TUMOURS OF AND AROUND THE PITUITARY SEEN AT THE THOMSON ROAD GENERAL HOSPITAL BETWEEN JULY 1965 AND DECEMBER 1967

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Brain tumours are not uncommon in any community. This series of 21 patients were treated between July 1965 and December 1967. It is discussed because when diagnosed before certain irreversible sequences set in, the disease is worth-while treating for the following reasons:

(a) Vision can be saved

(b) USEFUL life is prolonged. After treatment the patients return to work and in most instances to their usual work. In early cases of certain tumours it is totally removable and therefore is a cure.

This series is broken down as follows:

Chromophobe adenoma	-	14 patients
Craniopharyngioma	-	4 patients
Suprasellar meningioma	-	2 patients
Optic glioma	-	1 patient

SEX AND AGE DISTRIBUTION

The sex and age distribution is as follows:

	Age Range	Sex Ratio
Chromophobe adenoma -	23 - 57 years (bulk between 35-50 yrs)	M : F 9 : 5
Craniopharyngioma	8 - 19 years	1 : 3
Suprasellar meningioma	41 & 43 years	Both F
Optic Glioma	4 years	M

There is no significant race distribution in any of these diseases.

SYMPTOMS AND SIGNS

(i) Visual Disturbances

The main presenting symptom is visual deterioration which if untreated leads to blindness in a few years. In this series twenty of the twenty-one patients presented with this complaint. The only exception who had a large extension of a chromophobe adenoma into the frontal lobe presented with generalised epilepsy. Although he had a typical bitemporal hemianopia he had not noticed any change in his vision. But his insight into his own illness was poor.

When first seen, two patients with chromphobe adenoma were completely blind. A third was blind in one eye and he could only make out shadows with the other one. Two other patients' vision was so poor that they only could see shadows with either eye. Three of the patients with craniopharyngioma were completely blind. The fourth one was blind in one eye but was able to count fingers at two feet with the remaining one. The two patients with suprasellar meningioma were blind in the left eye but could count fingers at three feet with the right one. The patient with the optic glioma was almost blind.

The disturbance of vision is variable and may be trifling. In one patient the complaint was that the two halves of the television screen seemed to have a difference in contrast. In another it was that he could not see the words on the edge of a page. In others it was that they could not read a sentence as the words seemed all jumbled up. Still others noted a closing—in of their vision and a tendency to knock into things on their outer sides. Others again described their vision as having a permanent mist over their eyes similar to that of using glasses lightly fogged over with moisture.

The visual field defects are dependent on the region of the chiasma on which the pressure is exerted. This may be on the body of the chiasma, the anterior chiasmal angle and one optic nerve or the posterior chiasmal angle and one optic tract in the above order of frequency. The common defects are the following:

(a) The bitemporal hemianopia which may be complete or quadrantic. Whether it is upper or lower quadrantic

will depend on whether the inferior or superior surface is compressed respectively.

(b) The scotoma, an interesting one of which is situated between the caecum and the macula and has variously been named bitemporal, paracentral or caecocentral depending on where the surgeon was trained. This interesting scotoma may be due to pressure on the posterior part of the body of the chiasma from below where the papillo-macular bundle of fibres lie. When the scotoma is pericentral in one field suggesting a nerve compression and an outer, upper quadrantic defect is found in the opposite field, the compression is on the anterior chiasmal angle.

(c) The defect of posterior chiasmal angle compression which involves the optic tract is homonymous and incongruous.

Papilloedema is a rare finding and it means a very large tumour which is obstructing the third ventricle and the foramina of Monro. This was observed in the patients who became extremely drowsy and disorientated before operation.

Optic atrophy in these patients is due to pressure on the chiasma and is not secondary to papilloedema.

(ii) Other Symptoms

The other symptoms and signs in this series are as follows:

(1) Disturbance of endocrine function.
  (a) Disturbance of sexual function
  (b) Disturbance of growth function
  (c) Disturbance of urinary concentration
  (d) Somnolence
(2) Headache
(3) Epilepsy
(4) Cerebrospinal fluid rhinorrhoea.

Disturbance of Endocrine Function

This is due to damage to the pituitary or the hypothalamus or both.

Disturbance of Sexual Function

This is observed in six patients with chromophobe adenoma, all the four with craniopharyngioma and one with a suprasellar meningioma. The signs are the following:

(a) Loss of secondary sexual characteristics which may be scanty or absent moustache and beard, scanty axillary and pubic hair, fine and smooth skin, a female distribution of fat and poor development of the genitalia.

(b) Loss of libido in the male

(c) Amenorrhoea or absent menarche.

Disturbance of Growth Function

This is manifested as follows:

(a) Dwarfism.
In this series one patient with chromophobe adenoma is short and fat. The adiposity is said not to be a growth disturbance but due to excessive intake of food. All the four patients with craniopharyngioma are dwarfs and they are emaciated as well.

(b) Gigantism or acromegaly.
This is a feature of acidophil tumours. There is none in this series.

Disturbance of Urinary Concentration

Diabetes insipidus is more common as a transient post-operative feature of hypophysectomy. One patient with craniopharyngioma passed nearly 5 litres of urine daily. The specific gravity was 1·004. This was probably due to a disturbance of the supraoptic nucleus of the hypothalamus and its tracts by compression.

Somnolence

Jefferson suggested that this was due to injury of the hypothalamic sleep centres. Three patients with chromophobe adenoma, one with suprasellar meningioma and all four with craniopharymgioma complained of this. The tumours in these patients were extremely large and have extended into the third ventricle partially obstructing the circulation of cerebrospinal fluid and compressing the hypothalamus. I have noted that the operative and postoperative management of these patients has been very difficult. One such patient died during operation and in others with this extension, in spite of more than adequate steroid cover the maintenance of their blood pressure was difficult. Bakay in discussing Olivecrona's 300 cases stated this, "... hypothalamic extension is of great prognostic

importance. Compression of the hypothalamus and invasion of the third ventricle frequently make the outcome of operation unfavourable . . . Some of the rare cases of papilloedena have been found in connection with these neoplasms . . . The postoperative mortality rate of this group was high (71·4%)."

The other signs of endocrine disturbance, such as an elevation of the blood sugar concentration, disturbance of basal metabolic rate, elevation of the blood pressure, disturbance of adrenal function and others have been described and are found more often with the acidophil and basophil tumours. None of these symptoms were noted in this series.

Headache

This is not a common feature and in most patients there is no headache. But when the mass is large enough to partially obstruct the circulation of cerebrospinal fluid the headache can be very severe and is due to hydrocephalus.

Epilepsy

The mass can extend forwards into the frontal lobe or laterally into the temporal lobe, the former being the more common. In each case the manifestation maybe epilepsy. One patient of this series presented with generalised epilepsy. His ventriculograms showed extension into the frontal lobe. Generally epilepsy indicates a large tumour.

Cerebrospinal Fluid Rhinorrhroea

One patient complained intially of this. This is caused by the mass eroding the sphenoidal sinus and ulceration of the mucosa of the nasopharyhx over this area. The danger of purulent meningitis is great. Sometimes before leakage of cerebrospinal fluid takes place, the patient complains of a mass in his nose and epistaxis takes place. Though the mass points mainly downwards it is also common for it to have a suprasellar extension large enough to cause a visual field defect. Another observation is that the cerebrospinal fluid leakage takes place usually after treatment by deep X-ray therapy.

Since the symptoms and signs are due to the way the mass extends and destroys the neighbouring structures there will be many more symptoms and signs that have not been discussed.

RADIOLOGY

Confirmation of the diagnosis is by radiology. Dr. Oon has very ably discussed this. The X-ray signs are found in the plain films of the pituitary fossa and the optic foramina, air studies and carotid arteriograms.

Neurosurgeons generally require bilateral carotid arteriograms because a large aneurysm of the internal carotid artery in this region can simulate a pituitary tumour. Useful information on the blood supply to the mass can also be obtained. Otherwise more information may be obtained from air studies.

TREATMENT

The aims of treatment have been discussed. The treatment is by surgery and radiotherapy together or by radiotherapy alone. Postoperatively the patient is put on replacement hormonal therapy for life.

INDICATIONS FOR SURGERY

The indication for surgery is deterioration of vision. The visual deterioration may be the following:

i) Recent rapid deterioration of vision.
ii) Visual deterioration during treatment with radiotherapy.
iii) Visual deterioration after adequate treatment with radiotherapy.
iv) No improvement during radiotherapy treatment which was started because of deterioration of vision.

Davidoff stated that the use of supervoltage radiotherapy has now made operation unnecessary in 85-90% of the cases.

Long standing blindness, or a large extension of the tumour into the third ventricle and compressing the hypothalmus, or an extension into the frontal lobe or into the nasal fossa are contraindications to surgery. In the former the blindness is permanent and in the latter the operative and postoperative mortality is prohibitively high.

OPERATIVE MORTALITY

The operative mortality of this series, in which all the tumours are extremely large, is 10%. This is a small series. Grant with a series of 148 cases had an operative mortality of 10%. Perhaps the most illuminating figures come from the 300 cases of Olivecrona. His overall operative

mortality was 11%. In discussing operative mortality all neurosurgeons stress this point. The operative mortality is low for small intrasellar growth and very high when there is extrasellar extension. Jefferson reported a mortality of 2% in the intrasellar group and 33% in the group with extrasellar extension. When Olivecrona's series was so divided it showed an operative mortality of 6% for the intrasellar group and 35% for the group with the extrasellar extension. Analysing the latter group further it was found that when there was "hypothalamic extension" the mortality was above 71%.

PROGNOSIS

The prognosis with regards to vision after successful surgical decompression is related to the nature of the patient's preoperative visual fields. When the patient has been blind for some time, it is extremely poor. But when the fields only show scotomata or the slope of the isoptres demarcating the temporal defect is gradual, the prognosis is excellent. When the slope of the isoptres is steep the prognosis is poor. Homonymous hemianopic and atypical fields indicate poor results because such fields generally mean a large extension of the tumour.

In this series 50% recovered full or almost full fields with a visual acuity between 6/6 to 6/12. In 25% the fields improved and the patients were left at least with bilateral full nasal fields and often with the inferior temporal quadrants as well and a visual acuity of 6/18 or better. In the next $12\frac{1}{2}$% the field deteriorated in one side but the other side had a full nasal field with a visual acuity here of 6/6 to 6/9. The compression here was on the posterior chiasmal angle and on one optic tract producing a homonymous hemianopic field before surgery.

In the last $12\frac{1}{2}$% the fields and visual acuity remained static. The compression here was due to large, calcified tumours which made safe decompression and removal very difficult.

SURVIVAL RATES

This series is too recent and too small to be useful in the discussion of survival rates and recurrence. However there has been no objective evidence, except in the case of the optic glioma, of any recurrence yet. The earlier cases have now survived 3 years. Olivecrona's figures were interesting. For his patients with chromophobe adenoma 75% were alive 2 years after operation, 70% 4 years, 71% 6 years, 66% 10 years and 55% 14 years. His recurrence rate was 5% and the first recurrence was 5 years after operation.

The problems of the surgeon are mainly due to the relation of the tumour to the circulation in this region. The fear of the surgeon is always that of uncontrollable bleeding from the circle of Willis and the vessels in the tumour capsule especially when the tumour has extended into the third ventricle. It must be remembered that the earlier the patient is sent for treatment the smaller would be the tumour and in certain cases like the early, small craniopharyngioma complete removal is possible.

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