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Case Report: Pituitary Apoplexy causing Panhypopituitarism

Abstract

We present an atypical presentation of pituitary apoplexy as a pontine mimic with drowsiness and pin-point pupils. The patient had a known pituitary macroadenoma with suprasellar extension with hypogonadism and hypothyroidism.

Initial investigations revealed hyponatraemia and imaging suggested a localised cerebral ischaemia in the basal ganglia and no change in the pituitary adenoma. Only further assessment with MRI showed the extent of the haemorrhage. Management was conservative, although close involvement of neurosurgery and ophthalmology teams is essential.

We believe, that awareness of such unusual association is important for clinicians to consider apoplexy in the differential, even if the initial CT imaging is not suspicious.

Key words

Pituitary macroadenoma; pituitary apoplexy; pin-point pupils; hyponatraemia

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Introduction

Pituitary apoplexy is a rare endocrine emergency which can occur due to infarction or haemorrhage of pituitary gland. The term pituitary apoplexy or apoplexia refers to the "sudden death" of the pituitary gland, usually caused by an acute ischemic infarction or hemorrhage. Pearce Bailey described the first case of pituitary tumour-associated hemorrhage in 1898, but the term pituitary apoplexy referring to both necrosis and bleeding into pituitary tumours was first used in 1950 by Brougham et al. [1]

Contributors

Several predisposing or contributing factors for pituitary apoplexy include endocrine stimulation tests, bromocriptine or cabergoline treatment, gonadotropin-releasing hormone treatment, lumbar fusion in the prone position, pregnancy, pituitary irradiation, anticoagulation, thrombocytopenia, and erectile dysfunction medications.[2] Sheehan syndrome occurs in postpartum women in which there is necrosis of the pituitary gland secondary to ischemia after significant bleeding during childbirth. It will present with adrenal insufficiency, hypothyroidism, and hypopituitarism, but rarely with visual changes.[3] Most of the time, this entity is not included as a pituitary apoplexy as the gland did not have a pre-existing tumour, and visual symptoms are extremely rare. Sometimes, it may be the first manifestation of an underlying adenoma. [4]

Visual Involvement

An overview of 186 cases of apoplectic pituitary adenomas, suggested a common presentation with monocular or binocular blindness.[5] The range of vision loss and ophthalmoplegia seen reflects the variability of cranial structures compressed by mass effect. The pathophysiology of extraocular muscle limitation and facial paraesthesia occurs with compression of the cavernous sinus, which contains cranial nerves III, IV, VI, and the ophthalmic branch of V. Blood supply to adjacent structures may be also compromised, causing additional loss of function.[6] New or worse visual field defects are usually found in 67% of the macroadenomas showing increase in size.[7]

Hyponatraemia

Hyponatraemia is an acute sign of hypocortisolism in pituitary apoplexy. However, Syndrome of inappropriate diuretic hormone secretion (SIADH) although uncommon, could appear later as a consequence of direct hypothalamic insult and requires active and individualised treatment. [8]

Natural History

Sixty-eight percentage of patients with a macroadenoma had pituitary hormone deficiency in one or more axes, compared to 42% of those with a microadenoma, when managed conservatively with non-functioning adenomas. [9] Patients with pituitary apoplexy may have spontaneous remission of hormonal hypersecretion.[10]

Case presentation

A 74-year-old man, while on holiday in Blackpool, presented with acute onset of frontal headache, severe around the eyes with radiation posteriorly, blurred vision (mild), transient diplopia, and vomiting. He had a history of a pituitary adenoma managed with thyroxine and testosterone replacement.

Examination - On arrival, he was hyponatraemic, his Glasgow Coma Scale Score was 15, pupils were pinpoint, bilaterally.

Investigations - Initial CT Head - Subtle ischaemic changes in the left capsular basal ganglia region. No acute intracranial haemorrhage or pathology was identified. The previously known large pituitary mass was seen with known suprasellar extension.

CT Head was then repeated as patient was found to be drowsy. There was new increased density in the right posterolateral aspect of the pituitary adenoma which was suggestive of new haemorrhage. The size of the adenoma was unchanged.

Progress - The patient was then put on fluid restriction of 1L/day, had review and advise from endocrinology, ophthalmology and neurosurgery teams. An MRI Head with contrast showed large pituitary lesion with internal haemorrhage /apoplexy with neural compression. Neurosurgeons advised medical management given that visual fields were intact (as reviewed by ophthalmologist). The patient recovered and was discharged in the next 48 hours.

Discussion

Pituitary apoplexy, a rare clinical syndrome secondary to abrupt hemorrhage or infarction, complicates 2%-12% of pituitary adenomas, especially non-functioning tumours.[11] It may present with headache, vomiting, visual disturbances and biochemical as well as hormonal changes. A high index of suspicion is required in patients with known pituitary adenoma.

In our case, the initial CT head scan excluded any other causes such as cerebrovascular disease although there was a suggestion of ischaemic changes in the basal ganglia. Cerebral ischaemic changes as a result of compression by suprasellar extension of large macroadenomas is not uncommon. The initial CT scan did not show any change in size of the macroadenoma.

The pinpoint pupils and drowsiness, usual features indicative of brainstem or pontine involvement, were an unusual presentation in this case. This can be seen in patients with encephalopathy due to various causes and are mimic of pontine lesion.[12] Brain MRI often show infarctions at the midbrain and thalamus. Most patients with pituitary apoplexy however present with ptosis, followed by limited gaze and diplopia. Oculomotor nerve palsy usually occurs in patients with apoplectic adenomas, especially those with hemorrhage. Most commonly affected of the ocular motor nerves is the third cranial nerve, which can be total or partial- pupil sparing. The *levator palpebral superioris* is the most commonly affected, causing either a partial or complete ptosis. [13] Early treatment, pupil-sparing, and minor oculomotor symptoms are factors

associated with a good recovery. [14]

In this case the biochemistry suggested syndrome of inappropriate diuretic hormone secretion leading to hyponatraemia. This is rare and hypocortisolism is usually the likely cause of hyponatraemia in majority of presentations. An atypical presentation of pituitary apoplexy, also known as 'subclinical pituitary apoplexy,' may be considered and requires careful evaluation for hyponatraemia using serial urine osmolality, which is useful to distinguish hypovolaemic hyponatraemia from euvolaemic hyponatraemia.[15]

Pituitary apoplexy should be managed as a medical emergency and the medical team must have a high suspicion for the condition. They should initiate Hydrocortisone (to prevent adrenal insufficiency), urgent ophthalmology evaluation of visual field defects or oculomotor insufficiency. Urgent brain imaging (preferably MRI) is essential to exclude alternative diagnoses and especially in cases of atypical presentations. Early involvement of Neurosurgical team is essential and often the cornerstone of treatment. There are some who advocate early transphenoidal surgical decompression for all patients, whereas others adopt a more conservative approach for selected patients (those without visual acuity or field defects and with normal consciousness).[16] In our case, the patient responded to non-surgical management.

The 'watch and wait' policy seems reasonable for microadenomas but is probably not a safe approach for macroadenomas, which appear to have a significant growth potential; in these cases, given the lack of established medical treatment, the decision for surgical intervention should balance the presence of significant comorbidities and the anaesthetic/peri-operative risks at presentation against the probability of tumour enlargement and its consequences, as well as the possible loss of advantages associated with early operation.[7]

Learning points

- When a patient with a known pituitary adenoma presents with severe headache and visual disturbances, one should consider Pituitary apoplexy as a top differential.
- While oculomotor palsy, visual field defects and hyponatraemia due to acute hypocortisolism are common, drowsiness and pinpoint pupils are rare and require careful evaluation by imaging.
- Early involvement of ophthalmologist and neurosurgeons are key to safe management. Awareness of this rare condition may help reduce the delay in diagnosis and management of this condition.

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