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CASE REPORT

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Abstract

A 37 year old lady presented to the hospital with severe headache. The lady had prior episodes of headache that lasted for 1 to 2 days and was associated with nausea, vomiting, photophobia and phonophobia. She was on treatment for episodic migraine. However, this time she had a severe and worse headache which was not improving on her regular medications. She had associated diplopia that she noted initially and had ptosis at the time of presentation to the hospital as well. Lady had ptosis with medial rectus palsy, with normal size normal reaction of pupil. There was mild frontal sinus tenderness. Pituitary tumours are most common type of disorders. The most common disorders that bring a patient to the hospital are pituitary adenoma. The emergency, pituitary apoplexy is a significant aetiology as well, that brings a person to a neurosurgeon. Pituitary apoplexy is defined as clinical syndrome that may include headache, visual defects, altered mental status. The symptoms may develop rapidly or evolve slowly over days to weeks. The most characteristic presenting features are inappropriate pituitary hormone secretion and visual field effects. Less commonly headache and subtle signs of pituitary hormone deficiency, such as amenorrhea, loss of libido and lethargy, are the presenting symptoms. Temporal lobe epilepsy, cranial nerve palsies, CSF rhinorrhea and hydrocephalus are rarer features. However, isolated third nerve palsy with ptosis are rare presenting feature.

Keywords: Pituitary Tumor, Pituitary Adenoma, Third Nerve Palsy, Sella Turcica, Cavernous Sinus

Introduction

Pituitary tumours are most common type of disorders. The most common disorders that bring a patient to the hospital are pituitary adenoma. The emergency, pituitary apoplexy is a significant aetiology as well, that brings a person to a neurosurgeon [1]. Pituitary apoplexy is defined as clinical syndrome that may include headache, visual defects, altered mental status. The symptoms may develop rapidly or evolve slowly over days to weeks [2]. Visual field effects and inappropriate pituitary hormone secretion are the most characteristic presenting features. Less commonly headache and subtle signs of pituitary hormone deficiency, such as amenorrhea, loss of libido, lethargy, is the presenting symptoms. Temporal lobe epilepsy, cranial nerve palsies, CSF rhinorrhea and hydrocephalus are rarer [3]. However, isolated third nerve palsy with ptosis is a rare presenting feature [2]. More common causes of isolated third nerve palsy include intracranial and subarachnoid hemorrhage, cavernous sinus thrombosis, bacterial meningitis and midbrain infarction [4].

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Case Report

A 37-year-old lady presented to the hospital with severe headache. The lady had prior episodes of headache that lasted for 1 to 2 days and was associated with nausea, vomiting, photophobia and phonophobia. She was on treatment for episodic migraine. However, this time she had a severe and worse headache which was not improving on her regular medications. She had associated diplopia that she noted initially and had ptosis at the time of presentation to the hospital as well. Lady had ptosis with medial rectus palsy, with normal size normal reaction of pupil (Figure 1). There was mild frontal sinus tenderness. There was no focal weakness, no sensory symptoms and signs, normal reflexes. No papilledema.

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In view of the red flags of migraine, she was admitted and MRI brain, MRV, and MRI orbit were planned. The MRI (Figure 2) was suggestive of well defined T2 hypertensive 2.1×1.7×1.5 cm lesion in sella extending into suprasellar cistern that was abutting cavernous sinus and ICA suggestive of pituitary macroadenoma. MRV brain was normal. MRI Orbits showed there were T1 bright components on superior aspect of mass with compression of medial aspect of right cavernous sinus. No mass effect of optic chiasma or optic nerves. Bilateral grade 1 extension into cavernous sinus is present. Pituitary macroadenoma didn't cross the median intercarotid line on either side. However, involvement of right cavernous sinus was more on the right side.

The lady was referred to neurosurgery department for further management.

Discussion

Pituitary gland is inferior to hypothalamus in the Sella Turcica. Optic chiasma lies superior to it and the pituitary tumor leads to compression of optic nerves and optic chiasma. Progressive visual deterioration of visual field is the principal criterion. Humphrey computerized visual fields are useful. It should be done even if there is no contact between optic pathways and pituitary mass. The visual defects may occur due to prior impingement, possible and impending vascular shunting, or displacement of optic chiasma following the decompression [3]. The headache may occur due to the mass effect or the meningeal involvement [1,2]. The optic chiasma compression leads to the visual field defects, most commonly bitemporal hemianopia. As dense bitemporal loss may reduce binocular field by only 25%, Esterman's field are important to objectively assess fitness to drive [3]. Decreased visual acuity is a result of severe compression or direct invasion of optic nerve leads [1].

Pituitary Apoplexy is a result of either acute ischemic or haemorrhagic vascular accident involving the pituitary adenoma or adjacent pituitary gland. Pituitary adenoma is particularly prone to haemorrhage and necrosis [2]. Cavernous sinus lies lateral to the pituitary gland. It houses the cranial nerves third, fourth, fifth and sixth. Cranial neuropathies are the commonest neurological manifestations. The cranial nerves 2nd, 3rd, 4th, 5th, 6th and 7th may be affected in various combinations [4]. A lateral expansion of the adenoma can affect these 3 cranial nerves [1].

Isolated 3rd cranial nerve palsy as a presenting sign of pituitary tumour is rare. The mechanisms may include compression of lateral wall of the cavernous sinus. However, this usually occurs late in the course of the disease. Direct invasion of tumour through sinus wall may also occur. The mechanical compression of the 3rd cranial nerve against the unyielding inter-clinoid ligament of cavernous sinus wall tends to bring about a slow onset nerve palsy. Sudden onset of 3rd cranial nerve palsy has been attributed to the compromise of the vascular supply to the nerve due to compression of the vasa nervosum originating in the Internal Carotid Artery (ICA) [2, 5]. A rapid expansion of the tumour secondary to tumour growth may also lead to acute compression. As per a series, the average growth in tumour diameter is 0.6 mm per year [5].

The crucial differential diagnosis include- intracranial hemorrhage, carotid artery aneurysm, subarachnoid hemorrhage, bacterial meningitis, cavernous sinus thrombosis, midbrain infarction, PCA aneurysm [2]. Early diagnosis and treatment remains the cornerstone of treatment of Pituitary apoplexy. This includes iv steroids till surgery to avoid acute adrenaline insufficiency. Neurosurgical decompression by trans-sphenoidal approach is the definitive treatment. Early decompression may partially or completely restore pituitary function and preserve 3rd cranial nerve function [2]. The three primary treatment goals for pituitary adenomas are- reducing hormone hyper secretion and its clinical manifestations, decreasing tumor size to improve the symptoms of mass effect, and correcting hormone deficiencies [1].

Most prolactinomas can be treated medically with dopamine agonists. Surgery is reserved for few patients who either don't tolerate the medications, or who develop pituitary apoplexy despite medical treatment, or whose macroprolactinomas do not respond to medical treatment [3]. ACTH and growth hormone secreting tumors are not as responsive to medical management and surgery is

the principal treatment [1]. Depending on the type of hormone secreted by the tumour, other therapies are now available with Somatostatin Analog, Growth Hormone Receptor Blockers, Cortisol Receptor Blocker, and Enzyme Inhibitor [6-8]. Progress in Pluripotent stem cell-based works ensures novel disease models for pituitary adenoma are bound to be detected. Drug discovery and clinical breakthrough may result [7].

Conclusion

Presented herewith is the case of this middle-aged lady who developed sudden answered severe headache, different from her regular migraine episodes, associated with isolated 3rd cranial nerve palsy. Such presentation is commonly due to pituitary apoplexy. However, the lady had no acute vascular event. The lady was found to have pituitary macro adenoma and this is an unusual case since she had isolated oculomotor nerve palsy without involvement of any other cranial nerves. The sudden onset may have been due to the involvement of ICA and vasa nervosum that arises from ICA.

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Figures

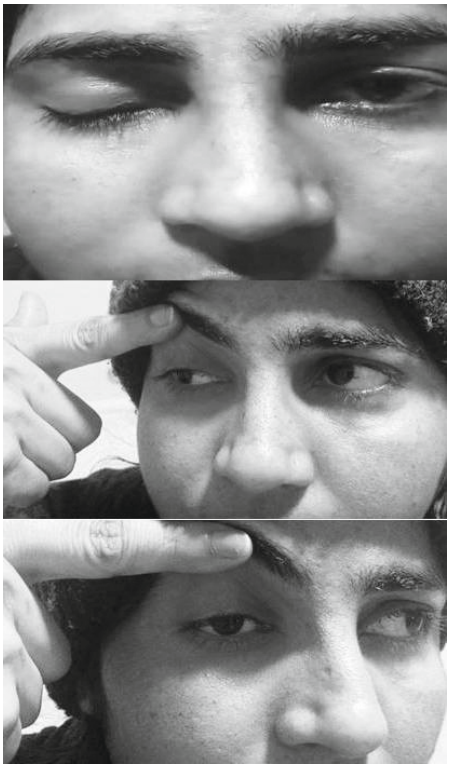

Figure 1: Eye signs of the patient: right eye ptosis and right eye medial rectus palsy can be made out.

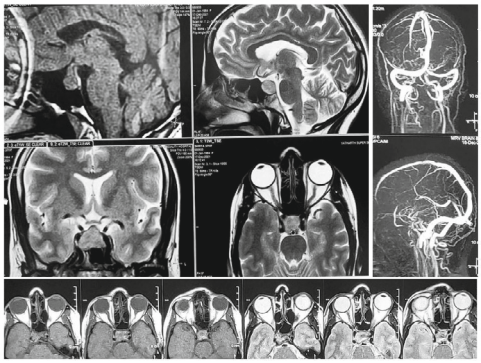

Figure 2: MRI images of the patient: lesion in sella extending into suprasellar cistern abutting the cavernous sinus and ICA suggestive of pituitary macroadenoma. The MRV brain was normal.