NEURO-OPHTHALMOLOGICAL FINDINGS IN CRANIOPHARYNGIOMAS

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SUMMARY :

Visual disturbances are common in patients with craniopharyngioma (CP). Thirty-five patients (17 adults and 18 children) with craniopharyngioma were examined neuro-ophthalmologically. Their visual acuity, visual fields, pupilla and fundoscopic examinations were evaluated in respect to intra-operative tumour localization, and age. Tumour localization and visual acuity were similar in both age groups. Normal fundi were more common in adult group, while papilloedema was more common in the paediatric group. Tumour localization was evaluated in four groups: Group I: intrasellar, Group II: parasellar extension, Group III: suprasellar extension, Group IV: extension into the third ventricle. There were more patients with normal visual acuity in Groups I and II, and more with defective visual fields in Group III. It is important for the ophthalmologist to recognize the visual findings of CP, which are usually the presenting complaint of this tumour.

KEY WORDS :

Craniopharyngioma, visual field, visual dysfunction.

INTRODUCTION

Craniopharyngioma (CP) is a benign tumour derived from the Rathke pouch which is part of the embryonal craniopharyngeal tube. It constitutes 3% of all intracranial tumours, and 8-13% of intracranial tumours in childhood (7). The most common complaints caused by CP are headache and visual disturbances (6,8). They mostly occupy the suprasellar cistern, and compress the intracranial portion of the anterior visual pathways. The compression of the tumour is depenent on its size, direction of growth, and the anatomy of the optic chiasm (7). While loss of vision is usually the presenting symptom in adults; in children, visual disturbances may be noted in later stages of the disease (4). We herein present our experience of the relationship of visual findings and tumour localization in children and adults with CP.

MATERIALS AND METHODS

Sixty-three patients were seen in the Department of Neurosurgery of Hacettepe University Hospitals between 1979 and 1988. Their case notes were reviewed, and 35 patients with fully-recorded visual findings in the Neuro-ophthalmology Unit were included in this study. The Patients' visual acuity, pupil reaction, visual field tests, and fundoscopic examinations were evaluated in respect to per-operative tumour localization, and visual function according to age. Patients younger than 18 years were included in the paediatric group (PG), and those older than 18 years were accepted as adults (AG). Post-operative evaluation of 13 patients who attended follow-up visits was also recorded. Localization of tumour was grouped as: Group I. Intrasellar localization, Group II. Parasellar extension, Group III. Suprasellar localization, Group IV. Extension to third ventricle, and larger tumours. All patients had Computed Tomography (CT) scans, and 18 had cerebral angiography.

RESULTS

There were 19 female and 16 male patients, and their ages ranged between 6 and 57 (mean 23 years, half were between 6 and 16 years). The AG consisted of 10 female and 7 male patients with a mean age of 36 years, and there were 9 females and 9 males in PG (mean age 11 years). Seventy-one percent of the patients had presented with the complaint of visual loss. Visual loss and/or visual field defect and/or fundoscopic abnormality (papilloedema or optic atrophy) was noted in 91.4%. Twelve patients in the PG (67%) were seen with the chief complaint of poor vision. Duration of symptoms in this group ranged between 3 days and 3 years (mean 10.5 months). Similarly 13 patients (76%) in the AG group presented with poor vision with a duration of 15 days ta 5 years (mean 13 months).

Total or subtotal tumour resection was performed in all but one patient in whom a ventriculo-atrial

shunt was placed as the tumour could not be resected. Thirty-one patients were given post-operative radiotherapy.

Tumour localization : The localization of tumours according to age groups is shown in Table 1. Tumour localization was similar in both groups. Fundoscopic findings of the patients with CP are summarized on Table 2. While there were more adults with normal fundi ($P \leq 0.001$) papilloedema was more common in the padiatric group ($P \leq 0.05$). There was no statistical difference between the visual acity of the groups (Table 3).

TABLE 1 : Localization of Mass According to Age Groups

Group	Paediatric Group	Adult Group
I	–	3
II	1	–
III	6	5
IV	11	9
Total	18	17

TABLE 2 : Fundoscopic Findings of Patients According to Age Groups (Number of Eyes)

Fundus	Paediatric Group	Adult Group
Atrophy	9	4
Pallor	6	11
Oedema	18	4
Normal	3	15

TABLE 3 : Acuity According to Age Groups (Number of Eyes)

Visual Acuity	Paediatric Group	Adult Group
20/20 - 20/40	15	20
20/50 - 20/100	3	3
20/200 - 20/800	8	6
Less than 20/80	6	4
Blind	4	1

Group I: (Table 4). Two patients out of 3 in this group had normal visual functions. Group II: (Table 4). The only patient in this group had severe visual loss. Group III: (Table 5). Totally, 73% of the patients had visual loss, and 62% had a visual field defect. Group IV: (Table 6). Totally, 68% of the patients had visual loss, and 27% had visual field defects. The types of visual field defect are shown in Table 7 according to localization. For a statistical comparison of visual field defects among groups; groups I and II were calculated together. Then, there was a significant difference between groups III and IV ($P < 0.05$) according to the presence or absence of a visual field defect. Also, there were more patients with visual field defects in group III when compared to groups I and II ($P \leq 0.05$). AS far as visual acuity is concerned, there were more patients with normal visual acuity in groups I and II than the other groups ($P \leq 0.05$).

TABLE 4 : Neuro-Ophthalmological Findings of Patients with Craniopharyngioma with Intrasellar Localization and Parasellar Extension

GROUP I : Craniopharyngiomas with intrasellar localization (3 patients)

Patient	Duration of Visual	Visual Acuity		Visual Fields		Fundi		Pupilla	Other
		OD	OS	OD	OS	OD	OS	OD/OS	
1	24 mo	20/20	20/20	N	Lower T def	P	P	+/+	–
3	–	20/20	20/20	N	N	N	N	+/+	–
8	–	20/20	20/20	N	N	N	N	+/+	–

GROUP II : Craniopharyngioma with parasellar extension (1 patient)

Patient	Duration of Visual	Visual Acuity		Visual Fields	Fundi		Pupilla	Oher
		OD	OS	OD/OS	OD	OS	OD/OS	
12	36 mo	p+p6	p+p+	Incongruous righthomonymous hemianopsia	A	A	RAPD.	–

N: Normal, T: Temporal, P: Pale, A: Atrophy, RAPD: Relative Afferent Pupillary Defect.

TABLE 5 : Neuro-Ophthalmological Findings of Patients with Craniopharyngioma with Suprasellar Localization

GROUP III : Suprasellar Craniopharyngiomas (20 patients)

Patient	Duration of		Vis.	Acuity	Visual Fields	Fundi	Pupilla	Other	
			OD	OS	OD,OS	OD	OS	OD/OS	
2	6	mo	20/40	20/30	Bil. low T. quadr.	N	N	+/+	–
5	1.5	mo	20/25	20/40	Right hom. up quadr	N	N	+/+	–
6	4	mo	h.m.	20/50	–, T hemianopsia	A	A	+/+	–
7	36	mo	5mfc	0	–,–	A	A	?	–
9	3	mo	20/50	20/400	Bil. T. hemianopsia.	A	A	RAPD	–
13	24	mo	20/200	20/200	–,–	E	E	+/+	–
14	20	days	20/20	20/20	N,N	E	E	+/+	6 CN.
15	1	mo	20/30	20/400	Bil. contraction	E	E	+/+	6 Cn.
16	26	mo	0	2mps	–, T. hemianopsia	A	A	+/+	–
17	–		20/25	20/25	N,N	E	E	+/+	6 CN.
18	3	mo	p+p+	20/400	–, contraction	P	P	+/+	–
20	12	mo	20/20	20/400	Bil. T. hemianopsia	P	P	RAPD	6 CN.
23	15	days	20/20	0	Uncooperative	N	A	RAPD	–
25	–		20/20	20/20	N,N	N	N	+/+	–
20	10	mo	20/40	20/200	OD T. hemianopsia OS central scotoma	N	P	+/+	–
30	15	days	20/100	20/400	OD paracentral.	P	P	+/+	–
31	4	mo	0	p+p+	–,–	P	P	+/+	–
32	2	mo	20/30	20/20	N,N	N	N	+/+	–
33	–		20/20	20/20	OD upper T defect. OS normal	S	S	+/+	–
35	5	years	20/20	20/20	Bil. T. hemianopsia	S	S	+/+	–

T: Temporal, N: Normal, P: Pale, A: Atrophy, E: Oedema, DAPD: Relative Afferent Pupillary Defect, CN: Cranial nerve.

Follow-up Examination Findings:

Four patients died in the post-operative period and 13 attended follow-up visits for a period of 2 months to 9 years after the operation (mean duration of follow-up was 2 years). The rest were lost to follow-up. Among 13 patients, 5 had unchanged visual acuity, one deteriorated, and 7 had improved more than two lines in the visual acuity chart. Similarly while 7 patients had unchanged visual fields, 6 improved more than 20 degrees peripherally. Fundoscopic examination disclosed unchanged findings in 8 patients, and a return to normal in one. Four patients' fundoscopy revealed progression to optic atrophy from previous papiloedema or pale discs. Only one patient in this latter group had deterioration of visual acuity.

DISCUSSION

Our female/male ratio was 1:2 which is consistent with previous findings (7). Similarly our figures also correlate with others as the most common complaint or visual disturbances (77%), and headache (80%) in patients with CP. Repka et al. (9) reported visual loss in all their patients with the same mean duration in both paediatric and adult patients.

Visual Acuity and Visual Fields :

It is known that the visual dysfunction caused by CP is mainly dependent on the localization, size and direction of extension of the tumour. Intrachiasmal lesions, if they grow upwards, may compress and flatten the optic chiasm. Tumours expanding towards the third ventricle may interfere with cerebrospinal fluid (CSF) circulation and may cause hydrocephalus. Despite more frequent localization of CP in the supra sellar area, rarely, it may be intrasellar, and growth towards the pituitary gland and upwards may be the reason for compression. Rarely does the tumour expand to the cerebrallar, and growth towards the pituitary gland and upwards may be the reason for compression. Rarely does the tumour expand to the cerebellum, brainstem, thalamus or hemispheres (7). In our series the tumour was localized at the

TABLE 6 : Neuro-Ophthalmological Findings of Patients with Craniopharyngioma Extending into The Third Ventricle

GROUP IV : Craniopharyngiomas with extension into the third ventricle (11 patients)

Patient No.	Duration of Visual Disturbance	Visual Acuity		Visual Fields	Fundi		Pupilla	Other
		OD	OS	OD,OS	OD	OS	OD/OS	
4	2 mo	20/400	20/400	Bil. T depression	P	P	+/+	–
10	3 days	0	20/20	Uncooperative	E	E	RAPD	Ptosis, nys.
11	1 mo	20/20	20/200	Bil. big blind spot	E	E	RAPD	–
19	4 mo	20/40	20/40	N,N	E	E	+/+	–
21	–	2mfc	2mfc	Uncooperative	E	E	–/–,	NR:+/+
22	–	20/20	20/20	N,N	E	E	+/+	–
24	?	20/800	20/800	Uncooperative	E	E	+/+	6 CN
26	?	20/100	20/70	Bil. T. hemianopsia	A	A	+/+	upgaze –, ET
27	–	20/20	20/20	N,N	E	E	+/+	6 CN
28	–	20/20	1mfc	Uncooperative	N	N	+/+	7 CN
34	1 mo	20/200	20/100	N,N	N	N	+/+	

P: Pale, E: Oedema, N: Normal, NR: Near Reaction, RAPD: Relative Afferent Pupillary Defect, nys: nystagmus, ET: esotropia, CN: Cranial Nerve.

intrasellar region in 3 patients, had extended towards the parasellar region in one, towards the suprasellar region in 20, and into the third ventricle and towards the hemispheres in 11 patients. The patient's age may be relevant to the symptoms and findings of craniopharyngioma. Loss of vision is reported to be a late finding in the paediatric age group, whereas it is one of the early symptoms in adults (4). Kennedy and Smith (6) found the rate of visual loss to be 50% in children and 64 % in adults, and Repka et al. (9) found 50% and 36% visual loss in paediatric and adult patients respectively, whereas Bartlett (1) separeted his patients into two groups aged 15-45 and older, and the rate of visual loss was 79% and 74% respectively. ALthough one would expect adults to note their visual symptoms earlier than children thus decreasing the delay in diagnosis and therapy; we found per-operative tumour extension to be similar in both groups. Also we found no differences between the AG and PG groups for the rate of visual loss. On the other hand in the series of Repka et al. (9) children presented with poorer visual function than the adults. The reason for our findings and theirs is delayed diagnosis of visual dysfunction. The visual findings of CP were classified into 4 groups: optic neuritis like; silent optic neuropathy in one eye and anterior juctional syndrome in the other eye; chiasmal syndrome with bitemporal defects; and tractus type visual loss (7). Therefore, loss of visual loss may aid the clinician to localize the tumour. Acute loss of vision which may mimic retrobulbar neuritis is due to an acute circulatory disturbance in the optic chiasm (2,5,7). For example, in one of our patients (Patient No: 30) lose of vision started acutely with paracentral scotoma in the right eye and central scotoma in the left eye. The findings in this particular patient were discovered after a traffic accident, and a mass was found in the suprasellar localization in close proximity to the optic chiasm; thus supporting the above postulate. Johnson and colleagues (5) described a patient with acute loss of vision who on normal CT had no abnormality, but enhanced CT, and Magnetic Resonance Imaging disclosed a suprasellar mass, a CP. This case shows how CP can easily be misdiagnosed as optic neuritis. Our most common visual field finding was bitemporal hemianopsia (Tablo 7). Kennedy and Smith (6) reported similar findings with 27% bitemporal, 20% prechiasmal, and 11% homonymous haemianopic defects.

Fundoscopic Findings :

It is reported taht papilloedema is more common in childhood CP whereas optic atrophy is more common in adults. Optic atrophy in childhood is mainly caused by direct by direct pressure of the tumour mass on the optic nerves (7). In the PG we observed 47% temporal pallor and atrophy (unilateral or bilateral), and 50% papilloedema. Whereas in the

TABLE 7 : Visual Field Defects in Various Groups of Tumour Localization

Defect	I. Group	II. Group	III. Group	IV. Group
	Total : 3	Total : 1	Total : 20	Total : 11
Bil. T. hemianopsia	–	–	1	4
Bil. T quadrantopsia	–	–	–	1
Bil. T. hemianopsia, central scotoma	–	–	1	–
Uni. T. quadrantopsia	1	–	–	1
Uni. T. hemianopsia, uni. central scotoma	–	–	–	1
Homonymous hemianopsia	–	1	–	–
Hononymous quadrantopsia	–	–	–	1
Central and paracentral scotoma	–	–	–	1
Bil. big blind spot	–	–	1	–
Contraction	–	–	–	2

AG the corresponding rates were 43%, and 12 respectively. (Table 2). Repka et al. (9) reported a rate of 50% of optic atrophy in the paediatric group and 30% in the adult group. Similarly we found more normal fundus in the AG whereas there was significantly more papilloedema in children.

Cranial Nerve Involvement :

It is generally accepted that sixth nerve paresis in CP is in fact a sign of increased intracranial pressure, and third nerve paresis is caused by intracranial haemorrhage. We also explained the sixth nerve paresis in 6 patients by increased intracranial pressure, as they had other confirmatory findings. We had one patient with horizontal nystagmus, one other with central seventh nerve paresis and hemiparesis, possibly due to tumour localization. Ingrahan and Scoot (4) noted 15% strabismus in paediatric patients with CP. We, however, had only one patient with strabismus who had the same visual acuity in both eyes which we believe to be a coincidental finding. One of our patients with paresis in upgaze was accepted as Parinaud Syndrome since she had a tumour in the third ventricle causing hydrocephalus with associated pupillary findings.

Pupilla Signs :

The patient with Parinaud Syndome had light-near dissocation. This finding has only reported one (3). She had upgaze for only a short time in the post-operative period.

CONCLUSION

CP are mainly suprasellar in localization, they can expand into the third ventricle, and, rarely, can be intrasellar or with parasellar extension. Therefore, it is of prima importance for neurosurgeons and ophthalmologists to recognize the visual findings which are usually the presenting complaint of CP.

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