

Visual findings as primary manifestations in patients with intracranial tumors

Nazife Sefi-Yurdakul

Department of Ophthalmology, Izmir Atatürk Education and Research Hospital, Izmir 35360, Turkey

Correspondence to: Nazife Sefi-Yurdakul. Halk Sokak, No: 26 Sahilevleri, Narlidere-Izmir 35320, Turkey. nsefi@yahoo.com

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Abstract

• **AIM:** To evaluate the visual findings as primary manifestations in patients with intracranial tumors.

• **METHODS:** The medical charts of the patients with intracranial tumors who initially admitted to the Neuro-ophthalmology and Strabismus Department with ocular complaints between August 1999 and December 2012 were reviewed retrospectively. The detailed clinical history and the findings of neuro-ophthalmologic examination were recorded. Ocular symptoms and signs, the types and locations of intracranial tumors, and the duration of symptoms before the diagnosis were evaluated.

• **RESULTS:** The mean age of 11 women (61.1%) and 7 men (38.9%) was 42.2 ± 11.0 (range 20–66y) at the time of intracranial tumor diagnosis. Initial symptoms were transient visual obscurations, visual loss or visual field defect in 16 cases (88.9%), and diplopia in 2 cases (11.1%). Neuro-ophthalmologic examination revealed normal optic discs in both eyes of 6 patients (33.3%), paleness, atrophy or edema of optic disc in 12 patients (66.7%), and sixth cranial nerve palsy in 2 patients (11.1%). Visual acuity ranged between normal vision and loss of light perception. Cranial imaging demonstrated craniopharyngioma ($n=1$), plasmacytoma ($n=1$), meningioma ($n=6$; olfactory groove and tuberculum sellae, pontocerebellar angle, anterior cranial fossa, frontal vertex, suprasellar region), and pituitary macroadenoma ($n=10$). The mean duration between the onset of visual disturbances and the diagnosis of intracranial tumor was 9.8 ± 18 mo (range 3d–6y).

• **CONCLUSION:** The ophthalmologist is frequently the first physician to encounter a patient with clinical manifestations of intracranial tumors that may cause neurological and ocular complications. Neuro-ophthalmologic findings should be carefully evaluated to avoid a delay in the diagnosis of intracranial tumors.

• **KEYWORDS:** intracranial tumors; neuro-ophthalmologic examination; optic neuropathy

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INTRODUCTION

Intracranial tumors may cause serious ocular signs and symptoms in addition to neurological complications due to increased intracranial pressure, cranial nerve impairment or brain compression. Therefore, prompt diagnosis of intracranial tumors can allow early treatment and avoidance of complications. Usually, the diagnosis of an intracranial tumor can only be established in the presence of usual symptoms or signs and sometimes these may lead to misinterpretation and misdiagnosis^[1-3]. Typical initial ocular symptoms are progressive loss of vision with or without optic nerve atrophy, visual field defects, and extraocular nerve palsies^[4-7]. Instead of progressive visual defects, occasionally, some of these lesions may cause acute visual loss as the presenting manifestation of intracranial tumors^[5,8-10].

In this study, we aimed to draw attention to careful evaluation of the findings in patients with intracranial tumors who were referred to the Neuro-ophthalmology and Strabismus Department with ocular complaints.

SUBJECTS AND METHODS

Subjects We retrospectively reviewed the medical charts of patients who were referred to the Neuro-ophthalmology and Strabismus Department of our hospital between August 1999 and December 2012 with ocular symptoms, and those who were subsequently diagnosed with intracranial tumors in the Neurology Department were included in the present study. Written informed consent was obtained from the patients in accordance with the Declaration of Helsinki. The study used routine data, which were collected in a standard manner on all cases.

Methods The diagnosis of intracranial tumors had been made on the basis of characteristic clinical and radiological findings. The detailed clinical history and the findings of a full ophthalmologic and neuro-ophthalmologic examination, including visual acuity, visual field, pupillary reactions, color vision, ocular motility, biomicroscopy of anterior segment, intraocular pressure measurement, and dilated fundus

examinations, were recorded. Additionally, the initial symptoms, signs prior to diagnosis of the tumor, and neurological, biochemical and hematological findings at diagnosis were recorded. Ocular manifestations, the types and locations of intracranial tumors were evaluated. The estimated duration of symptoms described by the patients was investigated.

RESULTS

This study included 18 patients of whom 61.1% ($n=11$) were woman and 38.9% ($n=7$) were man. The mean age at presentation was 42.2 ± 11.0 (range 20-66y). Decreased visual acuity, transient visual obscurations or visual field defect were the most common presenting symptoms ($n=16$, 88.9%), followed by diplopia ($n=2$, 11.1%). Eleven patients (61.1%) complained of progressive blurred vision. The remaining patients presented with acute visual deterioration for a period of less than one month. Visual acuity ranged between normal vision and loss of light perception. Visual field pattern differed from normal visual field to total loss of visual field. Incomplete right homonymous hemianopsia and enlargement of blind spot are shown in Figure 1 (case 18). The mean duration between the onset of visual disturbances and the diagnosis of intracranial tumor was 9.8 ± 18 mo (range 3d-6y). Dilated fundus examination showed normal optic discs in both eyes of 6 patients (33.3%), and paleness, atrophy or edema of optic disc in 12 patients (66.7%). Bilateral optic disc edema is disclosed in Figure 2 (case 18). Sixth nerve palsy was detected in 2 patients (11.1%). Non-specific headache was described by 22.2% of the patients. All extensive laboratory tests were within normal limits.

Before radiological investigations, one case (case 1) had pre-diagnosis of toxic optic neuropathy, one case (case 17) Leber's hereditary optic neuropathy, one case (case 6) ischemic optic neuropathy, and 2 cases (cases 3, 15) retrobulbar neuritis. After neuro-ophthalmologic examination, the patients were consulted by a neurologist. The definitive diagnosis of an intracranial tumor was made based on the characteristic clinical and radiological findings. Neurological examination and cranial imaging demonstrated craniopharyngioma ($n=1$), plasmacytoma ($n=1$), meningioma ($n=6$), and pituitary macroadenoma ($n=10$). The locations of the meningiomas were olfactory groove and tuberculum sellae ($n=1$), pontocerebellar angle ($n=1$), anterior cranial fossa ($n=1$), frontal vertex ($n=1$), and suprasellar region ($n=2$). A large anterior cranial fossa meningioma is demonstrated in Figure 3 (case 1). The size of the tumors ranged between 1 and 7 cm. Descriptive features of the patients are presented in Table 1.

DISCUSSION

Intracranial tumors generally cause progressive visual deficits and visual field loss (up to 95%) over weeks to months before the diagnosis^[11-13]. Initial symptoms and signs are often

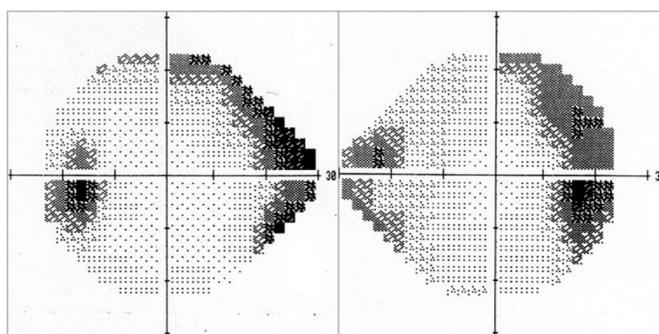


Figure 1 Incomplete homonymous hemianopsia and enlargement of blind spot (case 18).

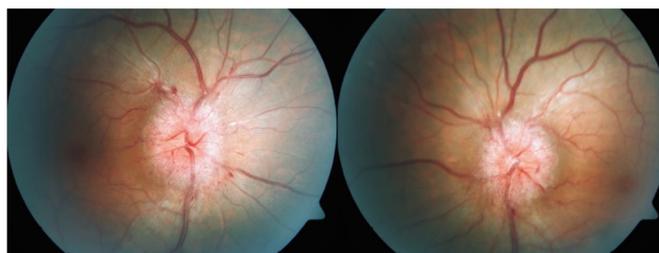


Figure 2 Bilateral optic disc edema (case 18).

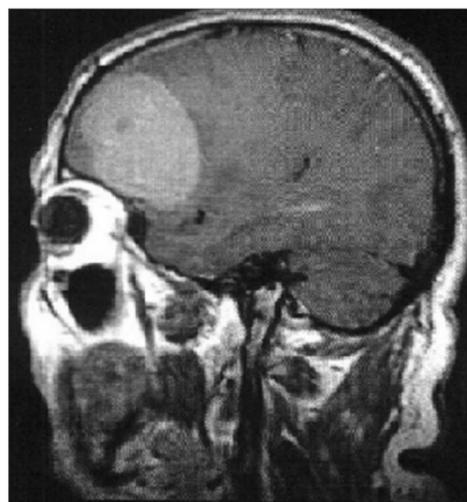


Figure 3 Anterior cranial fossa meningioma (case 1).

misinterpreted both by the patients and physicians, and appropriate investigations are consequently delayed. The duration of symptoms may be as long as 13y before the diagnosis^[3-5,14]. In the present study, the estimated duration of symptoms described by the patients before the diagnosis ranged from 3d to 6y, with a mean of 9.8mo.

The three most common types of intracranial tumors in adults are pituitary adenoma, meningioma, and craniopharyngioma, in decreasing order of frequency^[4-6,12]. Pituitary adenoma frequently manifests with bitemporal hemianopia, but is usually asymmetrical and unpredictable in its evolution^[6]. Clinical presentation is related with anatomic location of the chiasm. Cranial nerve paralysis and diplopia can develop because of parasellar extension of the tumor^[15-17]. In this study, neuro-ophthalmologic examination and cranial imaging demonstrated pituitary macroadenomas in 10 patients; all cases, except one with diplopia (case 14), were admitted due

Table 1 Descriptive features of the patients with intracranial tumors who were initially admitted to ophthalmology outpatient clinic with ocular symptoms

Case No.	Age (a)	Sex	Visual acuity	Visual field defects	Other findings	Optic disc	Diagnosis (localization)	Duration (D)
1	55	M	RE: P- LE: P- RE: P+	RE: Not done LE: Not done	Headache Anosmia	RE: Edema LE: Edema	Meningioma (anterior cranial fossa)	3d
2	66	F	RE: 20/50 LE: 20/20	RE: Hemianopsia	Cortical cataract	RE: Atrophy LE: Atrophy	Meningioma (suprasellar)	6y
3	26	F	RE: 20/20 LE: 1m CF	RE: Normal LE: Not done	Headache	RE: Normal LE: Normal	Meningioma (suprasellar)	7d
4	37	F	RE: 20/20 LE: 20/20	RE: BSE LE: BSE	Transient visual obscuration	RE: Edema LE: Edema	Meningioma (pontocerebellar angle)	3mo
5	44	F	RE: 20/20 LE: 20/20	RE: BSE LE: BSE	Transient visual obscurations (right eye)	RE: Edema LE: Edema	Meningioma (frontal vertex)	20d
6	51	F	RE: 20/20 LE: 2m CF	RE: Normal LE: Total loss	-	RE: Normal LE: Pale	Meningioma (olfactory groove-tuberculum sellae)	3y
7	46	F	RE: 20/30 LE: 20/200	RE: Hemianopsia LE: Hemianopsia	-	RE: Normal LE: Normal	Pituitary macroadenoma	1y
8	43	M	RE: 20/200 LE: 20/200	RE: Hemianopsia LE: Hemianopsia	-	RE: Atrophy LE: Atrophy	Pituitary macroadenoma	1y
9	50	M	RE: HM LE: 3m CF	RE: Not done LE: Diffuse loss	-	RE: Atrophy LE: Atrophy	Pituitary macroadenoma	1y
10	41	F	RE: 20/30 LE: 20/40	RE: Hemianopsia LE: Hemianopsia	-	RE: Normal LE: Normal	Pituitary macroadenoma	9mo
11	44	M	RE: 20/40 LE: 20/20	RE: Total loss LE: Hemianopsia	-	RE: Atrophy LE: Edema	Pituitary macroadenoma	2mo
12	42	F	RE: 2m CF LE: 20/20	RE: Concentric narrowing LE: Normal	-	RE: Pale LE: Normal	Pituitary macroadenoma	30d
13	51	F	RE: 20/30 LE: 20/30	RE: Hemianopsia LE: Hemianopsia	Transient visual obscuration	RE: Normal LE: Normal	Pituitary macroadenoma	1y
14	26	F	RE: 20/20 LE: 20/20	RE: Normal LE: Normal	Diplopia	RE: Normal LE: Normal	Pituitary macroadenoma	15d
15	36	F	RE: 20/20 LE: 20/25	RE: Normal LE: Normal	-	RE: Normal LE: Normal	Pituitary macroadenoma	20d
16	42	M	RE: 20/40 LE: 20/20	RE: Diffuse loss LE: Diffuse loss	-	RE: Edema LE: Pale	Pituitary macroadenoma	10d
17	20	M	RE: P+ LE: 3m CF	RE: Diffuse loss LE: Diffuse loss	-	RE: Atrophy LE: Pale	Craniopharyngioma (suprasellar)	15-30d
18	39	M	RE: 20/20 LE: 20/20	RE: Hemianopsia LE: Hemianopsia	Diplopia Headache	RE: Edema LE: Edema	Plasmacytoma (posterior fossa)	30d

RE: Right eye; LE: Left eye; P: Light perception; HM: Hand motions; CF: Count fingers; BSE: Blind spot enlargement; D: Time from onset of symptoms to diagnosis.

to visual or visual field defects. No patient primarily presented with endocrinologic symptoms. Before confirmation of the diagnosis with cranial imaging, one patient (case 15) was pre-diagnosed as retrobulbar neuritis. Besides the cases with pituitary macroadenoma, three cases with meningioma were pre-diagnosed as toxic optic neuropathy (case 1), retrobulbar neuritis (case 3), and ischemic optic neuropathy (case 6). The most dramatic case (case 1) among our patients had anterior cranial fossa meningioma, and he developed bilateral visual loss in three days. This patient described nonspecific headache and anosmia from time to time. He was initially referred to our clinic and pre-diagnosed as toxic optic neuropathy, and then eventually a large olfactory groove meningioma, 5.5 × 6.7 × 8.5-cm³ in size, invading the anterior cranial fossa was determined. Olfactory groove meningiomas most commonly present with symptoms of headache, anosmia, and personality changes [18,19]. Furthermore, they can reach to a

huge size without showing any symptom and be easily confused with toxic optic neuropathy, especially in patients who present with bilateral vision loss and visual field defects. Jung *et al* [18] reported a case with bilateral visual loss for over 2y due to a giant olfactory meningioma encompassing the entire frontal lobe and compressing the optic nerves. Bouyon *et al* [3] reported 5 patients with an initial presentation of retrobulbar optic neuropathy and in radiological imaging, that had to be repeated to make the diagnosis of meningioma, which was noted to be delayed from 18mo to 4y. Puchner *et al* [20] suggested that the commonly very late diagnosis of meningiomas as a cause of visual loss may be attributed to the low incidence of the tumor. Although benign in nature, craniopharyngiomas can contribute to significant morbidity. Karavitaki *et al* [21] and Overly [22] found visual field defects and an initial decrease in visual acuity in their patients with suprasellar craniopharyngioma. Chen *et al* [23] reported that change from

one type of field defect to another is one of the most typical signs of craniopharyngioma. In our craniopharyngioma case (case 17), tumor was located in the suprasellar region. He presented with severe vision loss in both eyes over the course of between 15 and 30d. The initial diagnosis of Leber's hereditary optic neuropathy was made. Cranial magnetic resonance imaging revealed the exact diagnosis of craniopharyngioma.

Ophthalmic signs as the initial manifestations of solitary intracranial plasmacytoma have been rarely described. Brannan *et al* [24] reported two patients with solitary plasmacytomas. One patient presented with optic neuropathy, the second with bilateral sixth nerve palsy. Similarly, a 39-year old man presented to our clinic with the complaint of diplopia and non-specific headache for about a month. After neuro-ophthalmologic examination, papilledema and right homonymous hemianopsia were identified and he was referred to the neurology clinic. Apparently, he had posterior fossa plasmacytoma. Plasmacytoma is a treatable intracranial tumor that should be considered in the differential diagnosis of patients who present with optic neuropathy or sixth nerve palsy.

In conclusion, the ophthalmologist may be the first physician to encounter a patient with clinical manifestations of intracranial tumors that may cause neurologic and ocular complications. We suggest that the possibility of intracranial tumors should be considered in the etiology of visual disturbances before reaching a definitive diagnosis. Neuro-ophthalmologic features should be carefully investigated to avoid a delay in the diagnosis of intracranial tumors.

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Conflicts of Interest: Sefi-Yurdakul N, None.

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