Pituitary apoplexy

Key concepts

- due to the expansion of a pituitary adenoma from hemorrhage or necrosis
- typical presentation: paroxysmal H/A with endocrinologic and/or neurologic deficit (usually ophthalmoplegia or visual loss)
- management: immediate administration of glucocorticoids, and transsphenoidal decompression within 7 days in most cases.

Definition

Pituitary apoplexy is characterized by a sudden onset of headache, visual symptoms, altered mental status, and hormonal dysfunction due to acute hemorrhage or infarction of a pituitary gland.

Epidemiology

In Wilson’s series, 3% of his patients with macroadenomas had an episode of pituitary apoplexy. In another series of 560 pituitary tumors, a high incidence of 17% was found (major attack in 7%, minor in 2%, asymptomatic in 8%) \(^1\). It is common for apoplexy to be the initial presentation of a pituitary tumor \(^2\).

The very low incidence of this complication hinders formulation of widespread guidelines on diagnostic and therapeutic management \(^3\).

This condition results in an estimated 1.5-27.7% of cases of pituitary adenoma, although the figure is probably closer to 10% \(^4\).

Mohr and Hardy noted typical symptomatic pituitary apoplexy to occur in only 0.6% of patients with significant hemorrhagic and necrotic changes in 9.5% of surgical specimens.

Frequency of intratumoral hemorrhage increases to 26% if using only MRI criteria without clinical evidence of apoplexy. However, hemorrhagic pituitary apoplexy may be fatal. Kurisu et al reported a
68-year-old man who developed pituitary apoplexy resulting in massive intracerebral hemorrhage and death 1 month later 5)  

**Sex**  

The male-to-female predominance is 2:1.  

**Age**  

The age range is 37-57 years.  

Male sex, non-functioning tumor, and macroadenoma are associated with an increased risk 6).  

**Classification**  

It is important to note that pituitary apoplexy may be divided into hemorrhagic or ischemic, each with unique neuroimaging findings.  

**Etiology**  

Sudden intrasellar expansion may occur as a result of hemorrhage, necrosis 7) 8) and/or infarction within a pituitary tumor and adjacent pituitary gland. Occasionally, hemorrhage occurs into a normal pituitary gland or Rathke’s cleft cyst 9).  

This condition stems from an acute expansion of a pituitary adenoma or, less commonly, in a nonadenomatous gland, from infarction or hemorrhage. The anterior pituitary gland is perfused by its portal venous system, which passes down the hypophyseal stalk. This unusual vascular supply likely contributes to frequency of pituitary apoplexy.  

**Pathophysiology**  

Some postulate that a gradual enlarging pituitary tumor becomes impacted at the diaphragmatic notch, compressing and distorting the hypophyseal stalk and its vascular supply. This deprives the anterior pituitary gland and the tumor itself of its vascular supply, apoplectically causing ischemia and subsequent necrosis.  

Another theory stipulates that rapid expansion of the tumor outstrips its vascular supply, resulting in ischemia and necrosis. This explanation is doubtful, since most tumors that undergo apoplexy are slow growing.  

Cerebral ischemia due to pituitary apoplexy is very rare. It may be caused by vasospasm or direct compression of cerebral vessels by the tumor.
Clinical features

DI may occur with pituitary apoplexy.

The clinical presentation varies widely and includes asymptomatic cases, classical pituitary apoplexy and even sudden death.

It is characterized by a sudden onset of headache, visual impairment, mental disorder, and hormone dysfunction due to acute hemorrhage or infarction of a pituitary gland.

Because of the acute symptomatology, many patients are referred to a neurosurgical department without prior endocrinological assessment.

Neurologists and neurosurgeons need to be aware of the endocrinological sequelae of pituitary apoplexy in order to avoid potentially lethal complications. Patients should be counselled to adhere to long-term endocrinological and neurosurgical follow-up.

An existing pituitary adenoma is usually present. The visual symptoms may include both visual acuity impairment and visual field impairment from involvement of the optic nerve or chiasm and ocular motility dysfunction from involvement of the cranial nerves traversing the cavernous sinus.

Paschou et al present a patient in his late 30s presented with sudden and severe frontal headache, fever, blurred vision, nausea, confusion, as well as oculomotor nerve palsy (CN III) with partial ptosis of the left eyelid, dilated left pupil and left eye globe deviation inferiorly and laterally. The final diagnosis was acute pituitary apoplexy complicating a pituitary macroadenoma. In this setting, headache is usually present due to stretching and irritation of the dura mater, and fever due to meningeal irritation or upward expansion leading to hypothalamic dysfunction. Decreased visual acuity and defects in visual fields are caused by upward expansion, which compresses the optic chiasm. Ophthalmoplegia can also be observed due to lateral expansion with invasion of the cavernous sinus.

Diagnosis

CT or MRI shows hemorrhagic mass in sella turcica and/or suprasellar region, often distorting the anterior third ventricle.

Cerebral angiography should be considered in cases where differentiating pituitary apoplexy from aneurysmal SAH is difficult.

Differential diagnosis

Seung et al., present an unusual case of bitemporal hemianopsia caused by a large anterior communicating artery aneurysm.
A 41-year-old woman was admitted to our neurosurgical department with a sudden-onset bursting headache and visual impairment. On admission, her vision was decreased to finger counting at 30 cm in the left eye and 50 cm in the right eye, and a severe bitemporal hemianopsia was demonstrated on visual field testing. A brain computed tomography scan revealed a subarachnoid hemorrhage at the basal cistern, and conventional cerebral catheter angiography of the left internal carotid artery demonstrated an 18×8 mm dumbbell-shaped aneurysm at the ACoA. Microscopic aneurysmal clipping was performed. An ACoA aneurysm can produce visual field defects by compressing the optic chiasm or nerves.

Seung et al., emphasize that it is important to diagnose an aneurysm through cerebrovascular study to prevent confusing it with pituitary apoplexy 13).

A 52-year-old woman, previously diagnosed with asymptomatic Rathke cleft cyst (RCC), came with a severe headache, along with visual dysfunction and symptoms of pituitary insufficiency. Fluid-attenuated inversion recovery magnetic resonance imaging demonstrated diffuse hyperintensity in the cerebral cisterns, whereas watery clear cerebrospinal fluid was obtained by lumbar puncture. Surgery performed 1 month after onset revealed a nonhemorrhagic lesion, with a final diagnosis of nonhemorrhagic RCC rupture.

Yokota et al., conclude that nonhemorrhagic RCC rupture and subsequent leakage of the contents into subarachnoid space were the underlying pathogenesis in the present case of RCC resembling apoplexy 14).

**Treatment**

Pituitary apoplexy treatment.

**Outcome**

see Pituitary apoplexy outcome.

**Case series**

Lammert et al., analysed data from 24 patients (m:f/16:8) with a median age of 64 yrs (38 to 83yrs) that underwent surgery for pituitary apoplexy regardless of time from symptom onset. Apoplexies were necrotic in 14 cases and haemorrhagic in 10 cases.

Preoperatively, 7 patients (29.2%) showed complete anterior pituitary insufficiency, 16 patients (66.6%) had partial anterior pituitary insufficiency and one patient (4.17%) had normal pituitary functions. Persistent panhypopituitarism was found in 7 patients (29.2%), whereas an overall improvement of pituitary function was noted in 13 (57.1%) patients. Preoperative prolactin (PRL) levels were significantly associated with recovery of endocrine functions, whereas specifically all patients with preoperative PRL levels of at least 8.8 ng/ml recovered partially or fully. Time to surgery (0-7 days vs. 1-4 weeks vs. >4 weeks) was not significantly associated with outcome.
The data emphasize that normal and high preoperative PRL levels are associated with better endocrine outcome after surgery. They conclude that patients benefit from surgical intervention even after delayed diagnosis with the serum PRL levels is being a valid biomarker for clinical decision making.

Case reports

Pituitary apoplexy case reports.

References

