

ORBITAL MENINGIOMAS – CLINICAL OBSERVATION

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SUMMARY – The aim of the study was to assess the characteristics and clinical features of orbital meningiomas during a 5-year period (2001-2005). Due to their typical posterior location, slowly progressive and painless visual loss, orbital meningiomas are intriguing and challenging lesions to diagnose and manage. This prospective study identified five cases (four female, mean age 46.5 ± 14.5 years and one male aged 68 years) of orbital meningiomas, treated at University Department of Ophthalmology, Split University Hospital in Split from January 2001 until December 2005. In each case, the diagnosis was confirmed by history review. In our study, the incidence of orbital meningiomas was 0.2 *per* 100,000 inhabitants, however, some cases may have been recognized and treated at other hospitals in Croatia. In this study, orbital meningiomas showed a female predominance (F/M, 4:1). The mean visual acuity was 0.7 (range 0.4-0.9). Discrete proptosis was found in two cases with a slightly blurred optic disc margin and discrete optic disc edema. Optic nerve discoloration was observed in three cases. In one case, meningioma showed intracranial extension. Mild optic disc discoloration and blurred optic disc margin with slow and painless visual loss, particularly in females, may be the first signs of orbital meningioma.

Key words: *orbital meningioma, optic nerve, clinical observation*

Introduction

Orbital meningiomas account for 1%-2% of all meningiomas or one third of all optic nerve tumors with an incidence of approximately 2%^{1,2}. With the modern neuroimaging technique, the incidence of primary orbital meningiomas may increase. There is a female predominance for all types of meningiomas³. Most orbital meningiomas arise from intracranial extension³. A review by Dutton 90% showed that 90% of meningiomas arose from intracranial extension and 10% were from the orbit³. Meningiomas in the orbit are challenging lesions to manage because of their typical posterior location and slowly progressive painless visual loss^{3,4}. The majority of orbital meningiomas present with a slowly progressive optic neuropathy characterized by a variable loss of visual acuity^{3,4}. Ophthalmoscopic examination may reveal optic nerve head edema, pallor or choroidal folds.

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Received January 30, 2007, accepted March 26, 2007

The pathognomonic triade of slowly progressive visual loss, optic atrophy and optociliary shunt vessels is almost pathognomonic for orbital meningiomas, but tends to occur relatively late in the course of the disorder^{3,5,6}. External findings like proptosis, chemosis, lid edema and limitation of upgaze are relatively rarely present³⁻⁵. Orbit computed tomography (CT) or magnetic resonance imaging (MRI) allow for excellent visualization of orbital meningiomas. The management of orbital meningiomas has been controversial^{2,5}. Orbital meningiomas have been treated by excision of the tumor or without surgical intervention. Surgical treatment is a method of choice but very often is followed by complications such as the loss of visual acuity and blindness^{3,5,7-9}. There is a low to no mortality with orbital meningiomas and therefore observation is a reasonable option, however, accompanied by further visual acuity decline^{3,10}. Fractionated radiation therapy has recently yielded promising results^{10,11}.

The aim of the study was to assess the incidence, characteristics and clinical features of orbital meningiomas during a 5-year period (2001-2005).

Material and Methods

This study was retrospectively performed in five patients (four female, mean age 46.5 ± 14.5 years, and one male aged 68 years) with orbital meningiomas, treated at University Department of Ophthalmology, Split University Hospital in Split from January 2001 until December 2005. In each case, the diagnosis was confirmed by history review. The diagnosis is based on appropriate clinical findings and neuroimaging of the orbit and brain. Each patient underwent routine ophthalmologic examinations including determination of visual acuity with Snellen charts, ophthalmoscopic fundus examination, measurement of intraocular pressure with applanation tonometry, and biomicroscopy of the posterior eye segment with Goldmann lens. Because of inadequately clear clinical picture, relatively preserved visual acuity and unexplainable discoloration of the optic nerve disc, the following studies were additionally performed: Goldmann kinetic perimetry, visual evoked potentials, ultrasonography of the eye and orbit, x-ray of the paranasal sinus cavity, CT or MRI of the brain and

orbit, Hertel exophthalmometry, color vision testing with Ishihara pseudo-isochromatic plates, and laboratory workup (complete blood count, erythrocyte sedimentation rate and routine biochemistry).

Results

In our study, the incidence of orbital meningiomas was 0.2 *per* 100,000 inhabitants, yet considering that some cases may have been recognized and treated at other hospitals in Croatia. In our patient series, there was a female predominance for orbital meningiomas (male to female ratio 4:1). The mean visual acuity was 0.7 (range 0.4-0.9). Discrete proptosis was found in two cases with a slightly blurred optic disc margin and discrete optic disc edema. Optic nerve discoloration was found in another two cases. In only one case, the meningioma showed intracranial extension. The cases are presented in Table 1. The first case was a woman aged 68 with the diagnosis ischemic optic neuropathy of the right eye, visual acuity 0.8 and optic nerve discoloration. CT of the orbit showed intraorbital tumor formation with

Table 1. Clinical characteristics of study patients

Case/sex/age	Diagnosis at admission	Fundus examination	Proptosis	Orbit CT or MRI	VEP	Visual field (Goldmann)
1/F/68	Right optic neuropathy	Optic nerve discoloration	No	Intraorbital optic nerve tumor	Prolonged latency of P100 wave	Altitudinal inferior defect
2/M/68	Suspected tumor of right optic nerve	Optic nerve discoloration	No	Intraorbital optic nerve thickening	Normal latency of P-100 wave	Moderate constriction
3/F/42	Left eye proptosis	Slightly blurred optic disc margin and discrete optic disc edema with discoloration	Yes	Presumed left optic nerve meningioma with intracranial extension	No	No
4/F/36	Suspected left optic neuritis	Slight optic nerve discoloration	No	Intraorbital tumor of optic nerve extending into optic canal	Prolonged latency of P100 wave	Bjerrum scotoma, moderate constriction
5/F/40	Right eye proptosis	Slightly blurred optic disc margin and discrete optic disc edema	Yes	Intraorbital thickening in front of optic nerve canal	Prolonged latency of P100 wave	Marked constriction

CT=computed tomography; MRI=magnetic resonance imaging; VEP=visual evoked potential

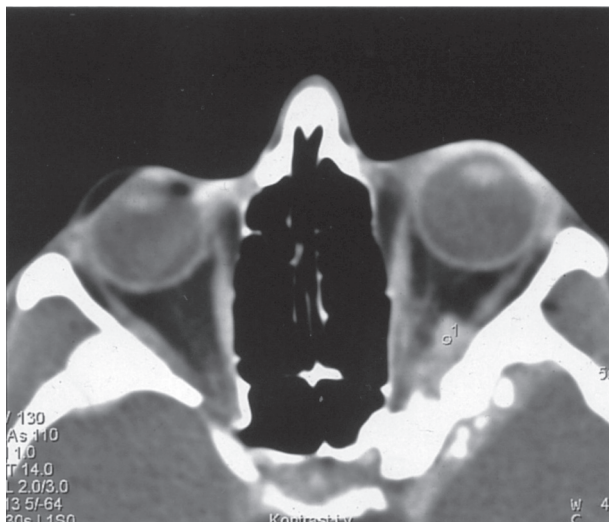


Fig. 1. Orbit computed tomography: suspected left optic nerve meningioma with intracranial extension.

propagation into the optic canal. The second case was a man aged 68 with suspected tumor of the right optic nerve and visual acuity of 0.9. Fundus examination revealed optic nerve discoloration. CT of the orbit showed optic nerve thickening. The third case was a woman aged 42 diagnosed with mild proptosis of the left eye due to unknown cause (18 mm of the left eye and 12 mm on the right eye with Hertel exophthalmometry), and with visual acuity of 0.8. Fundus examination showed slightly blurred optic disc margin and discoloration. CT of the



Fig. 2. Orbit computed tomography: left optic nerve meningioma with intracranial extension after surgery.

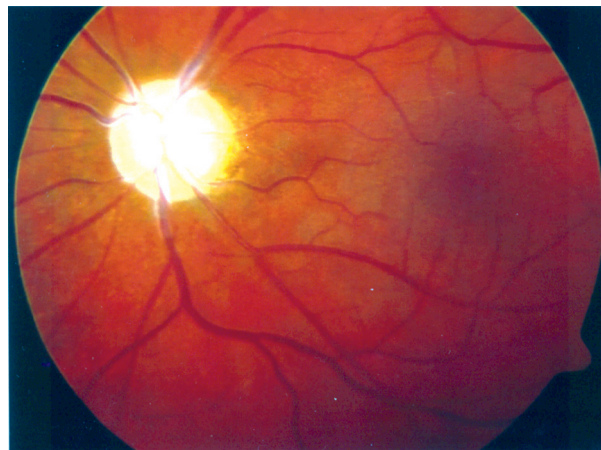


Fig. 3. Left optic disc atrophy following surgical intervention for orbital meningioma with intracranial extension.

orbit showed posteriorly located meningioma with intracranial extension (Fig. 1). The fourth case was a woman aged 36 hospitalized for suspected optic neuritis of the left eye, with visual acuity of 0.7. Ophthalmoscopic examination of the optic nerve head was unremarkable. MRI of the brain showed tumor formation in the optic nerve canal. The fifth case was a woman aged 40 diagnosed with diplopia and proptosis of the right eye, with visual acuity of 0.4. Fundus examination showed slightly blurred optic disc margin and discrete optic disc edema. Hertel exophthalmometry showed 21 mm on the right eye and 18 mm on the left eye. CT of the orbit and brain showed thickening of the optic nerve in front of the optic nerve canal.

Discussion and Conclusion

The diagnosis of orbital meningioma can be suspected from clinical findings and supported by results of orbit neuroimaging, in most cases with progressive visual loss, swelling of the optic disc or optic atrophy^{3,4}. Compared to other studies, our experience in the early detection of orbital meningiomas in our patients seems rather scarce; however, they have been recorded and noted at our hospital. The fact that it is atypical and unobtrusive as well as scarce in symptoms and signs in the early phase makes the clinical picture of orbital meningiomas very difficult to make. Therefore, we must thus bear in mind that besides some other similar eye pathology, a mild loss of visual acuity without a noticeable reason, with slight changes in the optic nerve head such as discoloration may be the first signs of existing

orbital meningiomas. In these patients, additional diagnostic testing, including CT or MRI of the orbit, should be considered. It is not always possible to differentiate orbital meningiomas from other lesions involving optic nerve, such as sarcoidosis, demyelinating optic neuritis or orbital inflammatory disease. We believe that a thorough, basic ophthalmologic examination and properly taken history are of utmost importance in case of suspected orbital meningiomas, in young people in particular. According to literature data, the treatment is rather controversial^{2-5,7,8}. Namely, surgical treatment for removal of a meningioma located in the posterior orbit/optic canal, which extends intracranially, seems as a logical solution (Fig. 2; case 3); however, results are often disappointing for the patient, as it may result in optic nerve atrophy and blindness (Fig. 3; case 3).

The natural history of orbital meningiomas is slow loss of visual acuity that progresses in most patients over years; also, these tumors do not metastasize¹². Therefore, observation is a reasonable option in patients with mild or no visual deficit¹³. Fractionated radiation therapy is promising and acceptable, however, with obligatory monitoring and observation of such patients^{11,13}. We do not restrain to conclude that, in the world of modern ophthalmology, orbital meningiomas may pose a diagnostic problem in the early stage of the disease.

In conclusion, slight optic disc discoloration and discretely blurred optic disc margin with slow and painless visual loss in younger persons, female in particular, may be the first sign of orbital meningiomas.

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Sažetak

MENINGEOMI ORBITE – KLINIČKO ZAPAŽANJE

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Meningeomi orbite zbog svoje stražnje lokalizacije i polaganog bezbolnog gubitka vida često predstavljaju dijagnostički i terapijski izazov. Cilj rada bio je ispitati incidenciju meningeoma orbite tijekom 5 godina (siječanj 2001. – prosinac 2005.) na Klinici za očne bolesti Kliničke bolnice Split. Retrospektivnom analizom su ispitane povijesti bolesti petoro bolesnika s kliničkom dijagnozom meningeoma orbita liječenih na našoj Klinici. Analiza je obuhvatila četiri žene srednje dobi $46,5 \pm 14,5$ godina i jednog muškarca starog 68 godina. Incidencija meningeoma orbite u našem ispitivanju bila je 0,2 na 100.000 stanovnika, iako je moguće da su neki bolesnici dijagnosticirani i liječeni na drugim klinikama u Hrvatskoj. Meningeomi orbita su bili češći kod žena negou kod muškaraca (4:1). Srednja vidna oštrina je bila 0,7 (raspon 0,4-0,9). Diskretna proptoza s nejasnim granicama i blagim edemom glave vidnog živca nađena je u dva slučaja, a diskoloracija vidnog živca u druga tri slučaja. Kod jednog bolesnika meningeom orbite je očitovao intrakranijsku ekstenziju. Blagi i bezbolni pad oštrine vida jednoga oka, diskoloracija i nejasne granice vidnog živca na očnoj pozadini, naročito kod žena, mogu biti prvi znaci meningeoma orbite.

Ključne riječi: *meningeomi orbite, vidni živac, kliničko zapažanje*



