"THE CLINICAL AND RADIOLOGICAL FEATURES OF CRANIOPHARYNGIOMA AS SEEN IN OUR LOCAL POPULATION AT THE KENYATTA NATIONAL HOSPITAL, KENYA.

A dissertation submitted in part-fulfilment for the degree of MASTER OF MEDICINE (RADIODIAGNOSIS) of the University of Nairobi, Kenya.

by

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DECLARATION

This dissertation is my original work and has not been submitted for a degree in any other University.

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REVIEW OF CRANIOPHARYNGIOMA AT K.N.H.

(8 YEAR PERIOD, 1973-1981)

SUMMARY

The main clinical and radiological features of craniopharyngioma as seen in our local population are presented. A total of 31 cases was studied. The peak age incidence was found to be 6-10 years. The male to female ratio was 1:1. Headache and visual loss were the commonest presenting symptoms (61.1% and 59.1% respectively). Visual field defect was the commonest clinical finding. Other clinical findings were those of pituitary hypofunction. Calcification in the supra- or infra- sellar region was the commonest abnormal radiological finding on plain skull radiographs (51%). Erosion with expansion of sella turcica was found to be (33%) while (21%) of cases showed sutural diastasis - carotid angiography revealed elevation of proximal portions of anterior cerebral arteries in 50% of patients. No shift of anterior cerebral artery from the midline was noted. Ventriculography showed dilatation of lateral ventricles in all patients who were subjected to the examination. Most patients studied were Kikuyus. The minority were spread among Luos, Kisiis and Kalenjins.

INTRODUCTION

Craniopharyngioma is one of the pituitary tumours and accounts for approximately 4% of all intra-cranial neoplasms and about 30% of all pituitary tumours (1). A variety of names has been used in the literature to describe these tumours: Rathke's Pouch Tumour, Craniobuccal Cysts, Supra-sellar Cysts, Simple Squamous Epithreliomas, Adamantinomas, Ameloblastomas, Hypophyseal Duct Tumours and Craniopharyngioma. Craniopharyngioma is the name used throughout this study. Craniopharyngioma is widely accepted to be of developmental in origin, arising from the remnants of Rathke's Pouch in the region of pituitary. Embryologically the pituitary gland is a composite organ formed from the neural and foregut elements. The latter gives rise to glandular part of adeno-hypophysis comprising the pars distalis 'Anterior lobe), pars intermedia and pars tuberalis. At about the age of four weeks the embryo develops a pouch from the stomadeum known as Rathke's pouch and this begins to extend upwards to meet a downward growth from the floor of the Diencephalon, which occupies a position immediately behind the pouch of the mouth. The orifice of Rathke's pouch gradually contracts and along slender duct is formed which becomes solid. This is called craniopharyngioma duct or pituitary stalk. The anterior wall of Rathke's pouch

becomes anterior lobe of pituitary and the neural pouch becomes the posterior lobe of pituitary. Remnants of the original cavity of Rathke's pouch often persist in a form of a cleft or cyst space lying between the pars distalis and posterior lobe. Cells lining this space - the so called rest cells are widely held to be the origin of craniopharyngioma.

The pituitary gland lies within the sella turcica of sphenoid bone and is separated from the rest of the cranial cavity by a fold of dura - mater called the diaphragm sellae, through which passes the stalk which connects the pituitary to the hypothalamus. The gland is related superiorly with optic chiasma laterally on each side by cavernous sinuses in which the internal carotid arteries lie. The 3rd, 4th, 6th nerves and the maxillary and ophthalmic branches of trigeminal nerve also lie in the cavernous sinuses. Thus a tumour growing within this region can present with a variety of symptoms. Clinical and radiological abnormalities including pituitary hypofunction, optic nerve involvement and raised intra-cranial pressure. Involvement of hypothalamus may lead to disturbances of temperature regulation and diabetes insipidus.

Craniopharyngioma as reported is predominantly tumours of childhood with onset of symptoms before the age of 15 years, but symptoms may have their onset at

any age. This study has been undertaken to find out the incidence of the main clinical and radiological features of craniopharyngioma of patients who attend the Kenyatta National Hospital. An attempt has been made to find out the distribution of the disease according to age, sex and tribe. The differential diagnoses of the disease are discussed.

MATERIALS AND METHODS

All files of patients admitted to the Kenyatta National Hospital with a principal diagnosis of craniopharyngioma were studied. The International Coding System of Diseases was used to get the files. The system has been in operation in this hospital since 1972. Only patients with confirmed histology of craniopharyngioma were analysed. The period covered was 1973 - 1981. A total number of 33 cases was studied, but only 31 cases were suitable for clinical and radiological analysis. The analysis was done with a view to extract information as regards the main presenting symptoms, main clinical findings on examination; the type of radiological investigation and results. The Roentgen results were either extracted from X-ray request form as per radiologist report or from the files if the X-rays were untraceable. The age, sex and tribal distribution were analysed.

RESULTS

AGE:

The peak age incidence was found to be 6-10 years (32.2%). The youngest patient was 6 months and the oldest in this series was 28 years. Julian and Hoffman have described craniopharyngioma in a child of one week (2). Our distribution according to age is tabulated in Table I. These figures confirm the findings of other authors that the majority of these tumours occur in childhood. 79% of our patients were below the age of 15 years. No patients were in the 3rd, 4th or 5th decade in this series. Table II shows distribution according to age as per Barnet study in 1932-1952.

TABLE I

AGE DISTRIBUTION OF CRANIOPHARYNGIOMA

31 PATIENTS

AGE GROUP IN YEARS	NUMBER OF CASES	%
0 - 5	6	19.3
6 - 10	10	32.2
11 - 15	9	27
16 - 20	3	9
21 - 25	1	3
26 - 30	2	6.4

HISTOGRAM SHOWING AGE DISTRIBUTION OF CRANIOPHARYNGIOMA

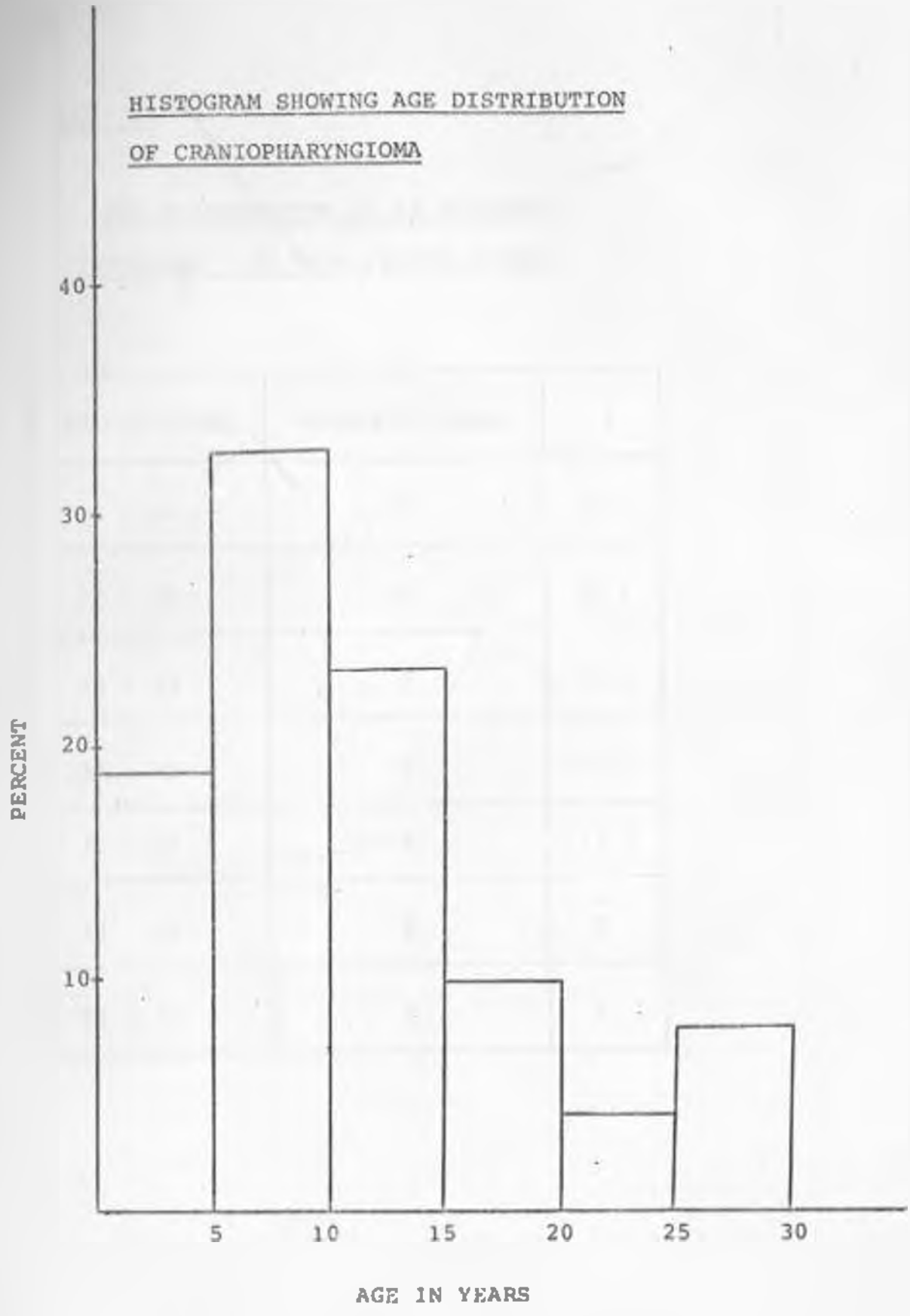


TABLE II

AGE DISTRIBUTION OF 62 PATIENTS

(BARNET - 20 YEAR PERIOD STUDY)

AGE IN YEARS	NUMBER OF CASES	%
0 - 10	10	19.2
11 - 20	18	29.1
21 - 30	9	19.1
31 - 40	7	11.1
41 - 50	8	11.5
51 - 60	4	5
61 - 70	4	5

SEX:

There were 15 females and 16 males. Most writers agree with my series in that the ratio of male to female regarding the incidence of craniopharyngioma is 1:1.

TRIBAL DISTRIBUTION:

Majority of the patients were Kikuyus. This can be explained by the fact that the hospital is within reach of Kikuyus who are the majority residents in and around Nairobi. The minority were spread among Luos; Kisii and Kalenjins.

SYMPTOMS AND SIGNS:

The main symptoms were those secondary to increased intra-cranial pressure and disturbance of visual pathways. 61.1% presented with headache as their main symptom while 59.1% presented with visual problems. Other symptoms elicited included vomiting, polydipsia, excessive sleep and convulsions. Two patients volunteered history of loss of libido. Table III shows distribution of symptoms.

TABLE III

FREQUENCY OF SYMPTOMS

SYMPTOMS	NUMBER OF CASES	%
Headache	19	61.1
Decreased Vision	18	59.1
Vomiting	2	6
Polydipsia + Polyuria	2	6
Excessive Sleep	2	6
Loss of Sexual Libido	2	6

DURATION OF SYMPTOMS:

The duration of symptoms was variable from patient to patient. In my series the period between the appearance of first symptoms ranged from 6 months to 30 months. Olive Crona series report range from 20 days to 31 years; while Love and Marshals the range is from 1 month to 31 years.

SIGNS:

The signs elicited were those due to pressure on optic chiasma, increased intra-cranial pressure and pituitary hypofunction. The commonest clinical findings in my series were those of visual field defect and optic atrophy (58%); and decreased visual acuity (45%). The field defect was that mainly of bitemporal hemianopia. Optic atrophy was a more common finding than papilloedema. The signs of pituitary hypofunction were those of growth disturbance and hypogonadism. Impaired growth was estimated on visual impression, comparing the age and the height of the patients: No bony age or growth hormone assay was done in any of these cases.

The gonadal deficiency was recognised by hypoplasia of sex organs and lack of secondary characteristics. In 2 patients both penis and testicles were hypoplastic, while in one female there were no breast visible at the age of 18 years.

TABLE IV

DISTRIBUTION OF CLINICAL FEATURES

SIGNS	NUMBER OF CASES	%
Decreased Visual Acuity	15	45
Visual Field Defect	18	58
Optic Atrophy	18	58
Papilloedema	2	6.4
Blindness Both Eyes	0	0
Blindness In One Eye	2	6.4
Stunted Growth	3	9.4
Hypogonadism	3	9.6

RADIOLOGICAL FINDINGS

All 31 patients had plain skull radiographs. Eleven patients were subjected to contrast neuro-radiology - study. Eight had carotid angiography and three had ventriculography. Pneumoencephalography was not done in any of the cases in my series.

RESULT OF PLAIN SKULL RADIOGRAPHY

The commonest radiological abnormality in plain radiography was calcification in either supra- or infra-sellar region (51%). Sutural diastasis was seen in 21% while erosion with expansion of pituitary fossa was noted in 30%.

TABLE V

RADIOLOGICAL ABNORMALITY	NUMBER OF CASES	%
Sutural Diastasis	7	21
Erosion With Expansion Of Pituitary Fossa	10	33
Calcification In Or Around Sella Turcica	17	51
Normal Skull	6	8

Calcification in the supra- or infra-sellar region was mainly noted in 11 - 15 age group; while as would be expected sutural diastasis was seen more in 0 - 5 years age group. Calcification varied in type from few punctate dots to a large densely calcified mass. As expected majority of cases of calcification was in the midline and just above the sella. 14 patients had supra-sella calcification while 3 patients showed calcification with the sella.

TABLE VI

DISTRIBUTION OF RADIOLOGICAL FINDINGS ACCORDING TO AGE

AGE GROUP IN YEARS	SUTURAL DIASTASIS	CALCI-FICATION	EROSION OR EXPANSION OF PITUITARY FOSSA	NORMAL SKULL
0 - 5	4	-	1	1
6 - 10	4	3	4	1
11 - 15	2	6	2	1
16 - 20	-	1	1	2
Over 20	-	-	1	-

There was no significant difference in the distribution of the radiological appearances between females and males.

RESULTS OF CAROTID ARTERIOGRAPHY

Patients who had carotid angiography were eight. Bilateral or single sided injection with cross compression was done.

TABLE VII

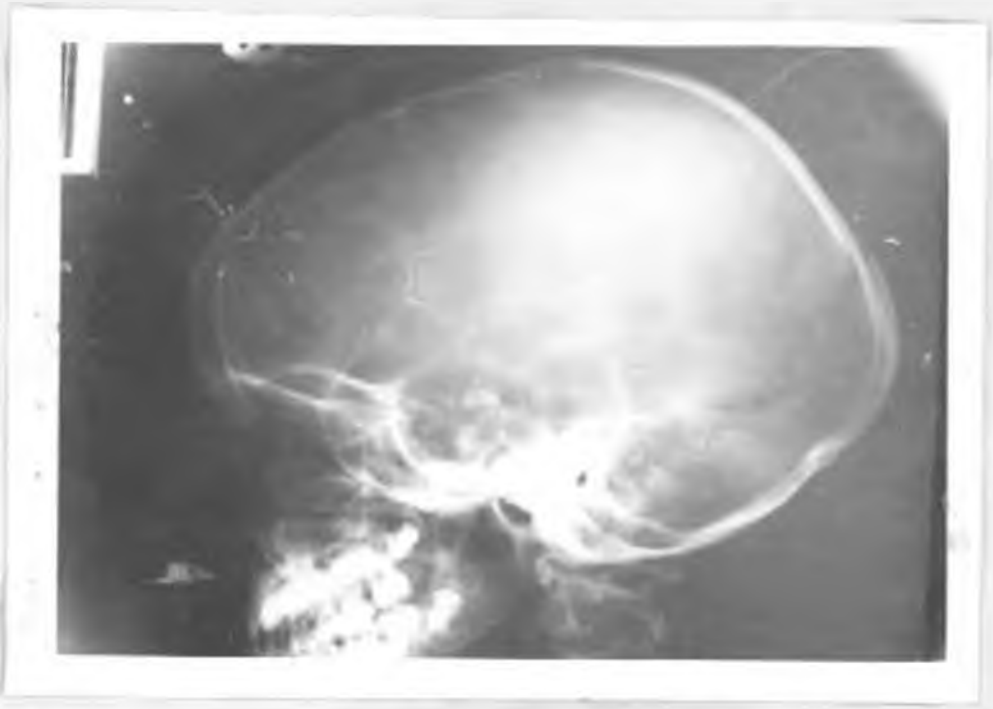
ABNORMALITY	NUMBER OF CASES	%
Shift of Midline	-	-
Elevation of A_1 of Internal Carotid Artery	4	50
Displacement of Internal Carotid Artery	0	0

VENTRICULOGRAPHY:

This was done in 3 patients. The films were untraceable but the report in the file indicated - there was bilateral dilatation of lateral ventricles in all 3 patients indicating third ventricular invasion of the tumour.

RADIOGRAPH SHOWING NORMAL LATERAL SKULL

RADIOGRAPH SHOWING SUPRA-SELLAR CALCIFICATION WITH FLATTENING OF POSTERIOR CLINOID PROCESSES



RADIOGRAPH SHOWING SUTURAL DIASTASIS AND SUPRA-SELLAR CALCIFICATION

ARTERIOGRAM SHOWING THE CHARACTERISTIC ELEVATION OF A_1 OF RIGHT ANTERIOR CEREBRAL ARTERY

DISCUSSION

Craniopharyngioma was first described by Erdheim in 1903 (1). Since then the literature about craniopharyngioma has been increasing enormously, yet because of the rarity of the tumour, statistical analysis are usually based on small number of cases. The craniopharyngiomas are encapsulated neoplasms of either oval or spherical in shape. The size of the tumour varies when clinically manifest from 2 to 10 cm. in diameter (1). The tumour may be cystic, solid or mixed.

The presenting features are directly related to the site of the tumour and surrounding structures. It arises in close proximity of hypothalamus, visual pathways, and pituitary gland. Thus the presenting symptoms are those of visual and endocrine disturbances in additions to symptoms and signs of increased intra-cranial pressure. In my series loss of vision and headache were the commonest presenting symptoms. This favourably agrees with other similar studies done in New York and MAYO clinics (1) (2). Three cases described in Kampala in one year period all presented with headache and diminution of vision as their main symptoms (7). Visual loss was varied in degree ranging from mild degree of decreased vision to total blindness. Two cases in my series presented with blindness of one of the eyes.

The endocrine disturbances were those of pituitary hypofunction and hypothalamic disturbances. Stunted growth and hypogonadism were the main clinical findings. Few pure hypothalamic disturbances were elicited. One patient presented with history of hypersomnia while one also presented with history of excessive sleep. The stunted growth was based on visual impression since none of the patient was done bone age assessment.

The peak age incidence in this series was 6-10 years; this agrees favourably with Julian and Hoffman (4). But in similar studies done by Barnett et al, the peak age incidence was within the second decade. This difference in peak incidence may be explained by varied timing to see the doctor; as shown by variable length of duration of symptoms.

The main clinical findings were those related to visual disturbances in which there were field defects in (58%). Julian and Hoffman in their series showed a visual defect of 86% in which over 80% were of bitemporal hemianopia. In my series the low figure may be probably be explained by inadequate eye examination done as shown in the case notes; however in my series also bitemporal hemianopia was the commonest field defect. Papilloedema was uncommon as compared with high frequency of optic atrophy. This may be explained

by the time factor - the interval between the onset of the disease and the time of eye examination.

Radiologically the commonest radiological finding was calcification in the supra- or infra-sellar region (51%). Most people find that between 60% to 75% of craniopharyngioma calcifiy. In my series, the figure is slightly lower, this may be genuine or may be attributed to poor standard of radiography which might have prevented the detection of some fine or minute calcification.

TABLE VIII : COMPARES RESULT OF AUTHORS

AUTHORS	PERIOD OF STUDY	NUMBER OF CASES	% CALCIFICATION
BARNET	20 years 1932-1952	62	73
LINDGREN AND DECHIRO	Not given	54	55
LOVE AND MARSHALLS	Over 20 years	100	56
ROUSERIO	Not given	26	76
MACKENZIE AND SOSMAN	10 years	35	76
BAILEY	1 year	3	100
EVANS	8 years	31	51

There was erosion with expansion of pituitary fossa in 33%, no attempt was made to find out which particular part of the pituitary fossa was most affected, this was because some of the radiological reports were extracted from files when actual films were untraceable. This was the main draw back in this study. Barnet and others however have reported the most hit are posterior clinoids and dorsum sellae. It must be appreciated that pituitary fossa radiological abnormality were by no means specific for craniopharyngioma. However Barnet described an unexpected finding of increased density of various portions of sella turcica of some of patients studied. Such findings are not recorded in my series. Sutural diastasis as would be expected was more common in the younger age group. This is also a non-specific findings of increased intra-cranial pressure.

The pneumoencephalographic changes (not recorded in my series) associated with craniopharyngioma are seen in the ventricular system and neighbouring subarachnoid cisterns. The lateral ventricles are commonly enlarged depending upon the extent of impingement of the tumour against foramina of monroe. The floor of frontal horn of enlarged lateral ventricle are occasionally deformed, the deformity resulting from the projection of the tumour into the frontal lobe and appears as a concave impression in pneumoencephalographic examination.

The third ventricle is usually displaced upward and backward to a lesser degree. The inferior arterior portion of third ventricle is occasionally deformed, the degree of deformity depending upon the size and extension of the tumour. The deformity is seen as a concave impression with concavity directed downwards. In most cases of pneumoencephalographic examination aqueduct of sylvius and fourth ventricle are normal in size and shape.

Arteriography which includes the examination of both carotid and vertebral arteries, their distribution and various drainage is occasionally done. Carotid angiography is the only examination which is usually done as recorded in literature in the study of craniopharyngioma. Vertebral angiography although rarely done can demonstrate the posterior extent of the tumour. Carotid angiography a part from demonstrating the extistence of space occupying lesion as may be shown by displacement of intra- or extradural branches of internal carotid artery may be used to exclude the possibility of the lesion being an aneurysm; which may present as a pituitary tumour, which may because of its frequent calcification in the supra-sella region may be misinterprated as a craniophryngioma.

DIFFERENTIAL DIAGNOSIS

These are conditions which produce signs and symptoms of increased intra-cranial pressure and do also produce calcification to some extent in the region of the sella.

(i) THE PITUITARY ADENOMA: The patients are usually more than 10 years. There are signs of increased intra-cranial pressure and hypothalamic disorders are absent or rare. The visual field defect are regular and symmetrical. Signs and symptoms of acromegaly may be present. If the pituitary adenoma is eosinophilic - the sella is symmetrically enlarged - calcification is rare.

(ii) OPTIC GLIOMAS: Patients are usually less than 10 years. The vision is rapidly affected. The sella is infrequently enlarged and shows J - shaped configuration in lateral skull radiograph. There may be widening of optic foramina. Calcification is quite common (33%) (1).

(iii) TUBERCULUM SELLAE MENIGIOMA: The patient is usually female overe middle age. There are bizzare field defects associated with Forster Kennedy syndrome. Endocrine disturbances are rare. The sella shows no significant change. Hyperostosis of the neighbouring bones may be seen.

CONCLUSION

The clinical and radiological presentation in our local population does not differ significantly from that described in literature. Thus a diagnosis of craniopharyngioma can be made with reasonable accuracy, by mere clinical history and plain film radiography. Thus a patient who presents with headache, disturbance of vision and plain skull films shows calcification in the supra- or infra-sella region a diagnosis of craniopharyngioma should be at the top of the list in the differential diagnosis.

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