Craniopharyngiomas—a summary of 85 cases

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SUMMARY The results of treating 85 patients suffering from craniopharyngioma are reviewed. The recurrence of the tumour after 20 symptom-free years after radical excision is reported. Claims for successful treatment of this illness should be reviewed in this light.

Craniopharyngiomas are curious tumours in that, instead of running a progressive course, the symptoms which they produce are often intermittent and variable; and although they are congenital tumours (Erdheim, 1904; Grinker, 1943) they may not cause symptoms until late in life. They declare themselves in a variety of ways. Disturbances of growth and sexual development, with and without evidence of raised intracranial pressure, are common during childhood, contrasting with the relative frequency of dementia in the elderly patient (Ross Russell and Pennybacker, 1961). Visual failure from compression of the optic pathway occurs with about equal frequency in all age groups. The duration of the illness, measured from the time of the first symptom to death, varies from a matter of a year or so to several decades. Recent reports of good results from radical excision (Matson and Crigler, 1969) and radiotherapy (Kramer, Southard, and Mansfield, 1968) make pertinent a review of the cases seen at the Radcliffe Infirmary over a 32-year period.

PATIENTS

Eighty-five patients were seen in the Departments of the Radcliffe Infirmary between the years 1938 and 1970. Seventy-two patients were operated upon and in 63 histological proof of the diagnosis was obtained. Eleven patients were not subjected to surgery because it was felt that either the symptoms were relatively trivial and of a non-progressive nature, or that surgery was unlikely to relieve the particular complaint. Two patients refused operation.

RESULTS

CLINICAL FEATURES There are 85 patients included in this series—42 are male and 43 are female. Thirty came under medical supervision before the age of

15 years. The remaining patients were distributed in a fairly even manner throughout the remaining decades, the oldest patient being 71 years of age at the time of presentation.

Evidence of raised intracranial pressure in the form of papilloedema was common, occurring in 43% of those less than 15 years of age. This is in contrast to dementia which occurred more frequently in the older patient, occurring in 31.5% of those over the age of 45 years.

The symptoms of these two groups of patients are summarized in Tables 1 and 2.

MANAGEMENT The surgical treatment of these tumours has been unsatisfactory, as shown by the fact that only 20 out of 73 cases on whom an operation was performed have survived 10 years. There are 14 additional patients who have the potentiality

TABLE 1

SYMPTOMS IN PATIENTS UP TO AND INCLUDING 15 YEARS OF AGE

Symptom	(no.)	(%)
Visual failure	23	76
Papilloedema	13	43
Growth disturbances	12	40
Mental symptoms	4	13
Hypopituitarism (alone)	1	3
Failure of puberty (alone)	1	3

TABLE 2

SYMPTOMS IN PATIENTS OF 45 YEARS OR OLDER

Symptom	(no.)	(%)
Visual failure	14	74
Dementia	6	32
Pituitary failure	6	32
Papilloedema	1	5

to live as long, making the maximum possible 10-year survival rate 43%.

Table 3 summarizes the surgical procedures and the frequency with which they were used. Often several operations were done on the same patient.

The polar graph Fig. 1 summarizes the results of the various methods of management. Each patient's history is represented by a line on this graph. The perimeter of the circle represents the time of the first operation. The further the line approaches the centre of the circle the longer is the survival. The time scale between the perimeter and the centre is linear and represents 30 years. This Figure shows that where it was possible to perform a radical removal of the

tumour the patients have consistently survived the longest period of time. Table 4 gives further details of this group. Tumour recurrence productive of symptoms has occurred in three cases; in two 20 years after the original operation and both patients died as a direct result. In the third case a cystic

TABLE 3	
PROCEDURES AND NUMBER OF TIMES EACH WAS USED	
Procedure	(no.)
Partial removal	112
Radical removal	6
Cyst aspiration	36
Ventriculocisternostomy	22
Radiotherapy	15

TABLE 4				
PATIENTS IN WHOM RADICAL EXCISION OF TUMOUR WAS POSSIBLE				
Case	Sex	Age at operation (yr)	Follow-up after radical removal (yr)	Notes
M.H.	M	1½	16	Cystic recurrence aspirated after 15 yr
G.M.	M	4½	4	Alive and well
A.B.	F	44	22	Radical removal performed 10 years after partial removal. Now aged 76—alive and well
R.C.	M	13	20 (dead)	Proven recurrence of tumour cause of death
B.H.	M	15	21 (dead)	Died of proven recurrence. Necropsy (Fig. 2) shows base of brain after removal from skull
D.K.	M	58	17	Died at age 73 yr

FIG. 1. Results of various methods of management.

recurrence was aspirated after 15 years, resulting in restoration of health. Figure 2 is a photograph of the base of the brain of one patient who died 21 years after radical excision of the tumour. The large recurrence is clearly visible.

Fifteen patients received irradiation at some stage in the management of the illness. One patient survives 29 years, one 17 years, three five years and one 18 months. Nine patients are dead.

FIG. 2. The base of the brain of case B.H. dying 21 years after radical removal of the tumour.

CASES NOT SUBJECTED TO SURGERY There were 11 patients in this group whose important details are summarized in Table 5. The long duration of symptoms requires emphasis: in three it exceeds 25 years.

DISCUSSION

The eighty-five patients were seen at the Radcliffe Infirmary during 32 years, in which period there were over 12,000 admissions to the Neurosurgical Department. Data obtained from the *Oxford Survey of Childhood Cancer* (Stewart, 1970—personal communication) reveal that there are approximately 15

TABLE 5

CASES NOT SUBJECTED TO SURGERY

Case	Sex	Age (yr)	Length of follow-up (yr)	Notes
E.S.	M	28	31	Practising veterinary surgeon
Dr.C.	M	57	13	Now aged 70—doctor living in retirement
M.B.	M	3	‡	Certified cause of death—cerebral tumour
D.G.	F	16	—	Untraced
D.W.	F	22	15	Married and well
A.S.	M	32	27	Died after gastrointestinal haemorrhage
A.C.	F	14	6	Certified cause of death—'Cerebral cyst'
D.R.	F	33	10	High grade mental defective since birth
G.J.	M	39	15	Diabetic—otherwise well
O.B.	M	8	27	Epileptic but able to work
A.S.	M	24	‡	Presented with advanced disease and dementia, died one month after air ventriculogram

deaths each year in children below the age of 15 years from craniopharyngioma in England and Wales. In the present series there were nine such children. By simple extrapolation there are about 150 new cases in England and Wales of *all* ages each year, not all of whom would require operation. This figure contrasts with that of astrocytomas which is about 25 times as large. These figures are gross approximations and are meant to show no more than the general magnitude of the problem.

Certain facts require emphasis: first, total removal, to judge from post-mortem specimens, is almost certainly impossible. Therefore, radical removal is probably a more accurate description. Figures 3 and 4 show the intimate relationship between the tumour and nervous tissue in one such specimen. That two patients in whom a total macroscopic removal was achieved have died of recurrence 20 and 21 years after the operation would support this view. A similar experience of recurrence after 'total excision' was reported by Gordy, Peet, and Khan (1949) where six out of 17 died of recurrence after an unspecified length of survival.

Secondly, in this series there are three patients who have been followed for periods greater than 25 years who have never been subjected to operation. This is in contrast to a number of patients whose total illness is measured in two or three years despite treatment. This serves to emphasize the great variability of the behaviour of these tumours and any claims for the success of radical surgery or radiotherapy must be assessed in the light of these observations.

PLACE OF RADIOTHERAPY One patient who received radiotherapy shortly after the primary surgery has lived for 29 years. He was irradiated at a time when

FIG. 3. An example of a craniopharyngioma in transverse section. Haematoxylin and Van Gieson, $\times 2$.

symptoms were recurrent, particularly headache. In this case it appears that irradiation had a significant effect. Kramer *et al.* (1968) report a series of six cases where irradiation was used as part of the primary treatment. All six cases are reported to be well between 13½ and 15 years later. In a survey of the cases seen at the Mayo Clinic (Love and Marshall, 1950), 10 cases were reported as experiencing remission or abatement of symptoms that could be attributed directly to radiotherapy. Tumour recurrence 20 years after surgery and the difficulties associated with radical removal would justify a long-term controlled trial to evaluate the precise part that surgery and radiotherapy should play in the management of these patients.

CASE FOR SPECIAL UNITS The endocrinological problems created by craniopharyngiomas and by attempts at radical excision (Tytus, Seltzer, and Khan, 1955; Kenny, Iturzaeta, Mintz, Drash, Garces, Susen, Askari, and Cowles, 1968; Matson and Crigler, 1969) both at the time of surgery and in the long term are formidable. The use of the operating microscope may make certain aspects of the surgery

FIG. 4. An enlargement of the area indicated by the arrow in Fig. 3 show the marked gliotic reaction adjacent to the epithelium. Haematoxylin and Van Gieson, $\times 20$.

safer; these factors, together with the rarity of the tumour, limiting any one person's experience, form a legitimate argument for the establishment of a few special centres for dealing with the new cases that occur each year.

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REFERENCES

- Erdheim, J. (1904). Über Hypophysenganggeschwülste und Hirncholesteatome. *S.-B. Akad. Wiss., wien. math.-nat. Kl.*, **113** abt III, 537-726.
- Gordy, P. D., Peet, M. M., and Khan, E. A. (1949). The surgery of craniopharyngiomas. *J. Neurosurg.*, **6**, 503-517.
- Grinker, R. R. (1943). *Neurology*, p. 602. Thomas: Springfield, Ill.
- Kenny, F. M., Iturzaeta, N. F., Mintz, D., Drash, A., Garces, L. Y., Susen, A., Askari, H. A., and Cowles, E. B. (1968). Iatrogenic hypopituitarism in craniopharyngioma: unexplained catch-up growth in three children. *J. Paediat.*, **72**, 766-775.
- Kramer, S., Southard, M., and Mansfield, C. M. (1968). Radiotherapy in the management of craniopharyngiomas. Further experiences and late results. *Amer. J. Roentgenol.*, **103**, 44-52.
- Love, J. G., and Marshall, T. M. (1950). Craniopharyngiomas (pituitary adamantinomas). *Surg. Gynec. Obstet.*, **90**, 591-601.
- Matson, D. D., and Crigler, J. F. (1969). Management of craniopharyngioma in childhood. *J. Neurosurg.*, **30**, 377-390.
- Ross Russell, R. W., and Pennybacker, J. B. (1961). Craniopharyngioma in the elderly. *J. Neurol. Neurosurg. Psychiat.*, **24**, 1-13.
- Tytus, J. S., Seltzer, H. S., and Khan, E. A. (1955). Cortisone as an aid in the surgical treatment of craniopharyngiomas. *J. Neurosurg.*, **12**, 555-564.