Death from neglected pituitary adenoma, a case report.

ABSTRACT: We present a case of a patient who presented with pituitary macroadenoma complicated with hydochephalus.He presented late to the referral hospital in a comatose state and died a few hours later.He was a 33-year-old,male who gave a history of highly accelerated symptoms lasting 1 week.Neurosurgical consultation as well as investigations with CT scan or MRI were not availed the patient,which could have been life saving. Autopsy findings were of a pituitary macroadenoma and signs of hydrocephalus and raised intracranial pressure which proved fatal.

Keywords: *Pituitary,adenoma,hydrocephalus*.

INTRODUCTION

Pituitary adenomas (PA) are intracranial tumours that may result in death(1). Autopsy studies suggest that microadenomas(<10mm) occur in 10 to 20% of the population (2, 3), while macroadenomas >10mm are quite rare(4, 5).PA commonly produce ocular,or endocrine symptoms (6), others symptoms relate to mass effect as well as obstruction to cerebrospinal fluid(CSF) flow.Giant PA >/=4cm have been reported(7-10).PA occur mostly in adults, but a case in infancy resulting in sudden death was reported(11).Equally occurrence in paediatric age range as well as adolescents(12,13) are reported.PA account for 10% of intracranial neoplasms,and most these arise from the adenohypophysis, other sites are involved by extension, infiltration or ectopic location(14,15).PA present a challenge to the clinician in determining the clinical significance, appropriate diagnosis, need for treatment and type of therapies/ therapies(16).PA may be functional(FA) or non-functional(NFA).Functional adenoma may produce a single hormone such as growth hormone(GH) secreting somatostatinomas,prolactin(PRL)prolactinomas, adrenocorticotrophic hormone(ACTH)corticotrophinomas,thyroid stimulating hormone(TSH) thyrotropinomas (17,18) and other rare hormone products(19) or mixed hormones(20).Clinical presentations may range from acromegaly or gigantism(21),Cushing's syndrome(22), Nelson syndrome, hyperprolactinaemia, and other rare endocrine syndromes.Most cases are sporadic, but some occur as part of multiple endocrine neoplasia1 (23,24),or other inheritable tumour syndromes(25),and may occur with other tumours(26,27).

Mortality in the NFA adenoma is commonly due to hormone deficiency , surgery, and mass effect as they often grow very large before detection(28,29).In one series patient died of circulatory complications, respiratory complications as well as infections(28), and in another series patients suffered occult hydrocephalus, and they died of complications following repeated interventions(30).The common presentation of NFA remain, headaches visual disturbance and the hormonal deficiencies(31).Among the functioning(FA),mortality from vascular disease is the predominant cause of death(28). In ACTH producing adenomas, control of cortisol and growth hormone secretion limits the cardiovascular risk(28).Pituitary apoplexy is a sudden vascular event in a PA that can also be life threatening(32,33), and has been reported to induce severe intracerebral haemorrhage(34,35), and death(36).Cases of hydrocephalus complicating PA were successfully treated through neurosurgical shunt procedures(37,38).In our environment an interplay of sociocultural and adverse economic factors frequently stand in the way of clinical care leading to disastrous consequences for patients. It is common to see patients consult traditional medicine practitioners due to cultural and economic considerations, only to appear orthodox medical care when it is very late.

CASE REPORT:

We present the case of a 33-year-old man with a history of headache of seven days duration.
Blurring of vision of 2 day,seizures which were intermittent and unconsciousness of a day's duration.
The headache was of sudden onset, noticed on the right temporal area and later became generalized. Described as the worse headache he has ever suffered. No known aggravating or relieving factor. Pain was said to radiate to the neck. Patient was initially ambulant but later could not undertake normal activities and could not sleep. There was no fever or associated but vomited repeatedly. The vomitus was non projectile, on bilious and comprised of recently ingested feeds.

He then developed tonic clonic seizure with an episode lasting 2 minutes, terminating in a cry and urinary incontinence.
Patient lapsed into unconsciousness on the way from the point of first consultation to the referral centre in the teaching hospital.
His past medical history was not contributory.
He was not known to indulge in alcohol or tobacco use. There was no previous history of hypertensive or diabetes mellitus, and had not recently travelled to the meningitis belt of the country.
Physical examination revealed a young man gasping for breath, was on oxygen at the time of reception at the referral hospital.
CNS EXAMINATION: He was unconscious with a Glasgow coma scale of 3/15 distributed as spontaneous eye movement(E1), verbal response(V1), motor response(M1).
Pupils were 5cm dilated and fixed, non-reactive to light (direct and consensual).
The neck was supple, Kernig's and Brudzinski signs were negative.
Cranial nerves 11, 111,1V, V11,1X,X were non responsive.
Muscle tone was reduced in all the limbs. Muscle power could not be assessed objectively.
There was generalized depression in limbs reflexes, both plantar and flexor.
RESPIRATORY SYSTEM:
The respiratory rate RR: was 42 cycles per minute and was gasping. The chest was resonant and breath sounds were vesicular.
CARDIOVASCULAR SYSTEM:Pulse rate was 140 bpm,small volume .The apex beat, was at the normal position, jugular venous pressure was not raised.
The blood pressure progressively fell from 120/60 mmHg on admission at 9am to being unrecordable at 12 noon.
Examination of the other organ systems did not reveal any significant find.
PRESUMPTIVE DIAGNOSIS:

(1) Subarachnoid haemorrhage secondary to ruptured aneurism.
(2) Space occupying lesion (Glioma)

Patient was admitted into the intensive care unit under the neurology unit.
INVESTIGATIONS AND RESULTS.

(1) Random blood sugar RBS 12.4mmol/l.
(2) Urinalysis was normal.
(3) HIV screening was Negative
(4) Complete blood count: Total wbc 5x 10/dl, Lymphocytes 47%,Neu.50%,eos.1% Platelets; in normal range.
(5) Serum electrolytes: Sodium:130mm/l, Potassium.3.3 mm/l calcium, magnesium and creatinine were being processed before patients' demise.
(6) Skull X ray was ordered but was not done because the patient was very unstable.
(7) Cranial CAT scan and MRI were, ordered but were not done before patient passed.

TREATMENT:

(1) 20 % Mannitol 250 ml over 4 hours, every 8 hrs. for 72 hrs.
(2) Normal saline 1 litre 8 hourly for 72 hours.
(3) Intravenous frusemide 20 litres after each dose of mannitol.
(4) Monitoring of vital signs every 30 minutes.
(5) Strict monitoring of fluid input and output.

OUTCOME: Patients died within two hours. Autopsy was ordered for.
AUTOPSY FINDINGS:
Patient was generally well nourished looking, male,1.62m in height and weighing 72kg.The pupil were dilated and fixed. Other physical examinations carried out were found to be in the normal range.
The main autopsy findings were in the central nervous system (CNS) and the bones surrounding the sella tursica. The brain weighed 1350g, The gyri were flattened bilaterally and the sulci narrowed. There was bilateral uncal and cerebellar tonsillar grooving. A large pituitary tumour measuring 3.4 x2 x 2 cm was found completely occluding the 3rd ventricle and projecting into the lateral ventricles. The lateral ventricles were dilated on both sides, the left measuring 12 cm long and 8 cm in the widest portion. The right measured 12 cm long and 7cm in the widest portion.The bones of the sella tursica were significantly

eroded, but the tumour was still covered by the diaphragma sella and the carvenous sinus had not been eroded into.

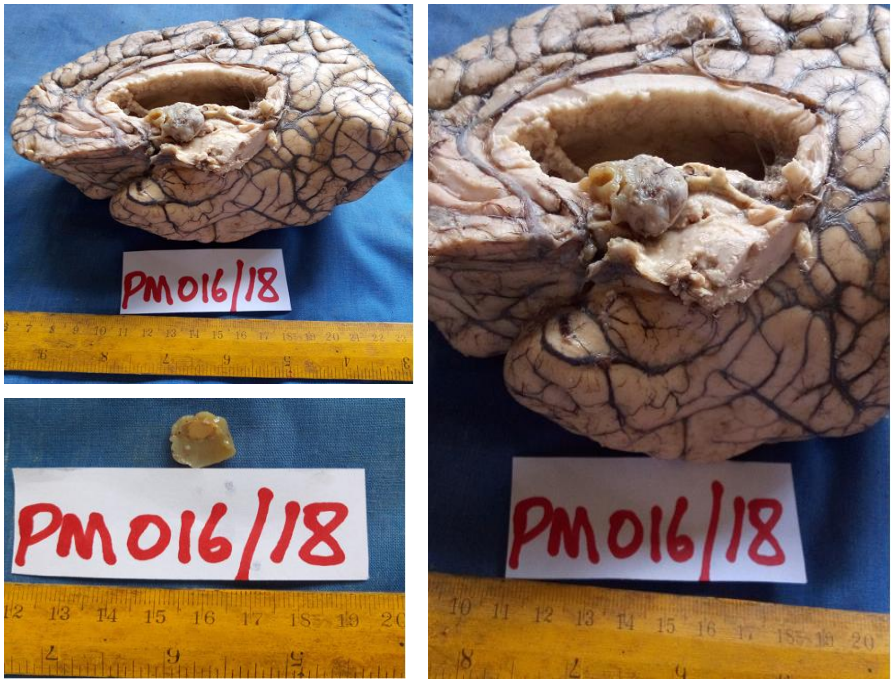

Figure 1: Left cerebral hemisphere showing a pituitary adenoma and markedly enlarged lateral ventricle.

A diagnosis of Pituitary adenoma causing Hydocephalus was made.
The cause of death was concluded to be coning due to hydrocephalus, which in turn was induced by a pituitary adenoma.

DISCUSSION:

Although the patient history suggested a symptom duration of 1 week, the findings at autopsy suggested otherwise. It is very likely that the pituitary adenoma had been for a while inducing symptoms which were not heeded to until it became too late. Adenomas that grow very large tend to be the non-functional types(6,28) in most cases, although one series in the United States reported 5 giant prolactinomas in (7).Patients with the functional adenomas normally seek medical attention quicker because of the endocrine symptoms they induce (16).Our index patient had no hormonal assay or immunohistochemistry to determine if the PA was functional or not. There was however raised random blood sugar which may suggest hypercortisolism, or even raised thyroid hormones. It is also important to state that the types of resources needed to save this patient are currently not in our centre. Early presentation would still have been beneficial to the patient because our hospital operates a robust referral system which would have been activated to save the patient's life. The lessons to learn in this case is the need for early hospital consultation when seemingly ordinary symptoms such as persistent headaches, blurring of vision are encountered. In our country the alternative medical practitioners often act as a barrier between patients and orthodox medical consultations, often with disastrous consequences.

CONCLUSION:This case highlights the common practise of patents arriving late for hospital treatment.Although the patient would have still had a tough time accessing the kind of care that will bring relief in this case.Government should intervene to change the circumstances that will improve patients welfare

CONSENT: Patients personal data was anonymised and permission for the use of specimen photographs was granted by the Department of Pathology, University of Calabar Teaching hospital.

ETHICAL APPROVAL: There were no ethical issues in this case and permission to report this case was granted by the institutional review board,

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