Hemorrhage into Craniopharyngioma as a Differential Diagnosis of Pituitary Apoplexy: A Case Report and Literature Review

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ABSTRACT

Published on 26th March 2015

Hemorrhage occurring in craniopharyngioma, with clinical presentation similar to pituitary apoplexy, is a very rare event; with only 20 cases reported. Hyponatremia, hemiplegia and upper motor neuron facial palsy occurring in such a situation is still rarer. We report the case of a 60-year-old lady who presented with right hemiplegia, diminished vision, right upper motor neuron facial palsy and hyponatremia. Computed tomography showed a hypodense lesion in the sellar and supra-sellar region with hemorrhage extending to left basal ganglia. Magnetic resonance imaging revealed a T2-hyperintense lesion with peripheral contrast enhancement in the sellar and suprasellar regions. The pre-operative diagnosis was pituitary macroadenoma with hemorrhage. The relatively avascular tumor was excised sub totally by craniotomy. Histopathological examination of the specimen revealed adamantinomatous type of craniopharyngioma. Literature review was based on 17 publications and a total of 20 patients. Their clinical features, locations of the tumor and histological types are tabulated.

Keywords: Craniopharyngioma, Pituitary apoplexy, Hyponatremia, Adamantinomatous

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Key Messages:

Craniopharyngioma presenting with intracranial hemorrhage and features of pituitary apoplexy and hemiplegia is very rare. This case report describes the clinical features, imaging and surgery done for such a patient.

INTRODUCTION

Craniopharyngioma account for 3.4% of all intracranial tumors.[1]These are mostly benign tumors arising in the sellar and suprasellar regions, some of which exhibit locally invasive properties. Intratumoral hemorrhage in these is very rare and only 20 such cases have been previously reported in world literature.[2-18] If such an event occurs, most patients present with features of pituitary apoplexy. Only one previous study[16]quotes such an event occurring in an adamantinomatous craniopharyngioma. A case in which hemorrhage occurring in a craniopharyngioma, extending to the capsulo-ganglionic region, and patient presenting with hemiplegia, facial palsy and hyponatremia has never been reported previously.

CASE HISTORY

A 60-year-old lady, hypertensive and diabetic, presented with one-week duration of right-sided weakness and sudden onset altered sensorium. Examination revealed cataract in her right eye and pseudophakia in left.

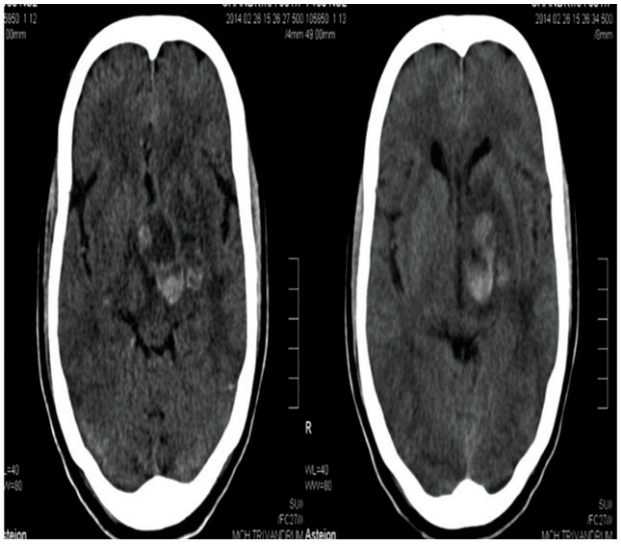

Figure 1. **Non-contrast Computed Tomogram brain images showing hypodense supra-sellar lesion with hemorrhage extending to left basal** ganglia

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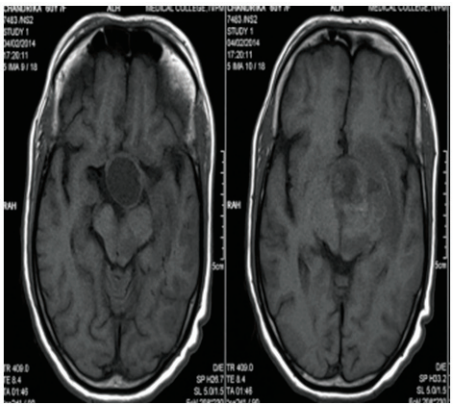

Figure 2a. **T1 weighted image (axial) showing the hypointense intra-sellar lesion extending to supra-sellar region**

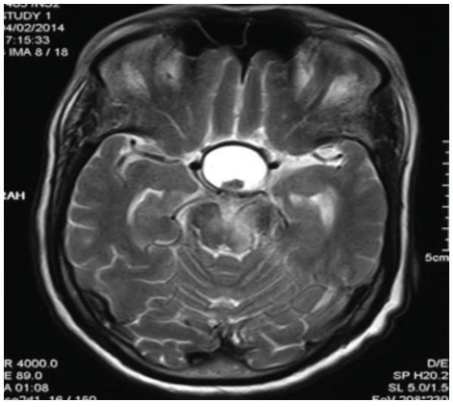

Figure 2b. **T2 weighted image (axial) showing the hyperintense lesion with nodular hypointense areas**

Glasgow Coma Scale (GCS) score at presentation was E4V1M5 and had only perception of light in left eye. She was having right-sided upper motor neuron type of facial palsy and right hemiplegia.

Blood investigations revealed that her serum sodium was 120 mEq/L and serum potassium 2.2 mEq/L. Renal function was normal. Her hormonal profile results (with reference range in parentheses) were Thyroid-stimulating hormone (TSH) – 0.48 microIU/ml (0.25-4.75), Free T3 – 1.48 pg/ml (1.82-4.62), Free T4 – 0.79 ng/dL (0.932-1.71) and Cortisol (morning) – 0.32 μg/dL (6.22-19.4). Pituitary hormone profile was normal for her age. Workup of hyponatremia led to the diagnosis of syndrome of inappropriate anti- diuretic hormone secretion.

Computed Tomography (CT) scan of the brain showed a well-defined lesion in the sellar and suprasellar region (Figure 1) with hematoma extending from the superior aspect to the left capsulo-ganglionic region with perilesional edema. Magnetic Resonance Imaging (MRI) of the brain revealed in T2-weighted image, a heterogeneously hyperintense lesion involving the sella and suprasellar region. The lesion (3.2 × 4.6 × 3.6 cm) was extending to the left capsulo-ganglionic region with hemorrhage (Figures 2a, 2b). Post-contrast moderate peripheral enhancement was seen with perilesional edema (Figures 2c, 2d). The pituitary and its stalk were not visualized separately. Magnetic Resonance Angiogram showed that both the optic nerves and anterior cerebral arteries were displaced laterally by the lesion (Figure 3). These suggested the possibility of pituitary macroadenoma with hemorrhage extending to left capsulo-ganglionic region.

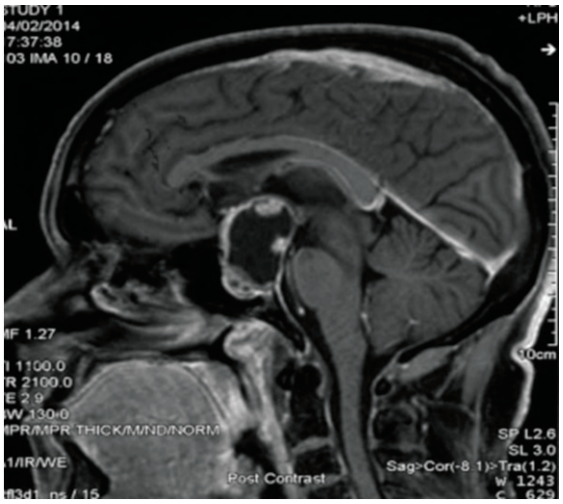

Figure 2c. **Post-contrast T1 weighted image (Sagittal) showing the vertical extent of the lesion**

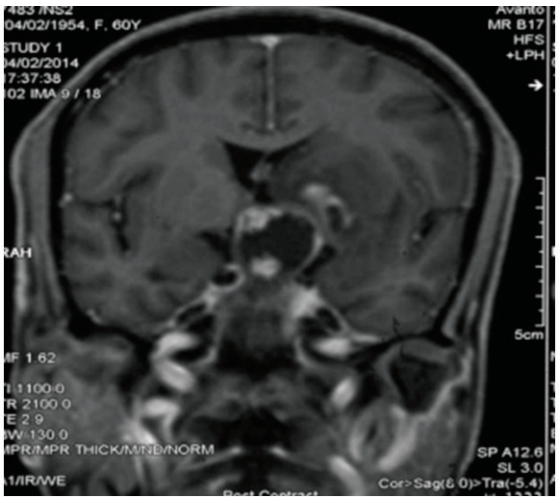

Figure 2d: **Post-contrast T1 weighted image (coronal) showing the horizontal extent of the lesion**

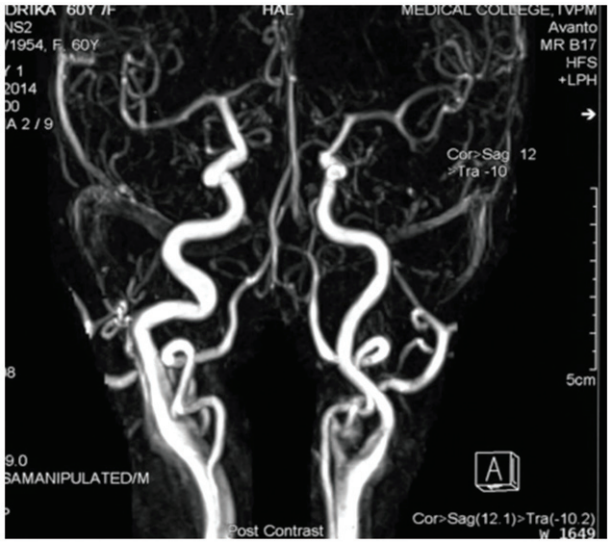

Figure 3. **Magnetic Resonance Angiogram showing the anterior cerebral arteries being splayed by the lesion**

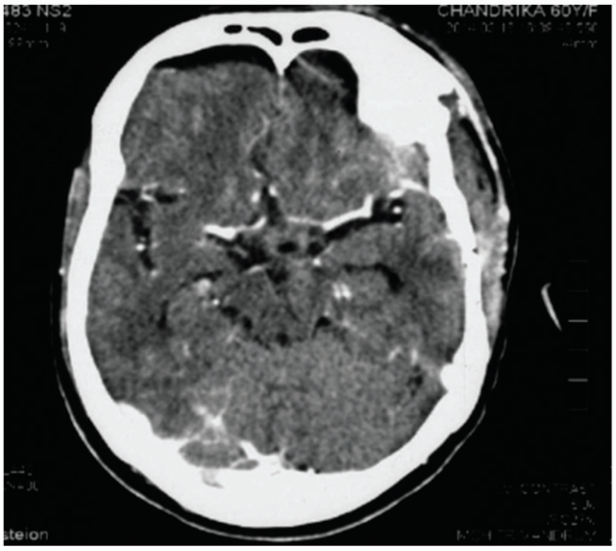

Figure 4. **Two months post-operative contrast Computed Tomogram brain showing resolution of both the lesion and hemorrhage**

Hyponatremia was corrected with fluid restriction and hypertonic saline (3%) infusions; and hypokalemia with potassium chloride. She was started on oral thyroxine, steroid, anticonvulsant and antiedema measures. Her GCS score improved to E4V2M6. She underwent left pterional craniotomy for excision of the tumor. Tumor was seen in the inter-optic space and had variable consistency, with some areas firm and others cystic. The cyst was aspirated draining a straw-colored clear fluid. Firm areas were relatively avascular with well-defined plane with surrounding parenchyma in most areas. It was adherent to left internal carotid artery and left optic nerve. The tumor was excised sub-totally, after which the optic nerves were found to be lax.

Post-operatively, she was fully conscious, had grade 2 power on right side and facial palsy had resolved. Hyponatremia got corrected. Hormonal values after six days of surgery were as follows: TSH – 0.21 mIU/ml, Free T3 – 2.05 pg/ml, Free T4 – 1.3 ng/dL and Cortisol –0.4 μg/dL. At the end of two months, they were respectively as follows: 0.02 mIU/ml, 94.04 ng/dL, 11μg/dL and 1.37 μg/dL. Sodium and potassium values had normalized. Post-operative CT brain showed significant resolution of the lesion and hematoma (Figure 4). She was followed up for a period of two months, at the end of which her GCS was E4V5M6 without motor deficits. Thyroxine was continued; steroid tapered and continued on a low dose. But she got only slight improvement in the vision of left eye and was lost to follow-up after two months.

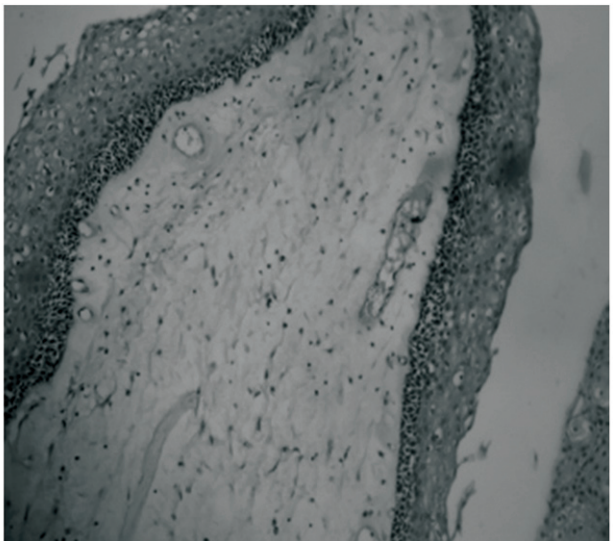

Figure 5a. **Photomicrograph of the lesion (20 x magnification) showing the basaloid cells and stellate reticulum**

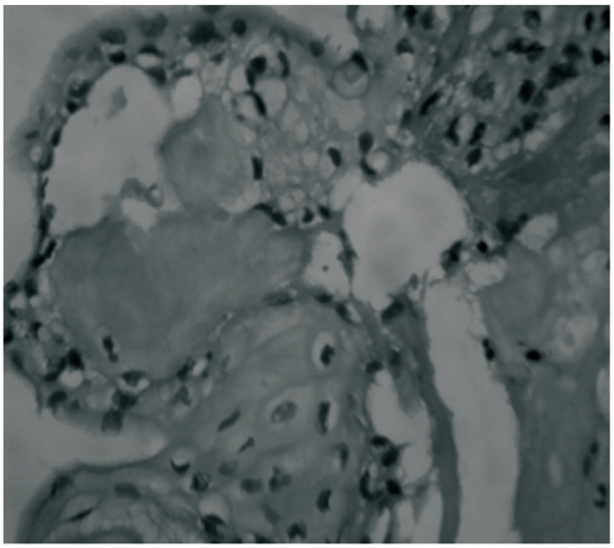

Figure 5b. **Photomicrograph of the lesion (100 x magnification) showing the characteristic "wet" keratin**

Histopathological examination of the specimen showed a neoplasm composed of stratified squamous epithelium along with stellate reticulum (Figure 5a). Wet keratin (Figure 5b) and fibro-collagenous tissue with inflammatory infiltrate were noted, confirming the diagnosis of adamantinomatous craniopharyngioma.

DISCUSSION

Craniopharyngiomas are relatively benign circumscribed epithelial neoplasms of the sellar region that arise from embryonic squamous cells of the hypophysiopharyngeal duct. Pituitary apoplexy is a clinical syndrome characterized by the abrupt onset of characteristic signs and symptoms, most commonly headache, nausea, visual disturbance, and ophthalmoplegia, in association with hemorrhage or infarction within the pituitary fossa.[19] Some craniopharyngiomas may extend from intra- to the suprasellar region. If such tumors bleed, the patient can present with features

Table 1. Showing relation between symptoms and location of pathology

Case number	Author (Year)	Age	Sex	Symptoms	Relation to sella	Pathology
1.	Kissel et al.,1965 [5]	?	?	Oculomotor paralysis	?	?
2.	Lloyd et al.,1977 [7]	?	?	?	?	?
3.	Kubota et al.,1980 [6]	?	?	?	Supra-sellar	?
4.	Wakai et al.,1982 [15]	>15y	?	?	Intra-sellar	?
5.	Kellen et al.,1988 [4]	37 y	M	Diplopia	Supra- and intra-sellar	Sq
6.	Yamamoto et al., 1989 [16]	59 y	F	Headache, nausea	Intra-sellar	Ad
7.	Yousem et al.,1990 [18]	16m	?	Visual disturbance	Supra- and intra-sellar	?
8.	Yousem et al.,1990 [18]	1-23y	?	?	Intra-sellar	?
9.	Masuda et al.,1990 [9]	63 y	F	Bitemporal hemianopsia	Supra- and intra-sellar	Sq
10.	Podgórski et al., 1991 [12]	?	?	?	Intra-sellar	?
11.	Makwane et al., 1996 [8]	46 y	M	Headache, vomiting	Supra- and intra-sellar	?
12.	Ishii et al.,1999 [3]	44 y	F	Headache, bitemporal hemianopsia	Supra- and intra-sellar	Sq
13.	Nishioka et al.,2000 [11]	49 y	F	Headache, nausea, visual disturbance	Supra- and intra-sellar	Sq
14.	Haraguchi et al., 2000 [2]	70 y	F	Headache, drowsiness, hyponatremia	Supra- and intra-sellar	Sq
15.	Yamashita et al., 2004 [17]	22 y	M	Bitemporal hemianopsia, deterioration of visual acuity	Supra- and intra-sellar	Sq
16.	Yamashita et al., 2004 [17]	29 y	F	Headache, bitemporal hemianopsia	Supra- and intra-sellar	Sq
17.	Rangel-Castilla et al., 2004 [13]	?	?	Visual disturbance	Supra- and intra-sellar	?
18.	Nielsen et al.,2013 [10]	<15y	M	?	?	?
19.	Nielsen et al.,2013 [10]	<15y	M	?	?	?
20.	Tosaka et al.,2014 [14]	69 y	M	Headache, blurred vision, unsteady gait	Supra-sellar	Sq
21.	Present case, 2014	60 y	F	Headache, altered sensorium, visual disturbance, right hemiplegia, right UMN facial palsy, hyponatremia	Supra- and intra-sellar	Ad

(? - Details unavailable, y - years, m - months, M - Male, F - Female, Ad - Adamantinomatous, Sq - Squamous - papillary, UMN - Upper Motor Neuron)
Courtesy: Table 1 in the report by Yamashita et al[17]

of pituitary apoplexy. Causes attributed to intratumoral hemorrhage include degenerative changes in blood vessels and the presence of numerous immature blood vessels.[11]

We made a PubMed and J-STAGE search using the key words "craniopharyngioma, tumor, hemorrhage" and "craniopharyngioma, pituitary apoplexy". Those studies demonstrating hemorrhage in craniopharyngioma with clinical presentation of pituitary apoplexy were reviewed. This revealed a total of 20 cases (Table 1) (including four in Japanese, one in French, one in Spanish and one in Polish language). So this becomes the fourteenth such case in English literature. This is the first case report of such an event occurring in an adamantinomatous type, other than that reported by Yamamoto et al.[16] Also, only one case has been reported till date in which there was associated hyponatremia.[2] No other case report states a patient developing hemiparesis secondary to hemorrhage in the suprasellar aspect of craniopharyngioma tracking into basal ganglia.

On reviewing the literature (including our case), the rare entity of craniopharyngioma bleed has been reported mostly in adults (75%) and most of them presented with an initial symptom of headache (57%). Females dominated the picture (54%). Most of the tumors were intra- as well as supra-sellar in location (64%). Squamous histology predominated in most situations (80%). The usual visual complaints were bitemporal hemianopsia, decreased visual acuity and diplopia.

It may be concluded that hemorrhage into an intra or suprasellar craniopharyngioma should be considered as differential diagnosis of pituitary apoplexy and imaging should be done promptly to differentiate between the two. Changes in hormonal profile can occur in both and it may be difficult to differentiate between these two entities preoperatively.

END NOTE

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Conflict of Interest: None declared

Editorial Comments

The case is being reported as 21st of its type in literature. Considering its rarity, it is apt that it should be brought to the attention of medical fraternity.

Cite this article as: *Das, Joe M, Rajmohan BP, Krishna Balachandran, Peethambaran, Anilkumar. Hemorrhage into Craniopharyngioma as a Differential Diagnosis of Pituitary Apoplexy: A Case Report and Literature Review. Kerala Medical Journal. 2015 Mar 26;8(1):33-37*

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