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Pituitary Tumours in Kampala, Uganda

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Summary. A brief survey of all pituitary tumours seen over the past 3 years are presented. We have also reported in detail three cases whose clinical findings suggested the presence of a pituitary neoplasm, but this diagnosis had to be revised after operation when no tumour was found.

Résumé. Un bref relevé de toutes les tumeurs pituitaires observées en trois ans est présenté. Nous rapportons également en détail, 3 cas où l'observation clinique indiquait la présence d'un néoplasme pituitaire, diagnostic qui a dû être revu après une opération qui n'a révélé aucune tumeur.

Twenty-five pituitary tumours have been seen at the Neurosurgical Unit, Mulago Hospital, over the past 3 years, 1969–71. In addition, one case of cyst of the pituitary fossa and two cases of 'empty sella syndrome' were encountered. This compares with five tumours (three craniopharyngiomas and two adenomas) diagnosed between 1953 and 1965 (Billinghurst, 1966) and five (two craniopharyngiomas and three adenomas) between 1965 and 1969 (Table 1). Seventeen of these twenty-five cases were previously reported (Bailey & Thomas, 1971). During this same 3 year period, sixty-eight other primary intracranial tumours were discovered (Billinghurst & Bailey, 1973) so that the overall incidence of pituitary tumours as seen here was 36%.

TABLE 1. Pituitary tumours—Mulago Hospital

	Craniopharyngioma	Adenoma
1953–65	3	2
1965–69	2	3
1969–71	5	20

CRANIOPHARYNGIOMA

There were five cases of craniopharyngiomas, all in children. Headache and visual disturbance were the commonest presenting symptoms (Table 2). By the time they came to hospital,

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TABLE 2. Craniopharyngioma

	Case no.				
	1	2	3	4	5
Age	11	9	11	7	9
Sex	F	F	F	M	M
Clinical presentation					
Headache	+	+	+	+	
Vomiting				+	
Visual disturbance		+	+	+	+
Ataxia				+	
Drowsiness	+				
Deafness					+
Increase in weight	+				
Convulsion					+
Examination					
Visual acuity	6/6 6/6	PL PL	No PL HM	FC FC	No PL
Optic atrophy		+ +	+ +	− +	Pap +
BP	125/85	100/60	140/90	100/70	85/55
Hypothalamic	+				
Radiological					
Raised i/c pressure	+	+	+		+
Path. calcification	+	+	+	+	+
Management	(a) Transventricular aspiration of cyst and biopsy (b) Torkildsen's operation (c) III venticulostomy	(a) Craniotomy. Drainage of cyst and partial removal of tumour	Partial removal	Drainage of cyst. Fistula made between cyst and III ventricle	Aspiration of huge cyst. Biopsy of tumour
Result	Well after 18 months	Good recovery. Readmitted after 7 months with hydrocephalus. Died suddenly while awaiting drainage procedure	Died from pseudomonas meningitis	Well	Died after 6 days from bronchopneumonia
Type of tumour	Cystic	Cystic	Solid	Cystic	Cystic
Other findings	Prefixed chiasm		Prefixed chiasm		

PL, Perception of light; HM, hand movements; FC, finger counting; Pap, papilloedema.

TABLE 3. Pituitary adenoma

	Case no.																		
	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20
Sex	F	M	M	F	F	F	F	F	M	M	F	M	M	M	M	F	F	M	M	M
Presenting symptoms																				
Headache	+	+		+	+		+	+	+		+	+	+	+	+			+	+	+
Visual disturbance	+	+	+	+	+		+	+	+	+	+	+	+	+	+	+	+	+	+	+
Acromegalic features		+				+										+				+
Hypituitarism		+		+	+	+	+				+					+				
Dementia										+			+							
Clinical signs																				
Acromegaly		+	+		+	+	+				+					+		+		+
Visual acuity (R)	6/18	?	–	4/4	6/24	6/9	6/6	6/12	–	–	FC	–	–	HM	–	6/9	?	6/6	HM	
Visual acuity (L)	FC	–	PL	PL	6/24	6/9	6/36	–	–	–	–	6/12	6/24	PL	–	6/6	?	6/9	6/36	
Optic atrophy	+	+	+	+		–	+	+	+	+	+	+	+	+	+	+		–	+	
Visual field defect	BT	?	?	N	?	N	BT	T	?	?	T	T	T	?	?	BT	?	BT	?	
Treatment by operation	+	R	+	+	+	+		+	R	+	+	R	+		R			R	R	

FC, Finger counting; HM, hand movements; PL, perception of light; BT, bitemporal hemianopia; T, temporal hemianopia in one eye; ?, visual fields could not be tested; R, refused treatment.

most of the children had serious visual defects with acuity reduced to finger counting or worse. The one girl with normal vision had a severe hypothalamic disturbance.

Skull X-rays showed evidence of long-standing raised intracranial pressure and all five had pathological suprasellar calcification. Full investigations, including air encephalography, were carried out on all patients who were subsequently operated upon. In four cases where the tumour was cystic, the cyst was emptied and part of the cyst wall removed. In one, a large fistula was created between the third ventricle and the cyst. Two later developed hydrocephalus. In one, a good result followed third ventriculostomy after a ventriculocisternal shunt (Torkildsen's operation) had failed. The other child died suddenly while awaiting a drainage procedure.

PITUITARY ADENOMA

In this group there were eleven men and nine women, average age 40 years. A deterioration of vision was the largest single factor that brought the patient to hospital, and was complained of by all except one patient who was referred by a psychiatrist because of acromegaly (Table 3). A delay in recognizing the symptoms of pituitary tumour, especially headache and hypopituitarism in women, until the onset of severe visual disturbance meant that most of the tumours were of large size. This may stem from the still widely held belief that intracranial tumours are very rare in tropical Africa. The seriousness of the visual defects is shown in Table 3.

Acromegaly was recognizable in nine cases. Four of these were operated upon, two refused operation and three were treated conservatively. In the absence of a radiotherapy unit, it has been our policy in the last 2 years to recommend operative treatment of acromegaly, even if there is no evidence of optic chiasm compression, as it is currently believed that early treatment can lead to some reversal of the acromegalic process probably by reducing the serum level of growth hormone. Our numbers are too few and follow-up too short in comment on this policy as yet.

Six of the eleven patients without acromegaly had an intracapsular removal of a pituitary tumour. Histologically all were chromophobe adenoma. Four others refused operation and one died shortly after admission in a state of shock (hypopituitary shock?).

It has not been easy to follow-up these patients as many live several hundred miles away and their financial circumstances do not always allow them to attend for review. We have therefore only meagre information about our long-term results.

In the presence of advanced visual deterioration and optic atrophy it is very difficult to improve the vision to a useful level. In a couple of cases, however, considerable early improvement has occurred. One very demented, blind man who just lay in bed and had developed bedsores improved sufficiently after operation to lead an almost normal life, albeit blind. These incidences of spasmodic success in seemingly hopelessly advanced cases has prompted us to seriously consider all patients as potential candidates for surgery. We must continue to emphasize that these benign tumours must be treated at an early stage when it is possible to achieve an 80% improvement in the visual status by operation (Ray & Patterson, 1971). Nothing is more disappointing to the surgeon than the patient's refusal to accept treatment for a condition that is curable. The high level of refusal to undergo operation for a pituitary tumour shows that we also have to educate the public to show that craniotomy is no longer a lethal procedure.

CASES SIMULATING PITUITARY TUMOUR

Case 1

This 38-year-old man presented in the Ophthalmology Clinic with a short history of visual deterioration. His corrected vision was then 6/4(R) and 6/24(L). The right visual field was full, but on the left, there was a centro-caecal scotoma. The left optic fundus was thought to be pale. Two months later, his vision had deteriorated to 6/18(R) and 6/24(L) and the left optic disc was now definitely pale. X-rays of the skull showed an enlarged, eroded pituitary fossa (Fig. 1). He had no complaint of headache, no loss of libido and no previous history of head injury or meningitis. Clinical examination revealed no other abnormality except mild essential hypertension (BP 180/120–160/100). The cardiovascular system was normal on examination as was the ECG. On two occasions, at lumbar air encephalography, air failed to enter the ventricles. As the clinical and radiological picture seemed to point to an intrasellar tumour with early optic compression, it was decided to carry out a surgical exploration.

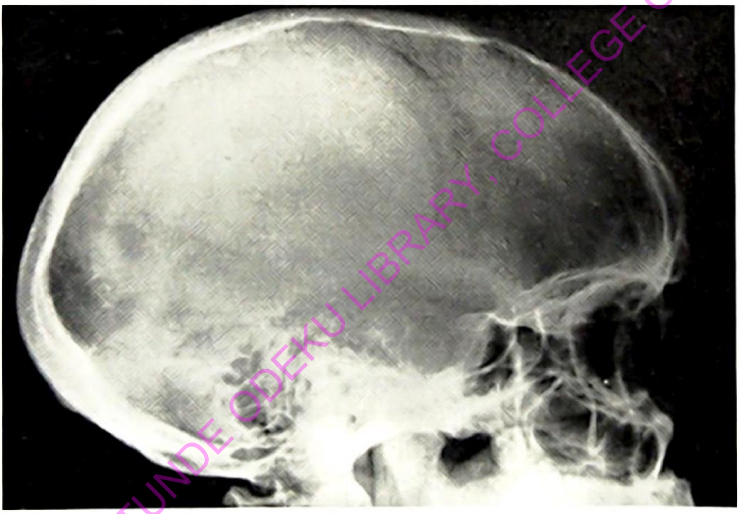

FIG. 1. Arachnoid cyst of the sella turcica. Plain X-rays show an enlarged, eroded pituitary fossa.

A sub-frontal approach to the pituitary fossa was made. There was a liberal amount of CSF in the subarachnoid space (protein 20 mg/100 ml, WBC $< 5/mm^3$). The right optic nerve looked normal, but the arachnoid over the chiasmatic cistern was thickened and opaque. A small piece of the arachnoid was stripped away and under it we found a cyst with a transparent, avascular capsule that lay in the sellar cavity. The cyst fluid was mildly xanthochromic (protein 5000 mg/100 ml, laboratory result—almost certainly too high. There was insufficient fluid to repeat the estimation, WBC < 5). After removal of the cyst fluid we found an empty sella turcica with many small blood vessels traversing the floor. The pituitary fossa was found to hold 10 ml of saline. The left optic nerve looked a little pale, but was otherwise normal.

Within 5 days of the operation the centro-caecal scotoma had disappeared. Eighteen

months after operation, visual acuity was J1,6/6 in each eye and the patient was symptom-free. Histological examination of the thickened arachnoid did not reveal anything specific.

Comment

We diagnosed this as an arachnoid cyst of the sella turcica.

Arachnoid cysts may be either congenital or acquired. The congenital variety are inter-arachnoid in position and probably arise by the passage of CSF into the perimedullary mesenchyme in foetal life, that is, a derangement in the development of the leptomeninges (Starkman, Brown & Linell, 1958). Acquired cysts, post-traumatic and post-inflammatory, lie in the subarachnoid space and become loculated as the result of adhesions. Without a definite history of trauma or infection, one can only postulate on the origin of the cysts when seen at operation. One only gets the opportunity at autopsy to be certain of the exact site of the cyst in relation to the meninges.

Cerebral arachnoid cysts are usually found either in the posterior fossa or in the Sylvian fissure and rarely occur in the sella turcica (Ring & Waddington, 1966; Robertson, 1954). Other cysts with which they may be confused include Rathke's pouch cysts and cystic pituitary adenomata (Paterson, 1948). Some cases described radiologically as arachnoid cysts are no more than dilated subarachnoid cisterns projecting into the sella turcica (Kaufmann, 1968).

Case 2

This 45-year-old man was referred from Dar-es-Salaam with a 15 year history of acromegaly and an 8 year history of hypertension and diabetes. For 3 years he had complained of headaches and over the past year had complained of loss of libido and failing vision.

Examination. The typical features of acromegaly were obvious. He had moderate hypertension, the BP varied from 170/120 to 140/95. The only neurological signs were a reduction of visual acuity to 6/24(R) and 6/18(L). The visual fields were full.

X-rays of the skull showed an enlarged pituitary fossa, and hypertrophied frontal air sinuses. Cardiac enlargement consistent with hypertension was seen on chest X-ray while a film of the hands demonstrated the typical tufting of the terminal phalanges found with acromegaly. It was technically impossible to lumbar puncture him and therefore AEG could not be carried out. Metabolic investigation including 17–ketosteroids (10·5 mg/24 hr), electrolytes, urea and cholesterol were all within normal limits.

His hypertension and diabetes were controlled with aldomet and insulin and under steroid cover, a sub-frontal exploration of the pituitary fossa was undertaken. The sellar diaphragm was a thin membrane and on opening this, the sella turcica was seen to be completely empty; there was no sign of any pituitary gland.

Comment

We considered that this was a case of burnt-out acromegaly and that the empty sella was the result of pituitary necrosis.

Case 3

This 30-year-old woman presented with a 6 month history of frontal headaches, rapidly progressive visual deterioration and impairment of memory. She also complained of feeling

cold, loss of energy, drooling saliva from her mouth and vomiting which had occurred daily for 2 months.

She had been married for 5 years and had one 4-year-old child. Amenorrhoea had been present for 5 months without other signs of pregnancy. There was no previous history of ill health.

Examination. She was slow and apathetic in speech and action. There was no dysphasia. There were stigmata of epiloia on her face and vague signs of myxoedaema such as cold extremities and sparse eyebrows. There was no goitre. The cardiac rhythm was irregular; BP 110/80; ECG was normal.

Visual acuity was reduced to finger counting on each side. Fundal examination showed pale discs especially on the temporal sides. She had an incongruous bitemporal hemianopia. There were no other neurological abnormalities.

Radiological examination of the skull showed an enlarged eroded pituitary fossa with a speck of calcification in the upper posterior part of the sella (Fig. 2). Chest X-rays were normal. Haematological investigations (included: haemoglobin 10·2g/100 ml, PCV 34. MCHC 30, WBC 5600/mm^3, serum cholesterol 182 mg/100 ml, fasting blood sugar 108 mg/100 ml, urea 14 mg/100 ml, electrolytes—normal, PPR—negative) added little to the clinical picture.

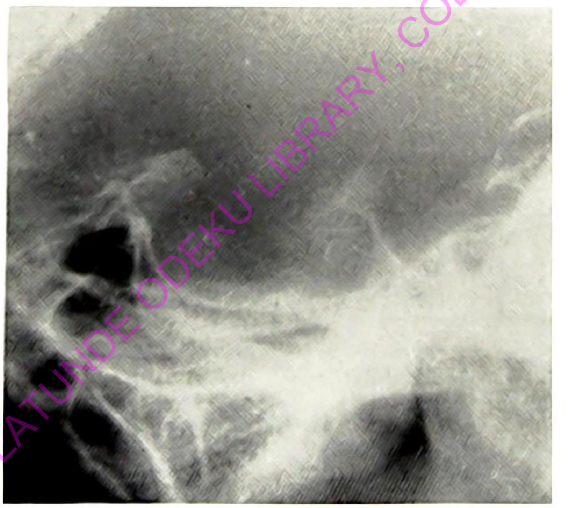

FIG. 2. Empty sella syndrome. Plain X-rays show an eroded pituitary fossa with a speck of calcification.

In view of the rapid deterioration in visual acuity and the seemingly obvious diagnosis of pituitary tumour, we decided to omit air studied and proceed to an emergency exploration.

A right sub-frontal approach was made to the pituitary area. The brain tension was slack after removing 15 ml CSF from the right frontal horn of the lateral ventricle. There was a copious flow of subarachnoid CSF. There were numerous fine avascular adhesions around the right optic nerve. The left optic nerve looked normal, and the chiasm was pre-

fixed. There was no pituitary enlargement and no chiasmal compression, in fact the sella looked empty.

The ventricular CSF was reported to contain 350 mg protein but no cells. Post-operative air encephalography showed a mild hydrocephalus but the air in the cisterns did not enter the pituitary fossa.

Comment

No firm diagnosis was made. The enlarged pituitary fossa was typical of a pituitary tumour. The intrasellar calcification may have represented a necrotic gland. The avascular adhesions were not considered significant at the time of operation but in retrospect may have indicated a former arachnoiditis. The mild hydrocephalus would support this. There were no other signs of sarcoidosis. Although small calcium deposits are seen on skull X-rays with tuberose sclerosis, we could find no reference to their appearance inside the pituitary fossa (Golden, 1964).

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REFERENCES

BAILEY, I.C. & THOMAS, J.D. (1971) Pituitary tumours in Uganda. *E. Afr. med. J.* **48,** 90–99.

BILLINGHURST, J.R. (1966) Intra-cranial space occupying lesions in African patients at Mulago Hospital, Kampala. *E. Afr. med. J.* **43,** 385–393.

BILLINGHURST, J.R. & BAILEY, I.C. (1973) Primary CNS tumours in Uganda. *Afr. J. Med. Sci.* **4,** October (in press).

GOLDEN, R. (1964) *Diagnostic Roentgenology*, Vol. I, pp. 1206–7. Williams and Wilkins, Baltimore.

KAUFMANN, B. (1968) The 'empty' sella turcica. A manifestation of the intra-sellar sub-arachnoid space. *Radiology*, **90,** 931–941.

PATERSON, J.E. (1948) Cystic pituitary adenomata. *J. Neurol. Neurosurg. Psychiat.* **11,** 280–287.

RAY, B.S. & PATTERSON, R.H. (1971) Surgical experience with chromophobe adenomas of the pituitary gland. *J. Neurosurg.* **34,** 726–729.

RING, B.A. & WADDINGTON, M. (1966) Primary arachnoid cysts of the sella turcica. *Amer. J. Roentgenol.* **98,** 611–615.

ROBERTSON, E.G. (1954) *Pneumoencephalography*, pp. 230–232. Charles C. Thomas, Springfield, Ill.

STARKMAN, S.P., BROWN, T.C. & LINELL, E.A. (1958) Cerebral arachnoid cysts. *J. Neuropath. exp. Neurol.* **17,** 484–500.