A cavernous sinus meningioma in a monophtalmus: a diagnostic and therapeutic dilemma
Un méningiome de sinus caverneux chez un monophtalme : un dilemm diagnostique et thérapeutique

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Abstract:
Cavernous sinus meningioma is a rare tumor. The clinical manifestations are variable and require a particular clinical and paraclinical approach. The therapeutic decision remains to be adapted on a case-by-case basis, especially in the presence of certain limiting factors such as monophthalmia. The aim of this report was to highlight the diagnostic difficulties and management characteristics.

Keywords : meningioma, cavernous sinus, diagnosis, management

Introduction
Cavernous sinus tumors (1% of all intracranial tumors) constitute a particular entity, not only because of their relatively unequivocal mode of clinical revelation: the cavernous sinus syndrome; but also their own therapeutic difficulties. Tumors of the cavernous sinus itself are mainly represented by meningiomas (41%). We report the case of acute ptosis following paralysis of the common oculomotor nerve revealing a meningioma of the cavernous sinus.

Observation
We report the case of a woman aged 50 who consults for a slightly painful ptosis with rapid progressive onset, evolving for 20 days. The patient’s past medical history was: inconsistent headache for 1 year and paresthesia of the left hemiface evolving for a few weeks.

Exotropia of the right eye complicating deep amblyopia since childhood.

Clinical examination showed: (Figure1)

Right eye: visual acuity 20/250 (Snellen), permanent exotropia, the examination of the anterior segment is without abnormality, the lens is transparent and the fundus reveals papillary dysversion with a chorioretinal atrophy.

Left eye: visual acuity 20/40 (Snellen), moderate ptosis with frontal muscle hyper action, a limitation of the elevation, lowering and adduction of the eyeball confirming a lesion of
the III nerve, hypoesthesia in the territory V1 of the trigeminal nerve and afferent papillary deficit. Examination of the anterior segment is without abnormality, the fundus is strictly normal, in particular no papillary edema.

Papillary OCT did not reveal any RNFL loss or thickening, with a symmetrical average thickness: right eye 109 µ, left eye 111 µ.

Lancaster test showed; elevation, lowering and adduction paralysis confirming III nerve damage. (Figure 2)

Visual field (Goldman) of the left eye was normal. However, visual field of the right eye was not feasible. (Figure 3)

Cerebral MRI showed an expansive lesion of the left cavernous compartment, including the internal carotid in its entirety, reducing its caliber, but this remains permeable. This lesion extends towards the pre-sellar Turkish saddle, towards the optic canal, and towards the internal face of the temporal lobe. The appearance evokes a meningioma of the cavernous sinus. (Figure 4)
Discussion

Cavernous sinus meningioma (CSM) is the most common primary cavernous sinus (CS) lesion. Tumors located in the CS represent 1% of all intracranial neoplasms, but 41% of them are CSMs. Overall, the incidence of skull base meningioma is 2 per 100,000 persons/year. These lesions are most prevalent in females in their third or fourth decade of life.(1)

The cavernous sinuses are located on either side of the sellaire region. They communicate via the superior orbital fissure with the orbit, via the round foramen (and the inferior orbital fissure) with the pterygopalatine fossa and via the oval foramen with the infratemporal fossa and the masticatory space. The lower, external walls and the roof of the cavernous sinuses consist of dura mater. The medial wall is either non-existent or incomplete, represented by a thin sheet of collagen.

They contain vascular structures: the carotid, surrounded by the sympathetic fibers, venous plexus receiving the upper ophthalmic vein and cerebral veins (middle, lower and the sphenoparietal sinus), which drain, in the transverse sinus and the jugular gulf.

They also contain cranial nerves: III (oculomotor nerve), IV (trochlear nerve), branches V1 (ophthalmic) and V2 (maxilla) of the trigeminal nerve, in the lateral wall of the cavernous sinus and the VI (abducens nerve), along the lower and lateral side of the intra-cavernous portion of the carotid.(2)

Due to the relationship of the cavernous sinus with the surrounding structures, its involvement must be evoked in front of any ophthalmological symptomatology, associating various types of the following signs:

Oculomotricity disorders manifested by a binocular diplopia following the compression of one or more oculomotor nerves, concerning the IIrd, Vth and IVth nerves and can cause complete ophthalmoplegia (in 30% of cases).

A decrease in visual acuity is found in 40% of cases. It is due either to compression of the anterior intracranial visual pathways (optic nerve or chiasma) by a lesion with superior development, or to compression of the optic nerve at the orbital apex by a lesion with anterior development via the superior orbital fissure. (1,3)

Other signs can be seen:

exophthalmos by compression of the ophthalmic vein, paraesthesia and neuralgia due to trigeminal injury, retro-orbital pain and headache, disruption of the hypothalamic-pituitary axis, neurological deficit by compression of the brainstem or acoustical facial packages or convulsive attacks by temporal irritation in case of posterior or lateral extension of the lesion and exceptionally cerebral ischemia by compression of the internal carotid artery.

Therapeutic options are multiple including; simple observation, radiotherapy, surgery or a combination of both procedures. (4)

Cavernous sinus meningiomas are surgically difficult to resect due to their tendency to invade adjacent neurovascular structures and their proximity to vital neurological structures.(5)

Meningiomas of the cavernous sinus can be treated by fractional stereotaxic radiotherapy or by stereotaxic radiosurgery. In fact, stereotactic radiotherapy remains an important therapeutic tool, used as primary or adjuvant, in cases of symptomatic and asymptomatic cavernous sinus meningioma depending upon; their location, size, grade of tumor as well as age and morbid state of the patient. Indication is based on reported findings showing a tumor size control similar to microsurgery while avoiding neurovascular complications. (4) A recent meta-analysis by Sughrue et al., reporting 2065 cases, has demonstrated a superiority of radiotherapy compared to surgery regarding the preservation of cranial nerve function. (6) However, stereotactic radiotherapy has certain limiting factors ; large tumors, significant extra cavernous extension and close location to radiosensitive vital neural tissue.(7)

Fractional stereotaxic radiotherapy allows the irradiation of a reduced volume with standard fractionation. Dividing the dose reduces the risk of severe damage to healthy tissue (4% of complications) compared to radiosurgery, which delivers a single high dose (14 to 42% of complications). (8)

Conservative management is often dynamic, requiring close initial clinical and radiological follow-up, every 3-6 months to exclude aggressive variants requiring earlier surgical management, then follow-up can be carried out twice a year for the next year and annually thereafter to assess the rate of tumor growth.(9,10)

In our case, visual acuity of the right eye is quantifiable but remains very limited compared to the visual acuity of the left eye associated with a permanent exotropia, hence the patient was considered to be monophthalmic on the functional level. Thus, our therapeutic conduct was conservative. The decision was to treat the patient with fractional radiotherapy and then perform a second control MRI to ask for the surgical indication.

A medium-term follow-up, 1 year after radiotherapy, showed a slight shrinking; 15% of the initial tumor size. However, functional symptomatology and visual acuity remained stable.

Conclusion

Imaging examinations remain an essential means of diagnostic highlighting; tumors of various origins. Diagnostic and therapeutic approach requires good anatomical knowledge and systemic analysis. The therapeutic strategy remains to be adapted according to the clinical features, complications and post-therapeutic results.
Declarations of interest:
The authors have no conflict of interest to disclose.

References