

The Grossly enlarged or "Missing" sella

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INTRODUCTION

Enlargement of the sella turcica as a mirror of diseases has been exhaustively discussed by many authors (Camp 1949; Thomson 1955; Mahmoud 1958; New 1966) but little has been written about the grossly large sella turcica.

As far back as 1910, Schuller has pointed out that on a plain film of the skull, radiological distinction between suprasellar and intrasellar tumours could not be definite. This has been widely accepted for many years but since then, attempts have been made by several authors (Camp 1950; Epstein 1951; Ross & Greita 1966; Du Boulay & Trickey 1970; Stargardt & Margolis 1972) to establish a definite relationship between sellar changes and the site of the primary lesion.

Our aim is to identify the site and possible nature of causative lesion on plain films in the grossly enlarged sella turcica, (as seen in the University Hospital, University of Malaya) and to suggest the ideal neuroradiological investigations.

MATERIAL

(a) Chromophobe Adenoma

A female, 26 years old had mental and physical retardation since birth. She can say a few words sensibly and obey simple commands. Recently she had urinary incontinence. There were features of cretinism with scaly, coarse skin, coarse voice and infantile limb proportions.

2 Hr. I^{131} uptake 3%

24 Hr. I^{131} uptake 3%

Visual fields showed bitemporal hemianopia with pallor on both discs. VAL 6/60 VAR 6/60.

Skull:- Demonstrate a typical "ballon" sella. The sella was grossly enlarged. The dorsum sella was very thin and bowed. Under-cutting of the anterior clinoids which are large and prominent was seen.

AEG:- Showed a large intrasellar lesion with a large suprasellar extension obliterating anterior 2/3 of III ventricle, and undercutting the anterior horns of the dilated lateral ventricles.

At operation - Chromophobe adenoma.
Pituitary tumour pushing optic chiasm back.
Both optic nerves were stretched and thinned out.

(b) Cystic Craniopharyngioma

A male, 28 years, had a history of left-sided headaches for 13 years, which became more severe after a motor vehicle accident in which he lost consciousness for 10 hours, 3 years ago.

Had history of failing vision, decreased libido, decreased concentration, and intellectual deterioration.

There was bitemporal hemianopia. VAR 6/9, VAL 6/12. The plain films show that there was destruction of the floor of the sella with under-cutting of the anterior clinoid processes which were prominent. The dorsum sella was thinned and not bowed. There were some irregular calcification just above the sella. Note position of yttrium implant.

Air ventriculogram:- showed a massive suprasellar mass with total obliteration of the III ventricle with upward extension to the anterior

horns of the dilated lateral ventricle.

(c) Aqueduct Stenosis

A 28 year-old male who had been mentally slow since childhood. He has been unable to retain employment as he was too "slow". Had ? hemiplegia 8 years ago and was admitted to a Hospital. There was improvement of this ? hemiplegia but noticed increasing weakness of left limbs for past 4 years.

Fundi – the optic discs showed no clear cut margin with peripheral pigmentation. VAR 2/60 VAL 2/60.

The plain skull films showed that the sella turcica was grossly enlarged, the enlargement more marked posteriorly. The dorsum sella was foreshortened and indistinct. Anterior clinoid processes were thickened and large with steep anterior walls.

AEG:- Air could not enter the ventricular system but instead into the subarachnoid cisterns and cortical sulci.

Air ventriculography:- showed a grossly dilated front end of the III ventricle occupying the whole of the sella turcica. No air seen in the cerebral sulci and confirms the diagnosis of aqueduct stenosis.

(d) Meningioma

A male, 40 years had headache, giddiness, and transient obscuration of vision for 4 years. Last 1½ years of loss of libido, and cold intolerance. Recently there had been personality changes, nocturnal anuresis and tinnitus. Noticed failing vision for past 6 months. Right amaurosis with left upper temporal quadrant defect. There was bilateral papilloedema with secondary optic atrophy.

The plain films show that the sella turcica was enlarged with almost absence of the dorsum sella and irregular floor. There was also absence of the lamina dura posteriorly with undercutting of the anterior clinoid processes and sclerosis of orbital plate

Left carotid angiogram:- highly vascular extensive but fairly circumscribed space occupying lesion measuring 9 cm in the widest diameter in left cerebral hemisphere occupying the frontal lobe and extending backwards and downwards invading the parietal and temporal region. Note the large anterior meningeal artery supplying this lesion. The anterior cerebral artery was pushed backwards.

At operation:- large vascular meningioma arising largely from the pterion and outer third of the

sphenoid wing and extended posteriorly to attach to the frontal horn of the lateral ventricle. Inferiorly, it extended towards the optic nerves were intact.

(e) Nasopharyngeal Carcinoma

A 45-year-old female complained of left-sided nasal obstruction and blood-stained nasal discharge for eight months and headaches for past five months.

Last 10 days she had paraesthesia and ptosis of both eyes. Both eyes exhibited no movement at all. Right pupil was fixed and ¾ dilated. Left pupil reacted sluggishly to light and ½ dilated.

On biopsy a large fungating growth at the left post-nasal space pushing the soft palate forward and the uvula to the right was seen. Enlarged matted lymph nodes at upper cervical region were palpated.

Biopsy results showed squamous cell carcinoma of the nasopharynx.

The plain skull films show that there was haziness of the sphenoid sinus with destruction of the posterior aspect of the floor of the sella and the dorsum sella. Undercutting of the anterior clinoid processes was seen.

The view of the neck showed a large irregular soft tissue mass in the nasopharynx.

(f) Primary Empty Sella Syndrome with Communicating Hydrocephalus

A male 60 years had periodic left-sided temporal headaches for 10 years, which became more severe during the past 6 years. Loss of libido and erection for 4 years. Visual fields showed a definite loss of both upper quadrants of temporal fields. VAR 20/30 VAL 20/40. No papilloedema or optic atrophy.

The plain films showed that there was extensive destruction of the body of the sphenoid. The dorsum sella was totally destroyed with undercutting of the anterior clinoid processes which were large and prominent.

AEG:- Large air-containing cyst within the sella which communicate with the basal cistern. Note dilatation of the III and lateral ventricles. Consistent with intrasellar subarachnoid cyst with the communicating hydrocephalus.

Discussion

This paper is not aimed to give a comprehensive account of all the known cases of grossly enlarged

sella as other well known lesions e.g. suprasellar astrocytoma, chordoma, plasmacytoma and aneurysms have been left out as they have not been seen in the University Hospital.

The cases discussed can be grouped into

(a) Intrasellar lesions - chromophobe adenoma.

(b) Suprasellar lesions - craniopharyngioma.

(c) Lesions remote from the sella

(1) Aqueduct stenosis

(2) Frontal meningioma

(3) Nasopharyngeal carcinoma

(4) Empty sella

(a) Chromophobe Adenoma (Fig. 1A & 1B)

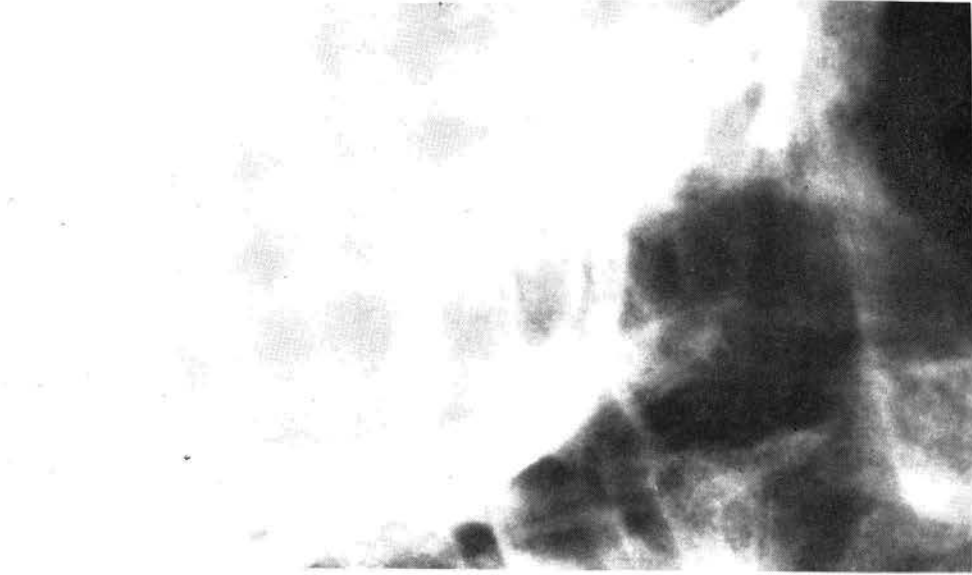


Fig. 1A Plain skull film of chromophobe adenoma showing grossly enlarged sella turcica and undercutting of anterior clinoids.



Fig. 1B AEG showing a large intrasellar lesion with large suprasellar extension obliterating anterior 2/3 of III ventricle.

The findings of a "Ballooned" sella on plain radiograph and clinical finding of a visual field defect must almost certainly indicate the presence of a pituitary adenoma. Only 7% of craniopharyngiomas and 4% of optic gliomas show a "ballooned" sella (Ross, 1966). The suprasellar extension of a pituitary adenoma is of utmost importance to the

neuro-surgeon and air study is the investigation of choice to demonstrate tumour size and extent. The choice between ventriculography or lumbar air encephalogram is dependent on the presence or absence of papilloedema.

(b) Craniopharyngioma (Fig. 2A & 2B)



Fig. 2A Plain skull film of craniopharyngioma showing destruction of floor of sella with irregular calcification just above the sella.



Fig. 2B Air ventriculogram showing a massive suprasellar mass with total obliteration of the III ventricle.

The presence of a grossly enlarged sella with suprasellar calcification is highly suggestive of craniopharyngioma. This is present in our patient. Craniopharyngioma exhibits calcification in the majority of cases in children and in less than half of the cases in adults. The dorsum sella is known to be short and flat topped in 50% of cases (Du Boulay, 1967). However in our case the dorsum is not short or flat topped but is seen to be angulated forward at less than 90° and there is irregular amorphous calcification. The extent of calcification do not necessarily indicate the full extent of the tumour and lumbar air encephalography or ventriculography is

necessary to delineate the full extent of the tumour.

Other tumours exhibiting calcification are

- (1) Chordomas.
- (2) Chromophobe adenomas (1.2 - 6%) (Camp 1950; Du Boulay & Trickey 1962)
- (3) Meningiomas (5 - 15%)
- (4) Aneurysm

Angiography may be necessary to ascertain the pathology of the tumour mass as craniopharyngiomas and chordomas are virtually always avascular.

- (c) Aqueduct Stenosis (Fig. 3A & 3B)

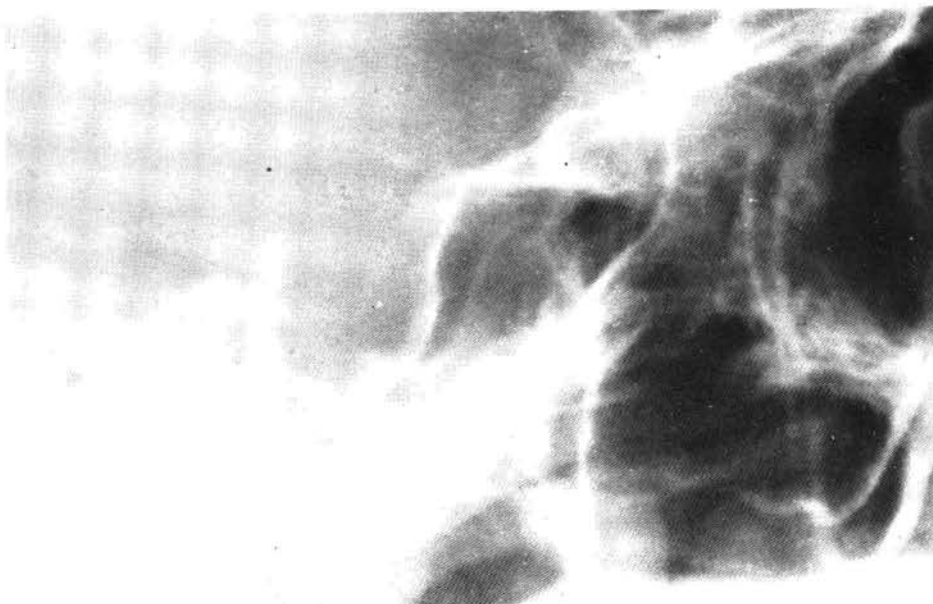


Fig. 3A Plain skull film of aqueduct stenosis showing sella turcica grossly enlarged with thickened and large anterior clinoid processes.

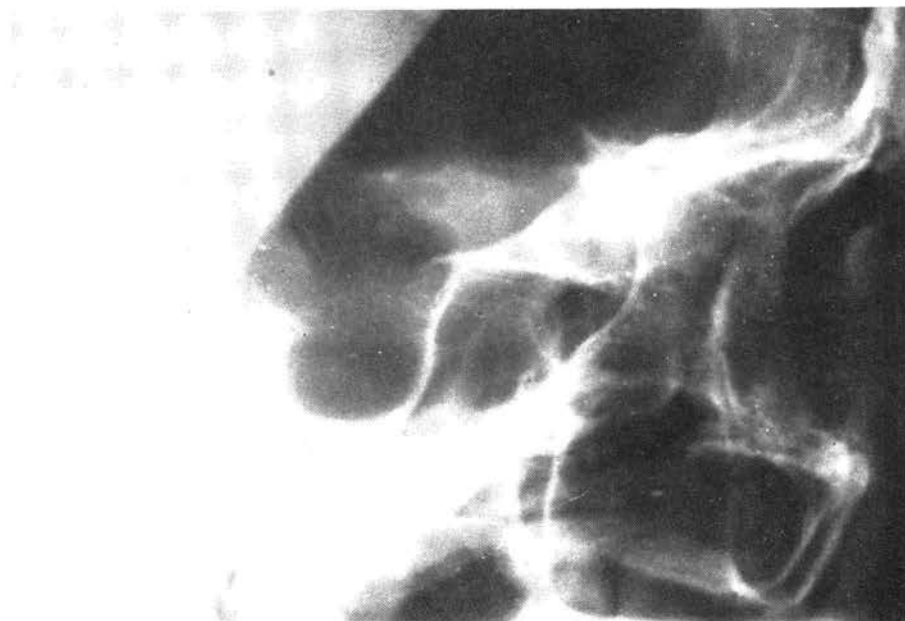


Fig. 3B Air ventriculogram showing grossly dilated anterior end of III ventricle occupying the whole of the sella turcica. No air seen in cerebral sulci or cisterns.

The classical plain film findings of aqueduct stenosis in an adult are:

- (1) Short dorsum (44%).
- (2) Long steep anterior wall consisting partly of a more or less vertically placed elongated sulci.
- (3) Massive anterior clinoid processes (Du

Boulay & Trickey 1970).

Our case shows all of the above findings.

The investigation of choice would be air Myodil ventriculography as this would demonstrate the presence of total or incomplete aqueduct obstruct.

- (d) Meningioma (Fig. 4A & 4B)

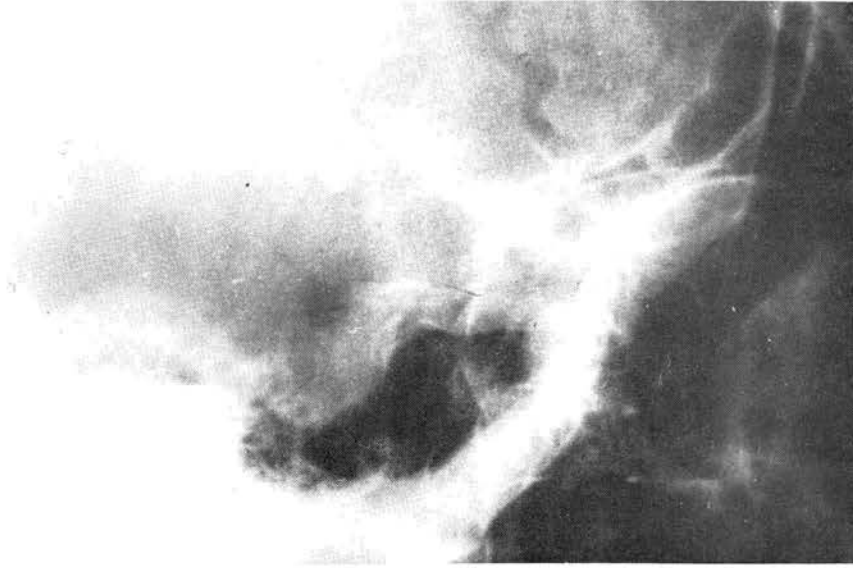


Fig. 4A Plain skull film of meningioma showing enlargement of sella turcica with absence of the lamina dura posteriorly and sclerosis of orbital plate.

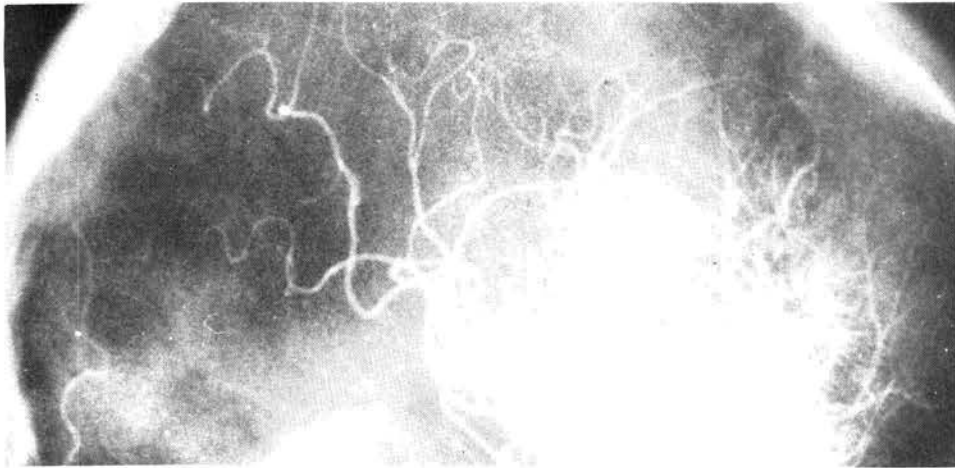


Fig. 4B Left carotid angiogram showing highly vascular circumscribed lesion supplied by large anterior meningeal artery.

Meningiomas distant from the sella, e.g. parietal or frontal regions can produce changes in the sella, similar to those produced by raised intracranial pressure. These are

- (1) truncation or absence of the dorsum sella.
- (2) loss of lamina dura or the dorsum sella.
- (3) undercutting of the anterior clinoid processes.
- (4) Large sella.

The findings are not specific but if sclerosis can be visualised as in our patient who has sclerosis of the orbital plate than the diagnosis of meningioma would almost be a certainty. Furthermore 15% of meningiomas calcify (Sutton 1971).

The next investigation of choice would be a brain scan. This will show an area of increased uptake. Angiography is invariably necessary to show the extent to the tumour and arterial supply.

(c) Nasopharyngeal Carcinoma (Fig. 5A & 5B)

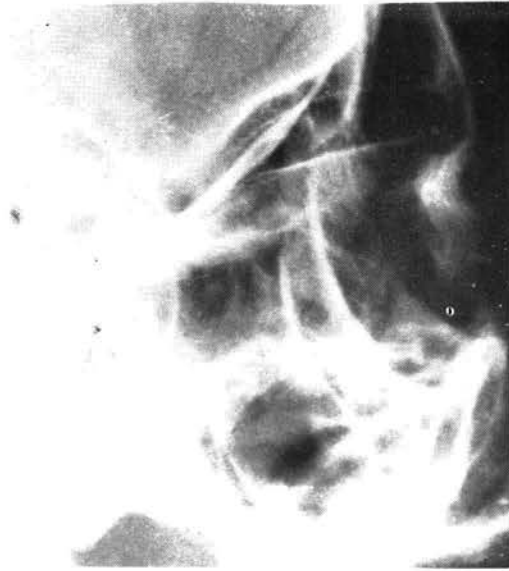


Fig. 5A Plain skull film of nasopharyngeal carcinoma showing haziness of sphenoid sinus with destruction of dorsum sella and posterior part of floor of sella.



Fig. 5B Large irregular soft tissue mass in the nasopharynx.

Nasopharyngeal carcinoma is known to destroy the sella by direct extension. As in our case there is direct extension through the sphenoidal sinus to involve the dorsum sella with destruction of the

posterior half of the sella turcica. The diagnosis can be implied from the large soft tissue mass in the nasopharyngeal space and a history of epistaxis.

(f) Empty Sella Syndrome (Fig. 6A & 6B)

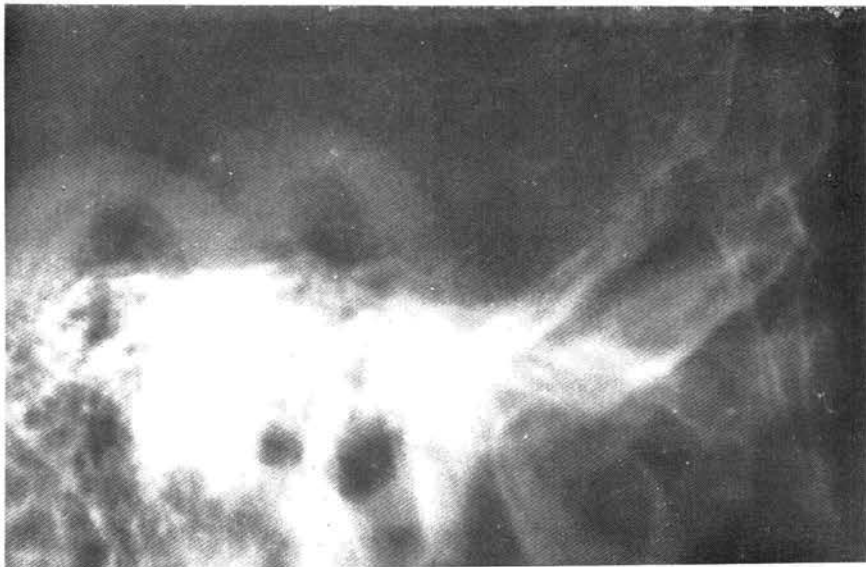


Fig. 6A. Plain skull film of primary empty sella showing extensive destruction of body of sphenoid with destruction of dorsum sella and undercutting of the anterior clinoids.

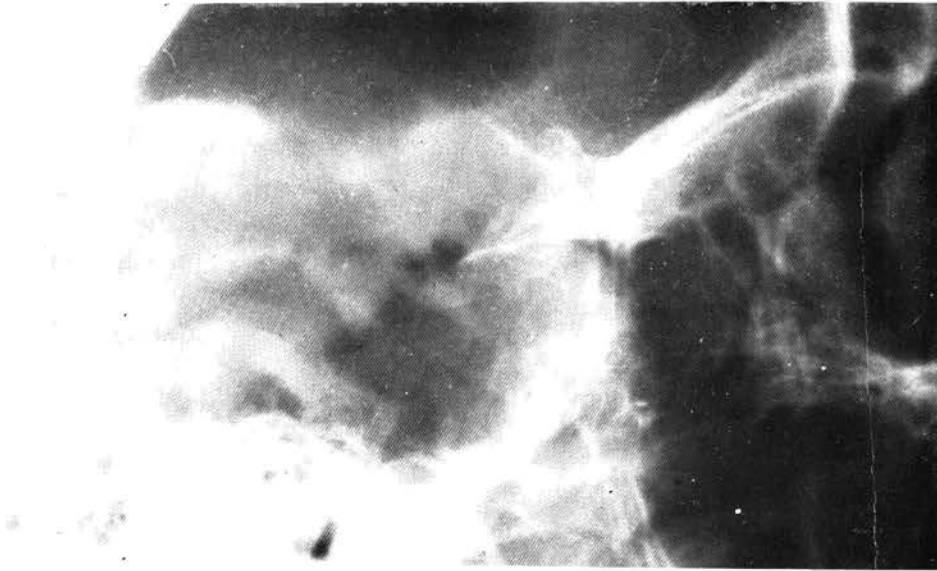


Fig. 6B. AEG showing large air containing cyst within the sella which communicates with the suprasellar cistern.

In this condition, the sella is large in 84% of patients. The cause is attributed to elevation of intracranial pressure which could be slight or intermittent, remodelling the anatomy of the sella through a congenital incompleteness of the sella diaphragm. This syndrome is seen mainly in females (Neelon et al. 1973). Our patient is a male and showed total absence of the dorsum sella with undercutting of the anterior clinoid process.

The presence of a grossly enlarged sella or "missing" sella without any radiological visualisation of calcification, raised intracranial pressure, sclerosis or extension from a nearby tumour could suggest an empty sella syndrome.

If there is no evidence of papilloedema the investigation of choice would be fractional lumbar air encephalogram.

SUMMARY

Six cases of grossly enlarged sella are discussed. The cases include chromophobe adenoma, craniopharyngioma, aqueduct stenosis, meningioma, nasopharyngeal carcinoma and the empty sella syndrome.

The plain film findings and the appropriate specialised neuroradiological investigations are discussed.

An attempt has been made to correlate the changes in and around the sella turcica on plain films to serve as a pointer towards the nature of the pathological processes. This will influence the selection of the ideal investigation or investigations necessary to demonstrate the extent and the pathology of the tumour.

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