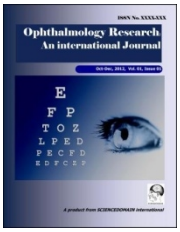

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Visual Pathway Tumor Presenting as Visual Disturbances without Extraocular Signs

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

Original Research Article

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ABSTRACT

Purpose: To evaluate visual pathway tumor presenting as visual disturbances without extraocular signs and assess the usefulness of various examinations.
Methods: Only 35 patients with intracranial tumors (22 males and 13 females) who were initially diagnosed at our hospital and confirmed by magnetic resonance imaging were enrolled, and previously known intracranial tumor patients and patients with strabismus or proptosis were excluded. Best corrected visual acuity (BCVA), pupillary reflex test, fundus examination, color vision test and visual field test were done to evaluate visual impairments. We investigated the incidence of intracranial tumors and sensitivity of tests.
Results: The most common tumors were pituitary tumors (60.0%), followed by meningioma (20.0%) and optic nerve glioma (5.7%). The BCVA ranged from 1.0 to no light perception and the visual acuity of 3 patients was 1.0 in both eyes. Positive relative afferent pupillary defect was seen in 71.4% and abnormal disc findings were found in 58.8% of patients. The color vision test was more specific (sensitivity: 76.6%), and all patients had abnormal visual field defects.
Conclusions: Pituitary tumor is the most common intracranial tumor. Among tests, a visual field test is a more sensitive test than other tests for detecting compressive optic neuropathy.

Keywords: Tumor; compressive optic neuropathy; visual field test; visual pathway; pituitary tumor.

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1. INTRODUCTION

Compressive optic neuropathy can result from numerous pathologies such as thyroid associated orbitopathy, intraorbital inflammation, and orbital and intracranial tumors. Among these, some intracranial tumors may be fatal, and therefore ophthalmologists should consider the possibility of an intracranial tumor in patients with compressive optic neuropathy.

Ophthalmological symptoms of intracranial tumors may include diplopia, proptosis, and visual disturbances, including a narrowing of the visual field. Visual disturbances usually have a slowly progressive presentation; however, in some cases of compressive optic neuropathy, a visual disturbance may have an acute or sub-acute onset [1]. Moreover, certain intracranial tumors are found incidentally. There are several subjective and objective signs of compressive optic neuropathy, including deteriorated visual acuity, abnormal color perception, relative afferent pupillary defect (RAPD), a defect observed during the visual field test), and optic disc swelling or atrophy. Diagnosis of compressive optic neuropathy based upon these signs is important in order to differentiate it from other optic neuropathies. Hence, in the present study, we evaluated the clinical characteristics of compressive optic neuropathy in patients who were initially diagnosed at the Kim's Eye Hospital (Seoul, Korea) and examined correlations between the ophthalmic examination and intracranial tumors.

2. METHODS

This retrospective study included patients with compressive optic neuropathy who were diagnosed with intracranial tumors on magnetic resonance imaging (MRI), visit to the Department of Neuro-Ophthalmology of the Kim’s Eye Hospital between January 2011 and December 2013.

Inclusion criteria for participation in the study were as follows: (i) the patients with visual problem which is related with visual pathway tumor was initially diagnosed at our hospital (ii) The visual pathway tumor had been confirmed by MRI, and (iii) the diagnosis of an visual pathway tumor was performed by one radiologist. Patients with strabismus and proptosis were excluded from this study, in order to solely evaluate the symptoms of visual disturbance related to intracranial tumors. In addition, we also excluded patients with previously known intracranial tumors and retrochiasmal lesions. This research study was reviewed and approved by the Institutional Review Board of the Kim’s Eye Hospital, and all patient procedures conformed to the guidelines of the Declaration of Helsinki.

General ophthalmological examinations including a visual acuity test, intraocular pressure measurements, and slit lamp and fundus examinations, were performed. Best corrected visual acuity was assessed using a Snellen chart at the initial visit. The relative afferent pupillary defect (RAPD) was graded using a neutral density filter bar with 6 steps (0.3–1.8 log units). To evaluate the optic disc, fundus photography was performed using a digital fundus camera (Canon CR6-45NM with Canon EOS 20D; Tokyo, Japan). The optic disc was assessed for abnormalities including swelling, disc pallor, and a large cup/disc ratio, which is defined as an optic disc with an intact temporal rim. An Ishihara color test, using 21 plates, was used to detect disturbances in color perception. As per the manual of the Ishihara color plate, the color vision test was considered abnormal when patients did not respond within 3 s. Goldmann perimetry (Takagi MT325-UD; Nagano, Japan) or Humphrey visual field perimetry (Carl Zeiss Meditec; Dublin, CA, USA) was performed where required.

3. RESULTS

Among the 89 patients who were diagnosed with a compressive optic neuropathy, 35 patients (22 male and 13 female; mean age, 43.7 years; age range, 6–77 years) met the criteria, and their medical records were reviewed. The most common tumors identified in these patients were pituitary tumors (60.0%), meningiomas (20.0%), and optic nerve glioma (5.7%) Fig. 1. Out of the seven meningioma cases, only two had originated from the optic nerve. A wide range of symptoms were identified among the 35 patients Fig. 2. Pituitary “incidentalomas” are, by definition, masses that are discovered by computed tomography or MRI, for the diagnosis of unrelated disorders (such as head trauma), cancer staging, or for other nonspecific symptoms such as dizziness and headaches.[2] The two "incidentalomas" (cases 2 and 4) were pituitary adenomas, one of which showed bilateral quadranopsia and the other, a unilateral arcuate defect.

Patients with compressive optic neuropathy had a range of visual acuity scores, from a maximum of 1.0 (normal vision) to no light perception Table 1. All 3 patients (cases 2, 4, and 11) with scores of 1.0 in both eyes had a pituitary adenoma Fig. 3. Further, 3 patients (patients 3, 24, and 29) had poor bilateral visual acuity and were in poor health, due to confounding pathologies such as dementia and uncontrolled diabetes mellitus. An RAPD was observed in 20/28 patients (71.4%). Eight of these patients had a negative RAPD score, of which 6 patients displayed a relatively symmetric visual field defect, and one patient had a small scotoma. One of the 8 patients with no RAPD was excluded from the visual field test because of poor visual acuity. Abnormal disc findings were identified in 20/34 patients (58.8%). The most common abnormality of the optic disc was optic disc pallor. Disc pallor was noted in 17/32 eyes with an abnormal optic disc, as well as in 8 with a large cup-to-disc ratio and in seven with an optic disc swelling. The color vision test was more specific (16/21 patients, sensitivity: 76.6%). For two of the 21 patients, results from the affected eye could not be obtained from the color vision test due to poor visual acuity. All patients who were able to take a visual field test exhibited abnormal visual field defects. A bitemporal hemianopsia was observed in 3/21 patients with pituitary tumors.

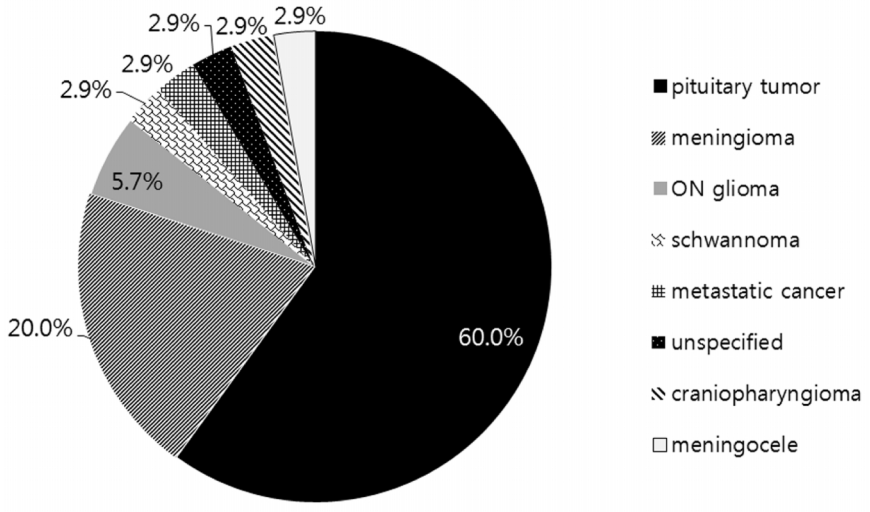

Fig. 1. The type of tumors

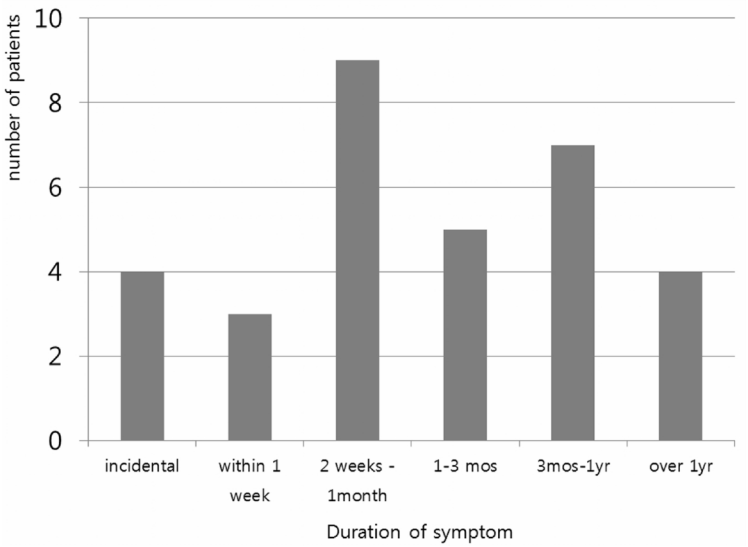

Fig. 2. The duration of time of onset

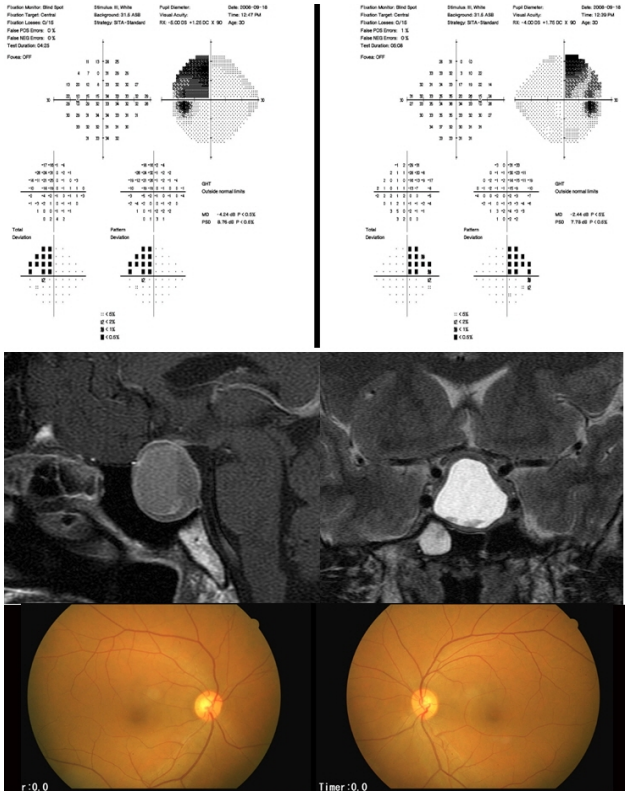

Fig. 3. Case 4; 30 year-old-man had a superior quadrant bitemporal hemianopia under routine examination (upper column). Lower column shows a pituitary adenoma compressing optic chiasm (right; T1 weighted sagittal section, left; T2 weighted coronal section)

Table 1. The full data of the 35 patients

Case	Sex	Age	Diagnosis	Visual acuity		Visual field test		Color test		Optic disc		RAPD
				Right	Left	Right	Left	Right	Left	Right	Left	
1	M	50	pituitary tumor	0.9	0.2	bitemporal		-		NL	pallor	OS equivocal
2	F	70	pituitary tumor	1.0	1.0	SA	NL	-		NL	NL	-
3	M	70	pituitary tumor	0.03	0.06	-	-	abnormal	abnormal	pallor	pallor	-
4	M	30	pituitary tumor	1.0	1.0	Q	Q	NL	NL	large C/D ratio	large C/D ratio	no
5	M	59	pituitary tumor	1.0	0.1	NL	diffuse	NL	abnormal	NL	disc swelling	OS (1.5)
6	M	44	pituitary tumor	FC	0.1	-	diffuse	abnormal	abnormal	large C/D ratio	NL	OD
7	F	62	pituitary tumor	HM	0.8	diffuse	SA	ND	abnormal	pallor	NL	OD
8	F	33	pituitary tumor	0.7	1.0	IA	NL	abnormal	NL	NL	NL	OD (1.2)
9	F	36	pituitary tumor	1.0	0.3	SA	hemianopsia	NL	abnormal	NL	NL	OD (1.2)
10	M	54	pituitary tumor	0.4	0.9	bitemporal				pallor	NL	no
11	F	54	pituitary tumor	1.0	1.0	NL	Q	NL	NL	large C/D ratio	disc swelling	OS (0.9)
12	F	70	pituitary tumor	0.5	HM	3Q	diffuse	-		pallor	pallor	OS
13	M	33	pituitary tumor	0.8	0.06	bitemporal		-		large C/D ratio	large C/D ratio	no
14	M	45	pituitary tumor	0.6	0.6	3Q	IA	-		NL	NL	-
15	M	39	pituitary tumor	1.0	0.1	3Q	SA	-		NL	NL	OS
16	M	66	pituitary tumor	0.3	1.0	3Q	temporal	-		pallor	NL	OD
17	F	29	pituitary tumor	1.0	0.7	NL	temporal	NL	abnormal	NL	NL	no
18	M	34	pituitary tumor	0.4	1.0	3Q	SA	abnormal	NL	large C/D ratio	NL	no
19	M	6	pituitary tumor	LP-	1.0	-	-	-		pallor	NL	OD
20	M	28	pituitary tumor	0.3	1.0	IA	NL	abnormal	NL	NL	NL	OD (0.3)
21	M	30	pituitary tumor	1.0	0.06	constriction	diffuse	NL	abnormal	NL	pallor	OS (0.9)
22	M	20	meningioma	0.6	0.6	NL	central	NL	NL	NL	NL	no
23	F	77	meningioma	0.2	0.6	3Q	NL	abnormal	NL	NL	NL	-
24	M	56	meningioma	HM	HM	diffuse	diffuse	-		disc swelling	pallor	OS equivocal
25	F	50	meningioma	1.0	FC	NL	diffuse	-		NL	pallor	OS (1.2)
26	M	54	meningioma	1.0	0.5	NL	Q	NL	abnormal	NL	NL	-
27	M	36	craniopharyngioma	0.5	0.8	Q	temporal	-		disc swelling	disc swelling	no
28	M	7	schwannoma	1.0	0.2	NL	central	abnormal	abnormal	NL	large C/D ratio	OS
29	M	75	unspecified	LP-	LP-	-	-	-		- pallor	pallor	no
30	F	63	metastatic carcinoma	HM	0.2	diffuse	3Q	ND	abnormal	pallor	pallor	-
31	M	10	Optic nerve glioma	1.0	0.2	Q	SA	-		-		-
32	F	39	Optic nerve glioma	FC	1.0	BS enlarged	NL	ND	NL	disc swelling	NL	OD
33	F	54	Optic nerve meningioma	1.0	FC	NL	diffuse	NL	ND	NL	disc swelling	OS 1.8<
34	F	40	Optic nerve meningioma	0.1	1.0	-	-	abnormal	NL	pallor	NL	OD 1.8<
35	M	8	meningocele	0.4	0.8	Q	diffuse	abnormal	abnormal	NL	NL	OD equivocal

3Q: visual field defect in the three quadrants. Q: quadranopsia, SA : superior arcuate defect, IA : inferior altitudinal, BS : blind spot, NL : normal finding, FC : finger count, HM : hand motion, LP : light perception, - : no data, ND : not detectable

4. DISCUSSION

A compressive optic neuropathy resulting from an intracranial tumor may present at any age. In the present study, the youngest age at which a compressive optic neuropathy was observed was 6 years. This patient had no light perception in the right eye and the chief complaint was exotropia. A pituitary tumor measuring 2.8 × 2.9 × 3.2 cm, was detected by MRI. Therefore, patients presenting with atypical optic neuritis or progressive visual disturbances, should also be clinically assessed for an intracranial tumor. Because compressive optic neuropathy is treatable, and in some cases curable, it should be distinguished from other optic neuropathies such as optic neuritis, ischemic optic neuropathy, hereditary optic neuropathy, and traumatic optic neuropathy.

Tumors affecting the optic nerve are usually divided into primary (optic nerve glioma and optic nerve meningioma) and secondary (meningioma, metastatic cancer, and pituitary adenoma) optic nerve tumors. We identified a primary optic nerve tumor in only 4 cases (two optic nerve gliomas and two optic nerve meningiomas). The low incidence of primary optic nerve tumors in the present study may be because these tumors are often accompanied by proptosis, and a primary optic tumor without proptosis is likely to be only detected in the advanced stages of the disease. The visual acuity of the 4 patients with primary optic nerve tumors varied from 0.2 (low) to finger count (high).

Pituitary adenomas were the most common tumor type identified in the present study. The clinical features of pituitary adenoma are diverse, and the visual acuity can range from hand motion to 1.0, together with large variation in visual field defects. High variability on the prevalence of visual field defects with pituitary adenoma has been reported. In one study, 94.6% had a visual field defect [3], whereas another study [4] reported that only 9% of patients with a pituitary adenoma had a visual field defect. In this present study, all patients with a pituitary tumor and those who underwent the visual field test showed a visual field defect.

A pituitary incidentaloma is a previously unsuspected pituitary lesion that is discovered by clinical imaging performed for an unrelated reason [5]. In these cases, by definition, the imaging study is not performed for a symptom specifically related to the lesion, such as visual loss, a clinical manifestation of hypopituitarism, or hormone excess, but rather more commonly for the evaluation of symptoms such as headache, or other head or neck complaints neurological or central nervous system, or a head trauma. The incidentaloma prevalence in those with a pituitary adenoma in our cohort was 2/21 (9.5%), with these two cases showing small quadranopsia without visual disturbance and color change.

The second most common tumor identified in this study was meningioma. Orbital meningioma may be either a primary tumor of the optic nerve arachnoid or a secondary tumor due to an extension of the meningioma into the orbit from a primary intracranial meningioma. Only 10% of orbital meningiomas are primary tumors of the optic nerve [6]. In the present study, 2/7 (28.6%) meningiomas were identified as primary tumors. Visual loss may be markedly decreased at presentation because of the compressive nature of the tumor. In this study, two patients with meningioma showed finger count and 0.1 at presentation. It is possible for optic nerve meningioma cases to be misdiagnosed as optic neuritis or amblyopia especially in children [7]. Therefore, the presence of an optic canal meningioma should be investigated in patients with unexplained visual loss, particularly when the symptoms are progressive [8]. Metastatic orbital lesions have been estimated to account for 1–13% of all orbital tumors reported, with the majority of metastatic lesions of the orbit and

ocular adnexa originating from breast carcinomas [9]. The results of this study identified only one metastatic cancer that originated from a breast metastatic carcinoma.

The present study used numerous clinical examinations to facilitate the detection and diagnosis of a compressive optic neuropathy. The most informative of the examinations employed were the RAPD and fundus examination, since the tests of visual acuity, visual field, and color vision were subjective, and therefore dependent upon patient cooperation. The prevalence of an abnormal optic disc detected by fundus examination was similar to that of RAPD (70% and 69%, respectively). It has been previously reported that RAPD increases linearly with an increased visual field defect [10]. However, Kardon et al. [11] reported wide variation in the correlation between RAPD and visual field defects with regard to different diseases. In some cases, RAPD may be normal in the early stages of the disease, or when visual field loss is equivalent in both eyes. In the present study, 6/8 (75%) non-RAPD cases had a similar defect as assessed by the visual field test. Of these six affected patients, 1 showed a small scotoma.

Optic disc findings are sometimes normal in the early stages of disease. In the present study, patients with a normal optic disc were noted to have relatively good visual acuity, with a visual acuity >0.5 identified in 6/11 patients (54.5%). The most common and important abnormality of the optic disc identified in this cohort was optic disc pallor. Optic disc pallor is an indicator that the disease has been present for >6 weeks, and therefore irreversible changes in optic nerve function should be suspected. Diagnosis of compressive optic neuropathy is therefore essential at an early stage, prior to damage occurring to the optic disc. In addition, optic disc swelling may be an initial symptom of compressive optic neuropathy [12].

A color vision test is a useful tool for compressive optic neuropathy diagnoses owing to its high sensitivity, and intracranial tumors with compression of visual pathways show abnormal color test results. However, the color test is not applicable when the visual acuity is lower than the finger count. Visual field tests showed abnormal findings in patients who were able to perform the color test. A visual field defect may facilitate the localization of the lesion; however, these defects are often nonspecific and delayed with respect to electrophysiological alterations [13]. Although bitemporal hemianopsia is a characteristic indicator of chiasmal disease, only 3/21 (14%) pituitary tumors revealed a typical bitemporal hemianopsia. Despite the low number of pituitary tumor cases, the present study may be valuable in determining the use of the visual field test in diagnosis and incidence of tumors in compressive optic neuropathy. A limitation of our study is the relatively small number of patients investigated. However, visual pathway tumor presenting as visual disturbances without extraocular signs is a relatively rare condition, and the present paper may give useful information to detect compressive optic neuropathy.

In conclusion, the visual field, color vision and RAPD tests and the optic disc appearance should be utilized routinely in the clinical practice to facilitate the diagnosis of compressive optic neuropathy. Among these tests, the visual field is the most sensitive. Although pituitary tumors are common, rare tumors such as metastatic cancer or optic glioma must also be considered during diagnosis.

CONSENT

All authors declare that ‘written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the IRB of Kim's Eye Hospital and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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