE 2020 criteria [5].

2. Case presentation

A 55-year-old man presented with a three-day history of a severe and gradually worsening retro-orbital headache on a background of type II diabetes and hypertension. It was associated with blurred vision in his right-eye and nausea. He had no significant medical, drug or family history.

Initial examination showed reduced acuity in the right-eye, but cranial nerve/upper/lower limb neurological findings were unremarkable. The patient was haemodynamically stable and underwent investigations.

2.1. Investigations

Haematological investigation revealed profound thrombocytopaenia with a platelet count of 18 (RR 150–400). Previous platelet counts on record were within normal limits. A hormone panel showed a prolactin level of >42,000 mIU/L (RR < 400), a cortisol level of 860 mmol/L (RR < 460) and an ACTH level of 26.9 pmol/L (RR 0–12).

A non-contrast CT brain demonstrated a heterogeneous hyperdense

* Corresponding author at: 30 Mount Street Constitution Hill, Sydney, NSW 2145, Australia.

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mass measuring 47x41x36mm within an enlarged pituitary fossa and extending into the suprasellar cistern with mass effect on the optic chiasm and right cavernous sinus. There was also thinning of the bone of the pituitary fossa with invasion of the right-anterior clinoid process and the sphenoid sinus. He was commenced on high-dose corticosteroids.

2.2. Differential diagnosis

The patient was commenced on intravenous immunoglobulin for presumed idiopathic thrombocytopaenia (ITP) due to low platelet count in the absence of splenomegaly and lymphadenopathy.

Within 4-h of his initial CT scan, the patient complained of complete loss of vision in his right-eye and a worsening headache. On serial examination he showed a right sided oculomotor nerve palsy, was only able to detect light in his right-eye on upward gaze and developed anisocoria with a sluggish direct light-reflex in both eyes. This prompted a progress CT brain which showed interval increase in the right-sided hyperdensity and anterior and posterior aspects of the mass (Fig. 1).

An urgent craniotomy was performed by the consultant neurosurgeon on call via the transcranial approach (due to the large intra-cranial extension) for evacuation of expanding haematoma. A massive haemorrhagic pituitary mass was seen to wrap around perforating branches of the right internal carotid artery and to tent the right optic nerve. Some tumour debulking was necessary to decompress the optic nerve and to control bleeding from perforating vessels. An extra-ventricular drain was left in place for further decompression.

2.3. Outcome

The post-operative period was complicated by refractory-hyperglycaemia (due to the high-dose steroids) and *Staphylococcus capitis* ventriculitis requiring antibiotics. Tumour histopathology demonstrated haemorrhagic pituitary macroadenoma and the patient was commenced on medical therapy in the form of cabergoline which would be highly effective in reducing the macroadenoma. He was unable to maintain his platelet count despite steroids and IVIG: further investigation via an autoimmune blood-panel and a bone-marrow biopsy, did not reveal an alternative diagnosis and a laparoscopic splenectomy was performed for management of steroid-resistant idiopathic thrombocytopaenia performed with intravenous platelet cover. A year from surgery the patient would show normal prolactin levels, significantly improved right-sided vision, and normal appearing pituitary on progress imaging.

3. Discussion

Factors precipitating PA have been identified in approximately 40% of cases, however the condition often occurs in the absence of obvious risk factors [4]. There are four main triggering-factors described within the literature with arterial hypertension said to have the largest impact. Other factors include pituitary stimulation (as in Gonadotropin-releasing hormone analogue use and pituitary testing), disturbances in coagulation (anticoagulation, bleeding disorders and thrombocytopaenia) and reduction in vascular-fluidity (seen during surgical procedures or lengthy admissions). Other common risk factors include diabetes, pregnancy, and head trauma [5].

While the exact pathophysiology of PA is unclear it is known that pituitary tumours have high metabolic requirement and can outgrow their blood supply. Ischaemia or infarction may also occur as the tumour mass expands and compresses on vessels against the sellar diaphragm. The haemorrhagic tendency of the tumour's intrinsic blood vessels also arises from their incomplete maturation and poor fenestration. While typical acute-haemorrhagic PA results from the extravasation of blood into the subarachnoid space, an alternate pathophysiological process exists where a clinically silent necrotic adenoma compresses on neighbouring structures causing destruction to the pituitary gland itself [6].

A degree of endocrine dysfunction is to be expected at presentation, with nearly 80% of patients having a deficiency of one or more anterior pituitary hormones due to mass effect on the gland. Prolactin-secreting tumours are the most common secretory tumour of the pituitary (40% of all pituitary-adenomas) and can are classified further by size (microadenoma [<10 mm], macroadenoma [>10 mm] and giant-prolactinoma [>40 mm]) [5]. This type of tumour is typically benign and well-circumscribed, arising from monoclonal expansion of lactotrophic cells of the posterior pituitary. Management typically involves medical-therapy with dopamine agonists and trans-sphenoidal surgery (if medical-therapy fails or in the context of large tumours in women wanting to become pregnant as pregnancy can stimulate growth).

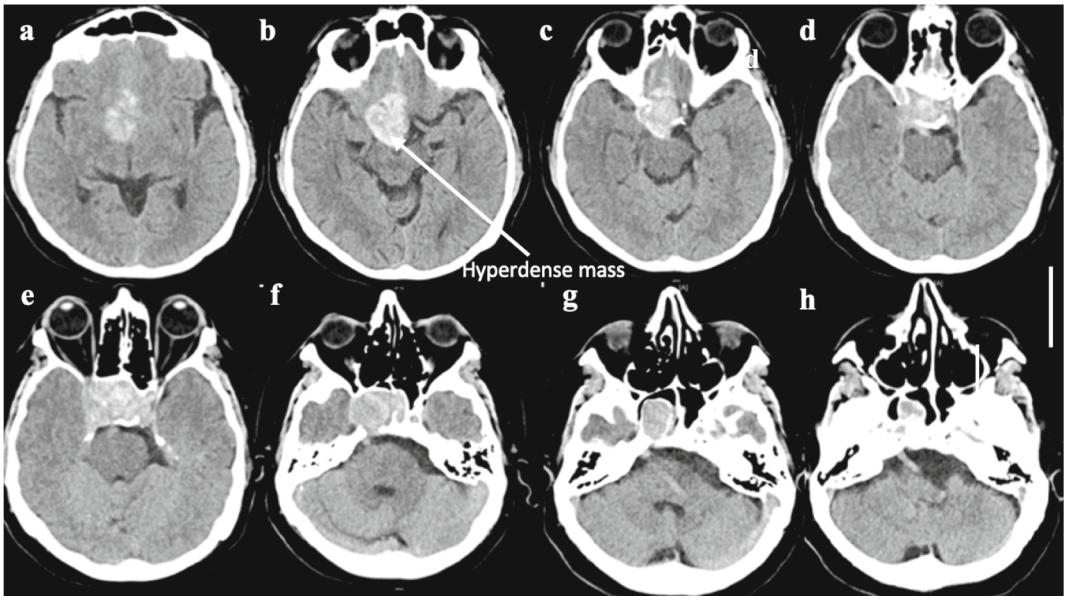

Fig. 1. Coronal sections of the progress non-contrast CT brain labelled a to h (cranial–caudal) showing a large hyperdense mass within the pituitary fossa (scale bar, 5 cm).
Abbreviations: CT, computed tomography.

In retrospect, many of the signs and symptoms of endocrine deficiencies are present but unnoticed prior to the apoplectic episode. Corticotropic insufficiency is the most common deficiency and gives rise to the most severe haemodynamic complications. Adrenal insufficiency leading to haemodynamic-instability and hyponatremia can be life-threatening and hence all patients demonstrating signs of PA should be started on empiric corticosteroid supplementation even before diagnostic confirmation [7]. Other commonly seen deficiencies include thyrotropic and gonadotropic deficiencies, both of which can be addressed once the patient's acute issues are managed.

The two main diagnostic tools used in the diagnosis of PA are computed tomography (CT) and magnetic resonance imaging. Patients presenting with the headache, visual disturbances or neurological deficits will typically undergo CT scan, which is an effective tool to visualize expansive lesions. With the appropriate clinical context and imaging showing a hyperdense lesion in the sella turcica, a diagnosis of PA is highly likely. Conditions such as meningiomas, germinomas, aneurysms and lymphoma may also present with similar CT scan appearance. Diagnosing active bleeding from a pituitary tumour on CT images is difficult with sensitivity ranging from 21% to 46%. Time and clinical context permitting, magnetic resonance imaging is considered a much more sensitive tool for diagnostic purposes [7].

There is no current consensus for the management of PA. Acute treatment involves haemodynamic-stabilization, correction of fluid and electrolyte imbalance and administration of corticosteroids [6].

Following this, surgical decompression is often the most rapid means of achieving a relief of compression of structures such as the optic-chiasm and nearby nerves. Risks specific to decompression include postoperative hypopituitarism and cerebrospinal fluid rhinorrhea (although this risk is not evident post-transcranial procedures).

This case highlights the importance of expedient investigation of visual disturbances as well as the correlation between physical exam, laboratory investigations and clinical imaging.

4. Conclusion

Pituitary apoplexy is a rare clinical syndrome that is often misdiagnosed or discovered late in its clinical course due to its relatively nonspecific clinical presentation. The case highlights the management of an exceedingly large adenoma in the context of coagulopathy contributing to rapid clinical progression, as well as a positive clinical outcome in a patient with restored vision and pituitary function.

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CRediT authorship contribution statement

1. **Dr Oliver Chow:** Study conception; Writing
2. **Dr Therese Pham:** Writing
3. **Brandon He:** Writing; Corresponding author
4. **Dr Amy Hort:** Writing
5. **Dr Damien Gibson:** Writing
6. **Dr Gemma Olsson:** Study conception; Writing.

Declaration of competing interest

Nil.

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