



## Case Report

## Unusual etiology of bilateral acute visual impairment: Optochiasmatic cavernoma haemorrhage



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## ABSTRACT

Optochiasmatic cavernoma haemorrhage is unusual etiology of bilateral acute visual impairment. This vascular hamartoma is extremely rare with a prevalence rate of 0.4–0.9% of the general population. They are frequently revealed by an optochiasmatic apoplexy. We present the case of a 38 year old woman admitted to the emergency department for bilateral acute visual impairment, associated with frontal headaches. A brain MRI led to the diagnosis of an optochiasmatic cavernoma haemorrhage. Because of the rareness, and the lack of knowledge regarding the natural history of this lesion, surgical resection is the preferred management option in most reported cases. However after clear and detailed explanations of the surgical procedure as well as the risk of visual loss our patient refused to undergo any interventions. So our attitude was to carry out a close follow-up. After one year the visual evolution was satisfactory, no clinical worsening has been noticed and the MRIs examinations showed the same aspect with the same size. The aim of this observation is to evoke the diagnosis of an optochiasmatic cavernoma haemorrhage in case of acute chiasmal compression syndrome within patients in the third and fourth decades.

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## 1. Introduction

Cavernomas correspond to a vascular malformation with a prevalence rate of 0.4–0.9% of the general population [1]. Most cavernomas are supratentorial while the optochiasmatic localisation is very uncommon [1,2]. They are frequently revealed by an optochiasmatic apoplexy [2]. Magnetic Resonance Imaging (MRI) plays a pivotal role in the diagnosis of optochiasmatic cavernomas.

## 2. Case presentation

A 38 year old woman with no past medical history was admitted to the emergency department for bilateral acute visual impairment, associated with frontal headaches. The ophthalmologic examination of left and right eyes was notable for severe loss of visual acuity, with a score of 4/10, Parinaud 2 after correction. The anterior segment examination was unremarkable as well as the fundoscopic examination. Automated perimetry examination found incongruent right homonymous hemianopsia (Fig. 1). MRI scan of the brain and orbit showed an extra-axial suprasellar mass located in the optic chiasma. The mass was grossly oval, finely heterogeneous with a typical pepper and salt

aspect and a surrounding rim which appeared hypointense in the T2 and flair sequences but hyperintense in the T1 sequence. The mass contains extra and intra lesion stigmata of haemorrhage (Fig. 2). These findings led to the diagnosis of an optochiasmatic cavernoma haemorrhage.

After clear and detailed explanations of the surgical procedure as well as the risk of visual loss, the patient refused to undergo any interventions. So our attitude was to carry out a close follow-up with regular ophthalmological examinations and MRIs.

After one year of follow-up no clinical worsening has been noticed. Visual acuity moved to (10/10) in the right eye and (6/10) in the left eye. Automated perimetry examinations showed an improvement essentially in the left eye (Fig. 3). MRIs examinations found the same aspect with the same size.

## 3. Discussion

A cavernoma is a vascular hamartoma which corresponds to a low flow vascular lesion composed of multiple sinusoid-like interspersed capillaries without any functional parenchyma in histological studies. The malformation grows as a result of recurrent internal haemorrhage. Cavernomas represents 10–20% of all cerebrovascular malformations [1,3]. The supratentorial localization is the most common (80%) whereas; the cranial nerves and the optic pathways localizations are extremely rare ( $\leq 1\%$ ) [1,2].

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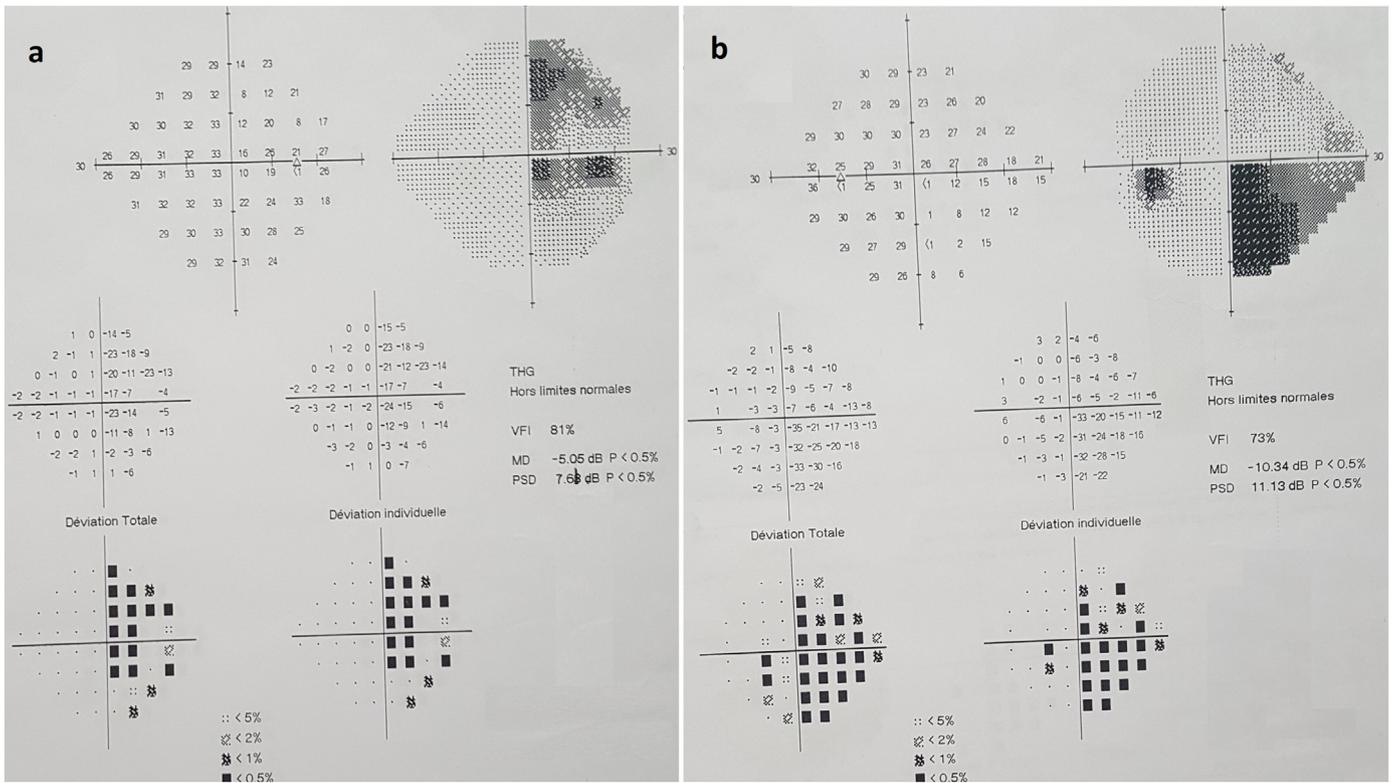


Fig. 1. Humphrey visual field; 24-2; standard white-on-white: showing incongruent right homonymous hemianopia. (a): Right eye, (b): left eye.

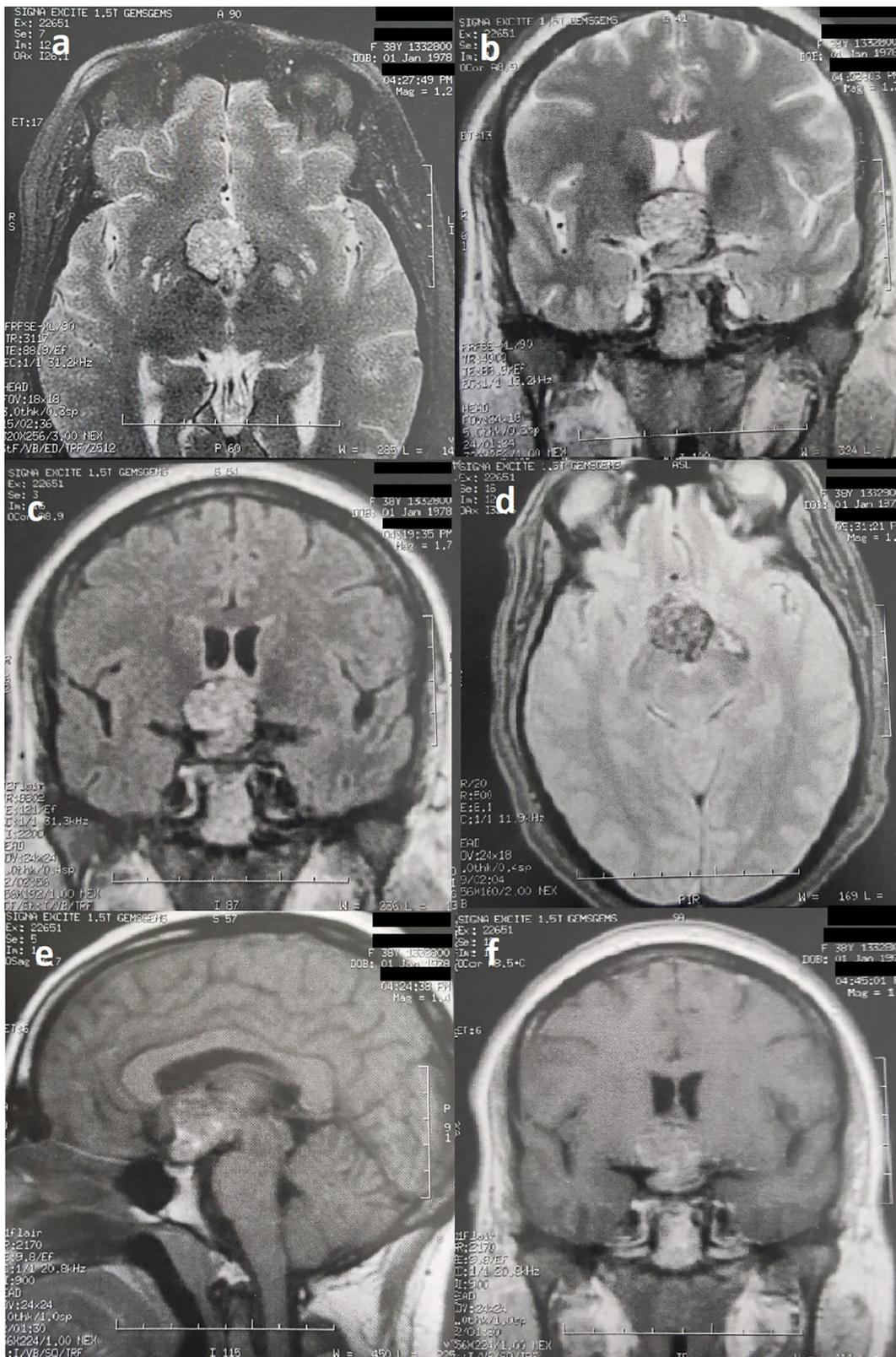
Optochiasmatic cavernomas become symptomatic essentially in the third and fourth decades of life and rarely at an earlier age [4,5]. It is frequently revealed by an optochiasmatic apoplexy corresponding to sudden visual deficiencies and often with an acute and frontal/retro-orbital headache which is the case of our patient [2].

MRI plays a pivotal role in the diagnosis of optochiasmatic cavernomas. Indeed this lesion has a specific presentation, it appears as a roundish lesion with heterogeneous signal in T1 and T2 sequences, surrounded by a thin hypointense hemosiderin rim in the T2 sequence, realizing a pop-corn or pepper and salt appearance which is typical of cavernomas [2,6,7]. Cavernomas present minimal or no enhancement after gadolinium injection [2].

Differential diagnosis of chiasmatic cavernomas includes optic gliomas, optic neuritis, arteriovenous malformation, intracranial aneurysm, metastasis and craniopharyngioma. However the clinical presentation, previous medical history, MRI findings and the clinical evolution can rule out these other possible diagnosis [7].

For our patient histopathological confirmation was lacking, nevertheless the typical clinical presentation, MRI findings and evolution were consistent with the diagnosis of cavernoma haemorrhage.

Because of the rare nature of optochiasmatic cavernomas and the lack of knowledge regarding the natural history of this lesion, surgical resection is the preferred management option in most reported cases [7]. For our patient we decided to carry out a close follow-up with



**Fig. 2.** A brain MRI showing an extra-axial suprasellar mass located in the optic chiasma. The mass is grossly oval, finely heterogeneous with a typical pepper and salt aspect and a surrounding rim which appeared in a hypointense in the T2 and flair sequences (a, b, c), but hyperintense in the T1 sequence (e, f). The mass contains extra and intra lesion stigmata of haemorrhage (d).

regular ophthalmologic examinations and MRIs imaging. The patient refused to undergo any procedures (surgical nor biopsy) due to fear of aggravation of her visual status. She judged her present visual acuity (10/10 right eye and 6/10 left eye) to be sufficient for her daily activities.

#### 4. Conclusion

Optic pathway cavernomas hemorrhages are very uncommon lesions, however we should think of this diagnosis in case of acute

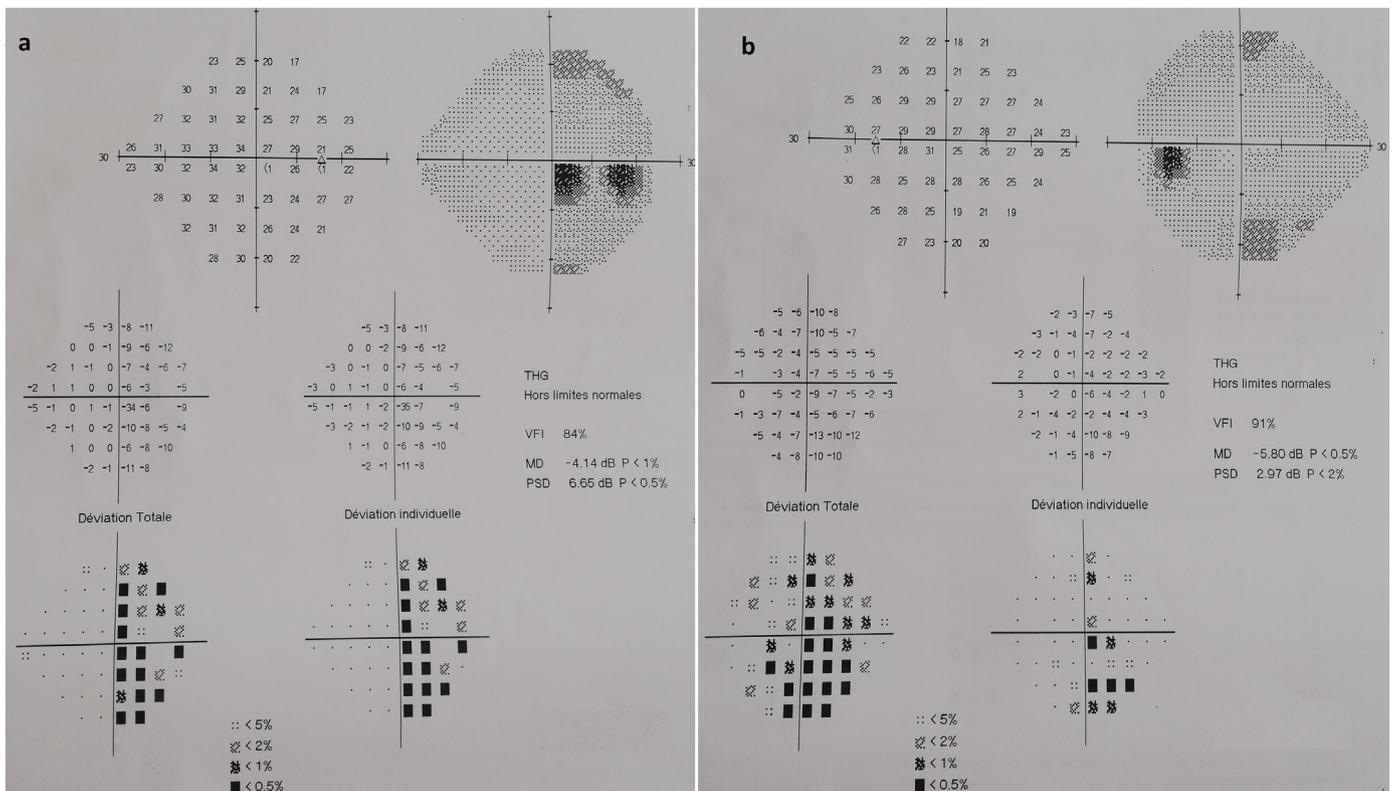


Fig. 3. Humphrey visual field; 24-2; standard white-on-white: after 12 month follow-up, we notice an improvement essentially in the left eye (b).

chiasmatic compression syndrome within patients in the third and fourth decades.

#### Conflict of interest

The authors declare having no conflict of interest.

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