OPTIC NERVE SHEATH MENINGIOMAS: CLINICAL FEATURES, FUNCTIONAL PROGNOSIS AND CONTROVERSIAL TREATMENT

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SUMMARY

Optic nerve sheath meningiomas (ONSM) are rare benign neoplastic lesions arising from meningothelial cells of the meninges.

As clinical features are highly variable, the diagnosis is often delayed.

From 1995 to 1999, 6 patients were diagnosed with ONSM in our department.

We compared our series with the literature data.

Visual prognosis is usually poor.

Despite a large literature, the treatment guidelines are still highly controversial.

KEYWORDS:

Atypical optic neuritis, MRI, optic nerve radiation therapy.

RÉSUMÉ

Le meningiome des gaines du nerf optique (MGNO) est une tumeur bénigne, rare,qui dérive des cellules meningothéliales de l'arachnoïde.

Les signes cliniques sont aspécifiques et le diagnostic est rarement précoce.

Nous avons collectés, de 1995 à 1999, 6 patients souffrant d'un MGNO.

Nous confrontons nos données aux données de la littérature.

Le pronostic visuel est généralement sombre: la vision est à plus ou moins long terme sévèrement réduite.

Malgré de nombreuses études, l'attitude thérapeutique est encore source de discussion.

MOTS CLÉS:

Névrite optique atypique,RMN,Radiothérapie du nerf optique.

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Table I: *Patients and ophthalmic signs at first examination*

PATIENTS	AGE (years)	SEX	INITIAL VA	PAIN	MOTIL DISTURB	PROPTOSIS	VISUAL FIELD	OPTIC DISC
1	68	M	10/10	+	-	-	paracentral scotoma	Optic disc edema
2	42	F	9/10	-	-	-	Enlargement blind spot	Optic disc edema
3	18	M	7/10	-	-	-	enlargement blind spot	temporal palor
4	40	F	10/10	+	-	-	paracentral scotoma	temporal palor
5	31	F	9/10	+	-	-	arciform scotoma	disc edema
6	37	F	7/10	-	-	-	central scotoma, altitud. defect	diffuse palor

INTRODUCTION

Optic nerve sheath meningioma (ONSM) is a benign tumor of the adult and may arise in the optic nerve sheath from meningothelial cells.
ONSM's account for 33% of all primary optic nerve tumors (4) and for 1,7% of all orbital tumors (14).
It is a rare disease with variable clinic presentations and the diagnosis is often delayed.
Despite a large literature, there is still much controversy regarding the natural history of this tumor and its appropriate management.
We reviewed our patients with ONSM, and we discuss our data reviewing the literature data.

PATIENTS AND METHODS

From 1995 to 1999,we collected 6 patients with an ONSM (mean age of 39 years old; 4 were female and 2 male).
All patients complained of some decreased vision, and visual acuity was mildly reduced in all patients (table I).
Slowly progressive loss of vision was usually reported, but 3 patients described a sudden onset of visual symptoms.
Peri- and retro-orbital pain was also noted by 3 patients.
Visual field defects were not specific but all were compatible with a compressive optic neuropathy (table I)
Chronic optic disc edema was observed in 3 patients and optic disc atrophy in the others.
Proptosis and oculomotor disturbances were absent in all patients.

Table II: *MRI diagnosis*

PATIENTS	DIAGNOSIS
	MRI
1	ONSM: orbital and intra- canalicular
2	ONSM: orbital and intracranial extension
3	ONSM: orbital
4	ONSM: orbital
5	ONSM: orbital (apex) and intracanalicular
6	ONSM: orbital and intracranial

Orbital M.R.I. (with gadolinium) allowed the diagnosis of ONSM in all patients.

M.R.I. showed a thickening of the optic nerve sheath complex, with an isointense or slightly hyperintense signal compared to the nerve in T1 and T2 weighted image, but with Gadolinium injection ONSM show marked enhancement of the tumor surrounding the hypointense optic nerve.The ONSM extended in the canal or intracranially in 4 patients (table II).
These MRI features allow to differentiate ONSM from Optic Nerve Glioma (ONG):M.R.I. show an enlargement of the optic nerve that is isointense relative to the gray matter in T1 weighted image.In T2, the tumor may show a variable signal intensity (homogenous or heterogeneous). After Gadolinium injection, ONG usually shows moderate enhancement.

FOLLOW-UP AND TREATMENT

Three patients had a progressive and severe visual loss over a period of 1 to 2 years, with an

Table III: *Follow-up of visual function*

PATIENTS	BEFORE TREATMENT		TREATM.	POST OP VA	LAST VA	FOLLOW UP
	FOLLOW UP	VA				
1	24 mo.	< 1/10	surgery	NLP	NLP	5 mo.
2	12 mo.	NLP	surgery			
6	12 mo.	6/10	surgery	6/10	< 1/10	24 mo.

intracranial extension in one of them. All had a tumorectomy without any visual improvement (table III).

Two other patients have been followed for at least three years without any visual deterioration. One had a very slowly progressive and severe visual loss, optic nerve sheath meningioma was strictly intracanalicular and surgery or radiation therapy have been declined by the patient.

DISCUSSION

Reviewing the literature since 1981, we observed that the clinical features were very similar to those reported in our series.

Patients were predominantly women (+/-61%) (4) with a mean age of 40 years. In 90% of the patients visual loss was one of the first symptoms. At the time of presentation, about 45% of the patients had a vision of more than 5/10 and a few of them complained of transient visual obscuration (2,4,14,15,16).
Visual field defects, as in our series were not specific but all suggestive of a compressive optic neuropathy.They were usually represented by a peripheral visual field constriction, central or paracentral scotomas, altitudinal defect and/or increased size of the blind spot (4,15).
Proptosis was usually very mild (median 2-5mm) and without a defined correlation with the visual acuity reduction (1,4,16).
Oculomotor disturbances were present in about 47% of the patients and most frequently related to mechanical restriction of the extraoculomotor muscles. Oculomotor nerve palsies were rare (5%)(4).
Orbital pain or generalized headaches were precisely reported only in few studies (+/-50%). Dutton, who reviewed the literature until 1992,observed that orbital pain seemed to be more frequently associated with an apical tumor. He suggested a possible compression of the sensory nerves (3).
Chronic disc swelling (50%) and optic disc atrophy (49%) have been reported as early findings (1,4,14,15).Optociliary shunt vessels, secondary to a retrobulbar compression of the optic nerve (1,14,15), appeared to be relatively infrequent (30%).However, their incidence might be higher because optociliary shunt vessels tend to appear some years after symptoms begin and might involute as optic atrophy is complete. In our series none of our patients presented optociliary shunt vessels.

EVOLUTION

From a histological point of view this tumor is benign and has a non-aggressive behavior for adult patients (2,14,16).
In our series, as in the literature, prognosis for life is excellent (15) but visual prognosis is usually poor.
In all studies, without treatment visual loss tends to increase in the majority of the patients. The evolution is usually unpredictable and the visual deterioration can be very slow or absent.
No significant relations between early visual decline and age (except for patients under 20 years), sex, tumor shape, optic disc aspect at the time of presentation and initial visual acuity were reported in the literature.Tumor location however seemed to have an important impact on the evolution of the visual function (14).In ONSM located at the apex of the orbit, the probability to conserve a good visual acuity is poor and the risk of intracranial extension higher (2).

THERAPY

The most appropriate therapy for the optic nerve sheath meningioma remains matter of controversy:

Table IV: *Ophthalmic signs reported by different authors*

AUTHORS	N° PTS	MEAN AGE	SEX	INITIAL V.A.	PAIN	MOTIL DISTURB.	PROPT OSIS	VISUAL FIELD	OPTIC DISC	DIAGNOSIS
Dutton, et al (1992) (4)	350	40 y	39%m	45% <=5/10	50%	47%	59%	35% periph. Constriction	49% O.D. atrophic	RX Skull
				31%: 4/10-CF				29% scotomas (central, para-, centrocecal)		CT SCAN
			61% F	24%: CF-NLP				16% altit. defect	48% O.D. swelling	MRI
								13% enlarg. B.S.		
Lindblom, et al (1992) (12)	13	40 y	8/13 f	...			–	...	9/13 O.D. swelling 11/13 O.D. atrophic	MRI ONSM (11/13 intra cranial ext.)
Polito, et al (1994) (14)	17	43.4 y	5 m	9 >=5/10		8	12	4 normal	10 O.D. atrophic	CT SCAN or MRI
				2: 4/10-1/20				4 periph. constriction		12 ONSM orbit
			12 f					3 scotomas	10 O.D. swelling	2 ONSM orbit + canal
				6<=CF				1 altit. defect	2 normal	3 ONSM orbit + intracranial extension
								1 enlarg. BS		

Table IV b: *Ophthalmic signs reported by different authors (continued)*

AUTHORS	N° PTS	AGE	SEX	INITIAL V.A.	PAIN	MOTILITY DISTURB.	PROPTOSIS	V.F.	OPTIC DISC
R DELFINI et al1996(2)	13	50	3m/10f	Reduced (76%)	rare	the majority	the majority	reduced	
LEE,et al 1996 (11)	1	43	female	10/10, asymptomatic	-	-	-	normal	swelling
KLINK, et al 1998(8)	1	40	female	8/10	-	-	-	enlarged blind spot, generalized depression	swelling

Table V: *Follow-up of patients without treatment*

OBSERVATION	N° PTS	FOLLOW-UP	MORTALITY	IMPROVED VISION	STABLE VISION	DECREASED VISION
Kennerdell, et al (1988)(7)	18	3-13 y.	-	0%	0%	100% after 5 y.
Dutton, et al (1992)(4)	64	-	0%	14%	86%	

Table VI: *Evolution after radiation therapy*

AUTHORS	RADIOTH.	N° PTS	F-UP	RECUR-RENCE	MORT-ALITY	IMPROVED VISION	STABLE VISION	DECREASED VISION
Itoi,et al (1988)(6)	Conven-tional	2	2 y.	-	-	100%	-	-
Kennerdell, et al (1988)(7)	Conven-tional	6	3-7 y.	-	-	100%	-	-
Dutton, et al (1992)(4)	Conven-tional	11	-	-	-	73%	9%	18%
Eng, et al (1991)(5)	3 -dim RT	3	-	-	,,,	,,,	-	
Lee, et al (1996)(11)	3-dim RT	1	4 y.	-	-	100%	-	-
Klink, et al (1998)(8)	Stereotactic R. surg.	1	2 y.	-	-	-	100%	-

Table VII: *Evolution after surgery*

AUTHORS	N° PTS	FOLLOW-UP	RECUR-RENCE	MORTALI-TY	IMPROVED VISION (N° PTS)	STABLE VISION (N° PTS)	DECREASED VISION (N° PTS)
Ito, et al (1988)(6)	5		,,,	-	1/5	3/5 (blind)	1/5
Kennerdell, et al (1988) (7)	10		+	-	if +, temporary	1/10 at 1 y.	4/10
Duton,et al (1992)(4)	148		25%	-	5%	1%	94%

Observation is generally recognized to be the most reasonable option in intraorbital meningioma with good function, because the slow and indolent growth pattern of ONSM (1,2,12) and the extremely low tumor-related mortality rate (11)(table V):

If visual deterioration progresses ***radiation therapy*** appears to be a promising and effective technique in slowing tumor progression and improving vision (3,5,10). But, because optic nerve sheath meningioma is a slowly growing tumor, high doses (+/- 5500-6000 cGy) have to be delivered (7) which have a high rate of visual and locoregional complications in all the studies: +/- 15% (11,13,14).

But recently, new radiation techniques have been applied with encouraging results in a few patients (table VI). Those new radiation modalities allowed an important reduction of the dose delivered to the surrounding structures but an increment of the dose delivered to the tumor.

Surgery has been for a long time the mainstay of therapy.

At present, because of the high rate of visual complications (30-40%) (1,2,15) such as central retinal artery occlusion, motility disturbances, visual field defects, phthisis, and the high rate of recurrences (65%)(9) surgery is reser-

ved for eyes without useful vision and for extraorbital spreading meningiomas.
Radiation therapy is usually recommended in case of partial excision to reduce the risk of recurrence (+/-25%).

CONCLUSION

Optic nerve sheath meningioma should be suspected in any patient presenting an usually progressive unilateral loss of vision, but can also be reported by some patients as an acute event. In the more recent literature as well as in our experience, peri- or retro-ocular pain appears a much more frequent and prominent symptom in ONSM than previously reported.
Unfortunately treatment is still a controversial issue, but new radiation techniques seem to be very promising for an effective and early treatment of ONSM. However, further multicentric studies are still necessary.

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