Craniopharyngiomas: Clinical Presentation and Endocrinological Evaluation.

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Ten patients with craniopharyngioma were studied in three years period. There were nine(90%) males and one (10%) female. Age of these patients ranged from 2-58 years. The major clinical features were headache (70%), deterioration of vision (70%) in the form of decreased visual acuity and field defects. Endocrinopathies were present in the form of short staturedness (40%), underdevelopment of secondary sex characters (40%), loss of libido and impotence (20%). Endocriological evaluation revealed Gonadotrophin deficiency in 50% pituitary - thyroid axis deficiency in 40% and serum cortisol deficiency in 40% patients. Eight percent patients had hyperprolactinemia. Growth hormone levels were found within normal range. On CT scan, only 20% patients had calcification, ninty percent patients had tumors in suprasellar region and 30% had parasellar extension. Sixty percent lesions were cystic and 40% were mixed-density lesions. Transcranial subfrontal approach was adopted and surgery was done under the microscope. In six patients total removal was possible and in the remaining four patients where total removal was not possible, postoperative radiotherapy was advised. The patients with hypopituitarism were given hormone replacement therapy accordingly.

Key words: Craniopharyngioma, endocrinological evaluation

Craniopharyngioma constitutes between 2.5-4 % of all the intracranial tumors[1]. The origin, natural history, operative removability, response to radiation and optimal therapy of the lesion have all been topics of debate[2]. This is the most common tumor of non-glial origin, representing 59% of all the suprasellar tumors in children and 20% of those in adults[1] There is bimodal age distribution, with the first peak at 5-10 years and a second peak between 55-65 years, but the tumor may become symptomatic at any age Recent large series had shown equal sex distribution[3].

It is said that craniopharyngioma may indeed have dual origin. The so-called childhood type may be of embryonic origin. The adult type, which occurs mostly in adults and consists mainly of mature stratified squamous cells, may be of metaplastic origin[3].

The common clinical manifestations fall into four groups: raised intracranial pressure, failing vision, endocrine disturbances and mental impairment. These are determined by the site of origin and direction of growth. In the majority the presenting symptoms are headache, vomiting and / or impairment of vision which may be in the form of field defects or decreased visual acuity. Endocrine disturbances are evident, in the form of obesity, stunted growth, hypogonadism, diabetes insipidus and hypopituitarism in about three quarters of patients but they occur very slowly. Mental impairment is present in over one third of the patients, varying from duilness and impaired memory to frank dementia[4].

About 90% of craniopharyngiomas are cystic, at least in part, lined with squamous epithelium and containing dark brown fluid like engine oil, which is full of cholesterol crystals that shimmer in the light. Small intrasellar tumors do occur, but 50% are suprasellar, causing no enlargement of the pituitary fossa, and growing mainly upwards towards the hypothalamus and third ventricle where they may cause obstruction of the foramen of Monro. They are thin walled and insinuate themselves into their surroundings, making total surgical removal difficult[5].

Craniopharyngiomas have distinctive histological features and a pronounced tendency to invade locally and to recur following therapy. Two variants have been described,the adamantinomatous and papillary craniopharyngiomas. Whereas adamantinomatous tumors occur at all ages, predominantly during childhood and early adolescence, papillary craniopharyngiomas have been reported less prone to recurrence[6]. Most reviews conclude that total excision should be attempted whenever feasible and sub-total removal with adjuvant radiotherapy should be done when resection is dangerous. In cases of recurrence, radical tumor removal, subtotal resection and / or radiotherapy are again the usual treatment modalities. Strict endocrinological evaluation and hormonal replacement therapy are essential in all patients in both immediate and long-term follow-up[1].

Material and Methods

This is a prospective study conducted at the Department of Neurosurgery, Lahore General Hospital, Lahore, which includes ten cases with craniopharyngiomas presented during the study period (January,1995- December,1997). A detailed history, clinical and ophthalmological

examination was done. Special consideration was given to clinical presentation and endocrine evaluation. In all the patients with clinical suspicion, skull X--rays were done and diagnosis was confirmed with computed tomography (C.T) / magnetic resonance imaging (MRI). The following hormone levels were checked for:

- Serum prolactin(PRL)
- Growth hormone(G.H)
- Serum cortisol
- Serum T4 & Thyroid stimulating hormone(TSH)
- Serum leutinizing hormone(L.H)
- Serum follicle stimulating hormone(FSH)

Results

Age and Sex Incidence

The age ranged from 2-58 years, with a mean age of 22 years. The maximum number of patients (60%) were in the 2nd and 3rd decades of life. Out of these ten patients with Craniopharyngiomas the males predominated with a male to female ratio of 9:1.

Clinical Presentation (table 1)

Headache was the main presenting feature in 7 patients. It was dull in nature, generalized and of moderate intensity.

Deterioration of vision was present in 7 patients. Among them two patients had complete loss of vision and three had grossly decreased visual acuity. One patient had bitemporal field defect and one had left homonymous hemianopia. Examination of fundi showed bilateral optic atrophy in 5 patients and two patients had bilateral papilloedema.

Table 1 Clinical presentation in ten patients with craniopharyngiomas.

Clinical feature	n=	%age
Headache	7	70
Deterioration of vision	7	70
Complete loss of vision	2	20
Decreased visual acuity	3	30
Bitemporal field defect	1	10
Homonymous hemianopia	1	10
Optic atrophy	5	50
Papilloedema	2	20
Endocrinopathies	7	70
Short stature	4	40
Underdeveloped secondary sex characters	4	40
Loss of libido & impotence	2	20
Obesity	3	30
Polyuria	2	20
Mental Changes	2	20
Seizures	1	10

Clinical evidence of endocrinopathies was present in 7 patients. Underdevelopment of secondary sexual characters was present in four cases. Four patients were short statured and two patients had loss of libido and impotence. Three patients were obese. Two had polyurea at the time of presentation.

Two patients were found lethargic, confused and disturbed memory.

Only one patient had seizures. They were generalized tonic clonic in nature.

Endocrinological Evaluation (Table 2)

Total of seven patients had hypopituitarism. Among these the following pattern was observed. Gonadotrophin deficiency in five patients Pituitary -thyroid axis deficiency in four cases . Serum cortisol levels were low in four patients Growth hormone levels were within normal range in all patients.

Serum prolactin levels were slightly raised in eight patients but they were below 100 ng / ml.

Table 2 Endocrinological evaluation in ten patients with craniopharyngiomas

PRL. ng/ml	G.H. ng/ml	S/ Cortisol microg/dl	T4-TSH nmol/l.-mIU/l.	LH-FSH mIU/ml
88	1.2	24.6	199-0.81	9.18-10.67
52	2.3	12.0	72-3.24	4.40-0.96
59	1.6	24.0	77-1.82	4.44-1.06
70.5	3.1	9.2	104-2.5	9.84-8.20
6.5	3.4	18.4	50-0.4	0.80-0.09
38	1.9	6.0	98-2.0	8.20-7.80
37	2.2	1.2	150-5.6	3.90-3.20
49	0.29	3.17	47-0.5	4.20-4.30
12.7	2.0	0.30	54-0.6	8.70.60
35	1.9	2.6	60-0.5	8.50-6.70

Radiological Investigations

X-rays skull

In only one patient sella was found eroded and in the rest of the patients it was found to be normal. Two patients (20%) had evidence of calcification in the suprasellar region.

Computed Tomographic (C.T) scan / Magnetic Resonance Imaging (MRI)

These investigations showed that in 9 patients (90%) tumor was in suprasellar region and in one patient (10%) tumor involved the sella as well. Four (40%) patients had parasellar extension. Six patients (60%) had cystic lesions and four (40%) had mixed density lesions.

Treatment

All patients underwent surgery as a primary mode of treatment. In all the cases transcranial subfrontal approach was adopted and surgery was done under the microscope. In six patients (60%) total removal of the tumor was possible. In the rest of the four (40%) patients, where tumor removal was incomplete, postoperative radiotherapy, as a second treatment modality was advised. The patients with hypopituitarism were given hormone replacement therapy accordingly.

Discussion

In the present study of ten patients with craniopharyngioma the age ranged from 2-58 years with a mean age 22 years. Landolt and Zachmann[7] had reported

the age range between 6-43 years with a mean age of 22 years. Crotty[6] has reported the age range from 10-74 years with a mean age of 44.7 years. In our study there is gross male predominance with a male to female ratio of 9:1. Landolt and Zachmann[7] have reported the sex incidence with a male to female ratio 11:3 and according to Crotty[6] there is equal sex distribution.

In the present study, the clinical presentation is dominated by the symptoms and signs of "mass effect" and "hypopituitarism". Headache and deterioration of vision was present in 7(70%) patients each. In one study Crotty[6] gave an incidence of headache as 84% and that of deterioration of vision as 68%. In our study headache was present in 70% patients and deterioration of vision in 68%. In our study 70% patients had clinical evidence of endocrinopathies. Growth failure was present in 40% of cases in our study but it was 75% in one study carried out by Landolt and Zachmann[7]. Loss of libido and impotence was present in 20% of cases in our study while it was 27% in Crotty's study[6] and 35% in a study carried out by Landolt and Zachmann[7]. In this study 20% patients had mental changes which is comparable to the figure of 22% reported by Crotty[7].

Endocrinological evaluation of the ten patients in this study revealed loss of endocrine function in 7(70%) patients while Landolt and Zachmann[7] reported this incidence to be 71.5% . In this study gonadotrophin deficiency was present in 50% and pituitary-thyroid axis deficiency was present in 40% patients while they were 88% and 46.5% respectively in a study carried by Landolt and Zachmann[7]. In this study serum cortisol levels were deficient in 40% patients and hyperprolactinemia in 80% of patients while these figures were 43% and 44% respectively as reported by Landolt and Zachmann[7]. Growth hormone levels were within normal range in our study while 77% patients were found deficient in growth hormone secretion as reported by Landolt and Zachmann[7].

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