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## CASE REPORT

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# Cavernoma of the Hypothalamus

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### ABSTRACT

*Intracranial cavernoma is a relatively rare condition. Cavernoma of the hypothalamus is even rarer. We report a case of hypothalamic cavernoma presenting with ptosis.*

*Key Words: Brain neoplasms; Cerebral hemorrhage; Hemangioma, cavernous*

## 中文摘要

### 下丘腦海綿狀血管瘤

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顱內海綿狀血管瘤較為罕見，下丘腦海綿狀血管瘤更極為罕有。本文報告一宗出現眼皮下垂的下丘腦海綿狀血管瘤的病例。

### CASE REPORT

A 32-year-old woman presented with a one-month history of right-sided ptosis in June 2008. She had no other neurological symptoms such as visual disturbances, seizures, headaches, weakness, or sensory loss. Moreover, there was no history of nausea or vomiting.

An ophthalmology review revealed a 1-mm ptosis of the right eye, no ophthalmoplegia and no visual field defect. Her pupils were equal and reactive. Fundoscopy examination yielded nil abnormal.

Hormonal assays yielded levels in the normal range: beta-human chorionic gonadotropin <2 IU/l (reference level, <5 IU/l), prolactin 10.5 µg/l (1-25 µg/l), cortisol

345 nmol/l (140-690 nmol/l), free triiodothyronine 3.73 pmol/l (4.0-7.4 pmol/l), free thyroxine 14.99 pmol/l (10-18 pmol/l).

Magnetic resonance imaging (MRI) [0.35T, Magnetom C; Siemens, AG, Erlangen, Germany] was performed, and showed a suprasellar mass arising from the hypothalamic region, posterior to the optic chiasm (Figure 1). The optic chiasm was displaced superiorly (Figure 2) and the pituitary stalk was displaced to the left. This mass demonstrated reticulated cores of low and high signal intensity consistent with areas of blood products (haemorrhagic) surrounded by a hypointense haemosiderin rim on T2-weighted images (T2WI). No significant enhancement was evident in the post-contrast scan. The pituitary gland was otherwise normal. Based

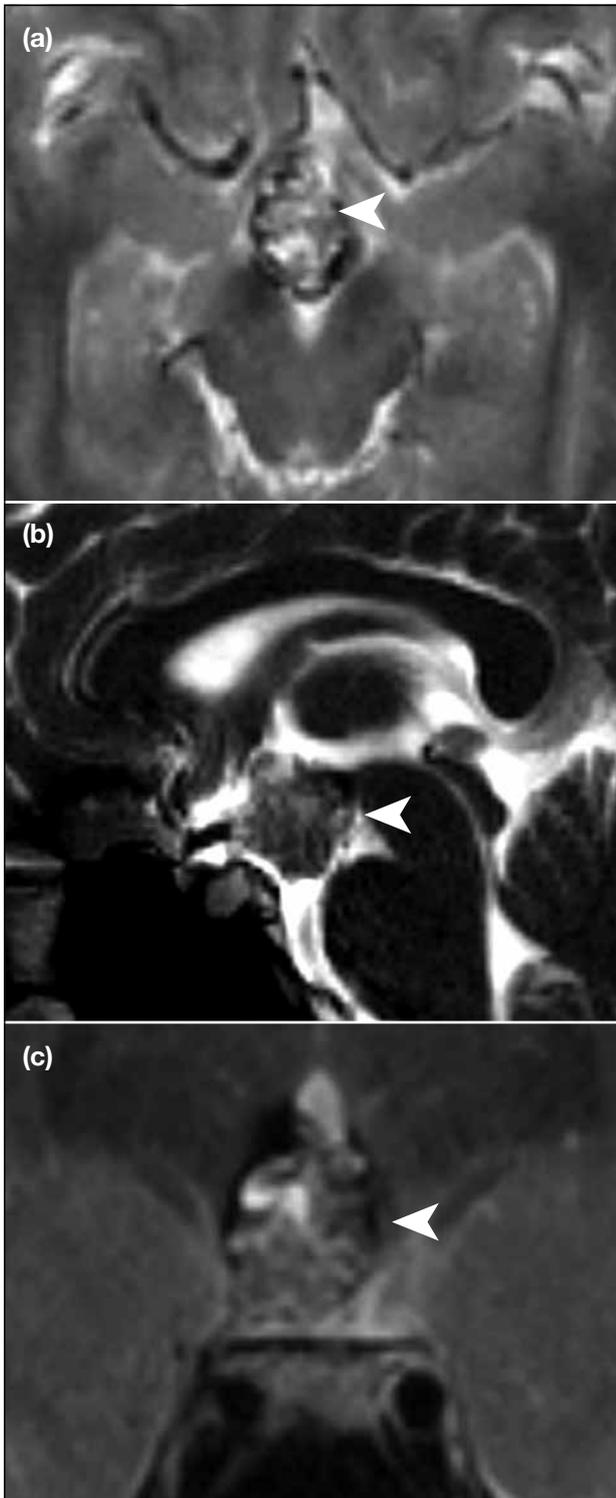
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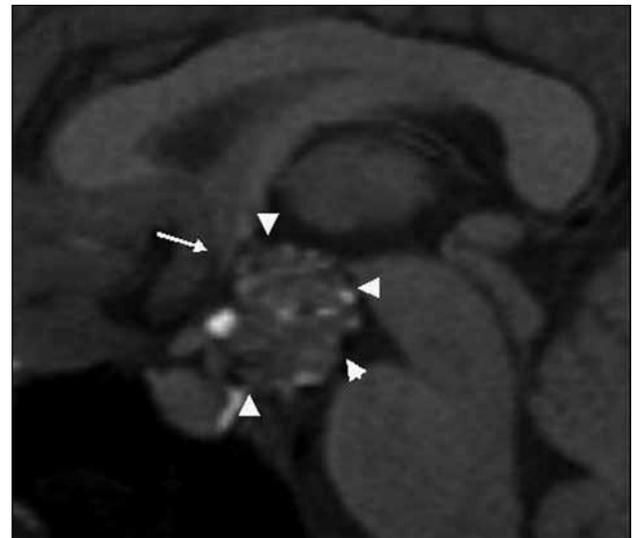
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Submitted: 18 Apr 2011; Accepted: 8 Jul 2011.



**Figure 1.** Magnetic resonance images of the hypothalamus obtained with a 0.35T scanner. (a) Axial spin-echo T2-weighted (TR: 3800 ms, TE: 90 ms), (b) sagittal T2-*Trufisp* (TR: 6.46 ms, TE: 3.05 ms), and (c) coronal spin-echo T2-weighted (TR: 3012 ms, TE: 119 ms) images all show well-defined lesions (arrowheads) with heterogenous signal intensity. Areas of haemorrhage are seen within the lesion (mulberry appearance) that are surrounded by a hypointense hemosiderin rim, which is in keeping with the diagnosis of cavernoma.



**Figure 2.** A sagittal spin-echo T1-weighted (TR: 330 ms, TE: 14 ms) magnetic resonance image obtained with a 0.35T scanner revealing the hypothalamus and suprasellar cistern showing the cavernoma (arrowheads) causing a mass effect and displacing the optic chiasm anteriorly and superiorly (white arrow).

on this characteristic ‘mulberry-like’ appearance, the diagnosis of hypothalamic cavernoma was made.

The ptosis subsequently resolved without any medical or surgical intervention. The patient was followed up with a planned repeat MRI should her symptoms recur.

## DISCUSSION

Intracranial cavernoma is a relatively rare benign condition.<sup>1-4</sup> Cavernomas are vascular malformations, which are thought to arise from failure of normal embryonic vascular development,<sup>5</sup> which give rise to a well-defined mulberry-like assembly of sinusoidal vascular channels lined by a single layer of endothelial cells. It contains collagen fibres in its walls but has neither muscle nor elastic fibres. The stroma shows fibroblastic proliferation without intervening neural tissue.<sup>2,4</sup> These findings distinguish cavernomas from other types of vascular malformations.<sup>1,3</sup>

The incidence of cavernomas in the brain is relatively low, overall they account for 5 to 13% of all central nervous system vascular anomalies. According to findings reported in several large autopsy series, they have a population prevalence of 0.02 to 0.9%.<sup>1,4</sup>

Cavernomas can arise anywhere in the brain, but are most frequently located in the frontal, temporal, and parietal lobes. Bernotas et al<sup>3</sup> reported that these three

sites accounted for 81% of all such lesions and that they were multiple in 13% of his patients. These anomalies are also found in the subcortical hemispheric area, deep cerebral white matter, basal ganglia, brain stem, and cerebellar hemispheres.<sup>6</sup>

Cavernoma in the hypothalamus is very rare.<sup>3,5</sup> To our knowledge, only about 10 cases have been reported.<sup>6</sup> Simard et al<sup>1</sup> reviewed 138 cases of cavernomas, only one of which occurred in the hypothalamus.

The clinical presentation of hypothalamic cavernoma is variable. Predominantly, patients have visual symptoms and headache. Kurokawa et al<sup>7</sup> reported a 27-year-old woman who presented with headache and a visual field defect. On neurological examination, she had an incomplete left homonymous hemianopsia, but hormonal assays yielded nil abnormal. Hasegawa et al<sup>8</sup> reported another case of a cavernoma and glioma in the hypothalamus. The patient presented with headache, visual disturbance and unsteady gait followed, and then had a generalised seizure. Physical examination revealed a right homonymous hemianopsia. Her serum prolactin level was elevated and she had anterior hypophyseal hypofunction, but levels of serum alpha-fetoprotein, carcinoembryonic antigen, and beta-human chorionic gonadotropin were all within normal limits. Mizutani et al<sup>9</sup> reported a 32-year-old patient who presented with progressive right visual loss with left frontoparietal headache. Examination revealed temporal hemianopsia of the right eye and a small inferotemporal scotoma of the left eye. Hormonal examinations were normal. Our patient presented with a focal oculomotor nerve deficit and no other nerve involvement.

Although virtually all cavernomas show signs of repeated microhaemorrhage, clinically significant haemorrhage is reported to be relatively uncommon.<sup>2,4</sup> However, in his review Simard et al<sup>1</sup> reported that one-third of the patients (40/138) presented with haemorrhage, eight of whom presented with recurrent haemorrhage. The risk factors for developing a clinically significant haemorrhage are a positive family history of intracranial cavernomas and patient's prior history of intracranial cavernoma haemorrhage. It has been found that pregnancy also increases the risk of haemorrhage, as the endothelial proliferation is related to the hormonal state of pregnancy.<sup>4</sup>

Other than its rare location, the radiological appearance of hypothalamic cavernomas is not different from

cavernomas of other parts of the brain. As in cavernomas at other sites, those that are hypothalamic are occult on angiography.<sup>9</sup> Perhaps this could be due to small, incompletely formed feeding vessels and slow flow, which causes dilution of contrast and / or thrombosis.<sup>1,2</sup> However, angiography may reveal hypovascular masses,<sup>2</sup> which may even cause lateral displacement of both anterior choroidal arteries, as reported by Mizutani et al.<sup>9</sup>

A plain computed tomography will demonstrate a heterogeneous mass that is slightly denser than the surrounding brain in the anterior region of the third ventricle and chiasmatic cistern.<sup>9</sup> It may appear hypodense or hyperdense, depending on the presence of cystic spaces, blood and calcium.<sup>1</sup> Calcification is frequently seen.<sup>2</sup> Most lesions show mild or no contrast enhancement although homogeneous or ring enhancement can occur.<sup>1,2</sup>

Using MRI, cavernomas appear isointense on T1WI and show mixed signals, and reticulated cores surrounded by hypointense rims on T2WI.<sup>6,9</sup> Poor contrast enhancement is a feature,<sup>7</sup> although Wang et al<sup>6</sup> reported a case of well-enhancing cavernoma in the hypothalamus. Their size varies, ranging from 0.3 to 5.0 cm as reported by Bernotas et al.<sup>3</sup> MRI is the investigation of choice as it can detect haemoglobin degradation products such as methaemoglobin, haemosiderin, and ferritin. It is highly sensitive and specific for these small angiographically occult lesions.<sup>4</sup> Cavernomas are characteristically small, and contain non-symptomatic haemorrhages that are typically confined to the lesion; only occasionally they become clinically significant.<sup>4</sup>

Furthermore, cavernomas are generally recognised as dynamic lesions, which grow prodigiously at one time and shrink considerably at others. Overall, they rarely remain quiescent,<sup>2,4</sup> the postulated reason being repeated microhaemorrhages followed by organisation, fibrosis, and calcification and / or recanalisation after intraluminal thrombosis.<sup>1,4</sup> We believe these features are also present in cavernomas of the hypothalamus, and could be the explanation for the ptosis that resolved spontaneously in our patient. Regrettably, we do not have a repeat MRI to confirm this possibility.

Although cavernomas are vascular malformations and not true neoplasms, they may mimic neoplasms clinically and radiologically. They may increase in size and even show enhancement in the post-

contrast scans.<sup>2,6</sup> The differential diagnosis of lesions in the hypothalamus in this age-group includes: germinoma, dermoid cyst, ganglioglioma, choristoma, hemangioblastoma, suprasellar aneurysm, and rarely metastasis.<sup>5</sup> All these lesions, except dermoid cyst, show some degree of enhancement. Germinomas may even show prominent contrast enhancement. In our patient however, no enhancement was noted in her MRI scan. Absence of fat signal intensity precluded the diagnosis of dermoid cyst.

The management of patients with cavernomas depends on their symptoms and their locations. An algorithm for management of cavernoma patients has been developed by the Department of Neurological Surgery at the Northwestern Memorial Hospital.<sup>4</sup> As cavernoma of the hypothalamus is a deeply seated lesion, it poses a challenge to the neurosurgeons. Kurokawa et al<sup>7</sup> recommended a “zygomatic approach” with the Neuronavigator system for accurate intraoperative anatomical evaluation so as to completely excise the lesion and avoid injury to the surrounding structures. The combination of functional MRI and diffusion-weighted imaging (DWI) with different directional sensitisations allows for assessment of functionally important cortical areas as well as additional visualisation and orientation of large fibre tracts.<sup>10</sup> This can further guide the surgical approach using frameless stereotaxy, and assist the neurosurgeon in avoiding interruption of pertinent penetrating fibres in the white matter.<sup>4,10</sup> In our patient, the managing team opted for conservative management.

## CONCLUSION

With the increasing availability of computed tomography and MRI machines, the incidental detection

of cavernomas has increased. MRI offers a more reliable means of diagnosing deeply seated, angiographically occult, hypothalamic cavernomas. In centres where facilities of DWI and frameless stereotactic navigation are available, MRI may further help to guide neurosurgical intervention for deeply seated cavernomas such as those in the hypothalamus.

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