Med. J. Malaysia Vol. XXXI No. 3, March, 1977

The Empty Sella Syndrome

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Introduction

THE RADIOGRAPHIC APPEARANCE OF an enlarged sella turcica is usually associated with an expanding intracranial lesion often a result of an intra- or peri-sellar neoplasm (Agrawal et al, 1966). Occasionally, pneumoencephalography shows that the enlargement is not occupied by a tumour mass but admits air instead as a result of the extension of the subarachnoid space below the clinoid processes. This constitutes the "empty sella" syndrome.

Busch (1951) first described the empty sella when the diaphragm sellae was incomplete or formed only a small peripheral rim and the pituitary gland was not grossly visible as viewed from above at necropsy. Colby and Kearns[4] (1962) described what is referred to as the secondary "empty sella syndrome" in a patient who had radiotherapy. With recurrence of visual symptoms he was operated on and the sella at operation was found to be empty without evidence of a tumour mass. We are reporting two cases encountered at the University Hospital illustrating the primary and secondary empty sella syndrome. Weiss[10] and Neelon[8] emphasized the need to differentiate between these; the latter occurring after surgical or radiotherapeutic procedures whereas the aetiology is quite different in the former.

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Case Reports

Case 1

OHS, a 60 year old Chinese male was first seen in another hospital in 1971 with a 10 year history of left sided temporal headache. He had no symptoms or signs to suggest hypopituitarism. His visual fields were intact. He was admitted to the University Hospital in December 1973 for a pneumoencephalogram.

On examination the patient was an intelligent slightly obese male. His pulses were normal and his blood pressure measured 160/100 mmHg in the supine position. He had normal hair growth. His visual acuity was not impaired and his fundi were normal. There was a bitemporal upper quadrant hemianopia. No other abnormal signs were detected in the other systems. Psychological assessment showed normal verbal intelligence on the Wechsler Intelligence Scale but non-verbal performance showed a discrepancy suggesting an organic abnormality.

Laboratory investigations revealed normal thyroid and adrenal functions. The insulin tolerance test was normal. Examination of the cerebrospinal fluid revealed no abnormality, the pressure was not elevated. Urinalysis showed the specific gravity to be within normal limits. Plain skull radiograph showed an enlarged sella (Fig. 1) and a pneumoencephalogram demonstrated an enlargement of the lateral, third and fourth ventricles with air filling the enlarged sella (Fig. 2). A brain scan was normal.

He was discharged as no surgery or radiotherapy was deemed necessary. He subsequently had an

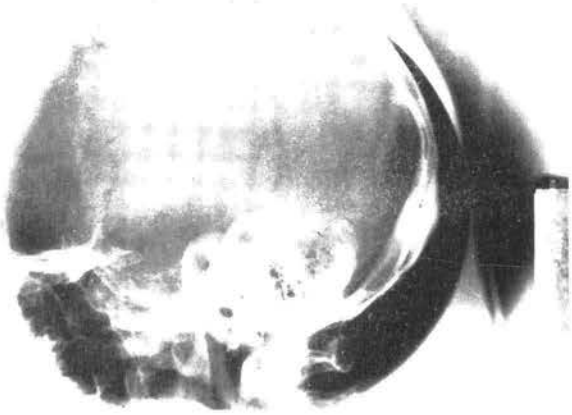

Fig. 1
Lateral projection – showed an enlarged sella with destruction of posterior clinoid processes and dorsum sella.

Fig. 2
Lateral projection taken in "overhanging head" position showing the following features:
(1) Enlarged lateral and third ventricles.
(2) Absence of air in cerebral sulci.
(3) Trapping of air in the suprasellar cisterns.
(4) Subarachnoid air filling the enlarged sella.

isotope cisternography at the Royal Perth Hospital, Australia in May 1974 and this confirmed the presence of a communicating hydrocephalus. Pituitary function was normal and there was no evidence of diabetes insipidus. He is being followed up regularly at the University Hospital.

Case 2

LSY, a 35 year old housewife was admitted in November 1970 with complaints of amenorrhoea and infertility. There was no history of sexual dysfunction, visual disturbances or headache.

On examination the patient was obese and hirsute. Her pulses were normal and the blood pressure measured 170/100 mmHg. Her visual acuity and fundi were normal. There was bitemporal hemianopia. The rest of the physical examination was normal. A plain skull radiograph and tomograms showed an enlarged sella with a double floor contour. There was undercutting of the anterior clinoid and erosion of the posterior clinoid processes (Fig. 3). A diagnosis of chromophobe adenoma was made and the patient underwent a course of radiotherapy.

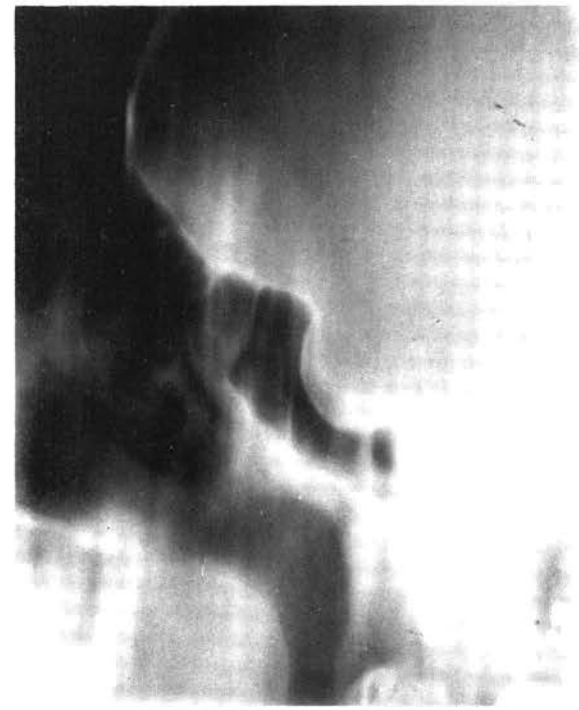

Fig. 3
Lateral tomogram of sella turcica.

In August 1972 patient was readmitted with complaints of sudden blurring of vision, diplopia and headache. She developed a transient right 3rd nerve palsy. There was no evidence of endocrine dysfunction. A right carotid angiogram showed no abnormality. A pneumoencephalogram revealed an empty sella (Fig. 4). The patient was last seen at the University Hospital in June 1974 and no deterioration in her visual fields was noted.

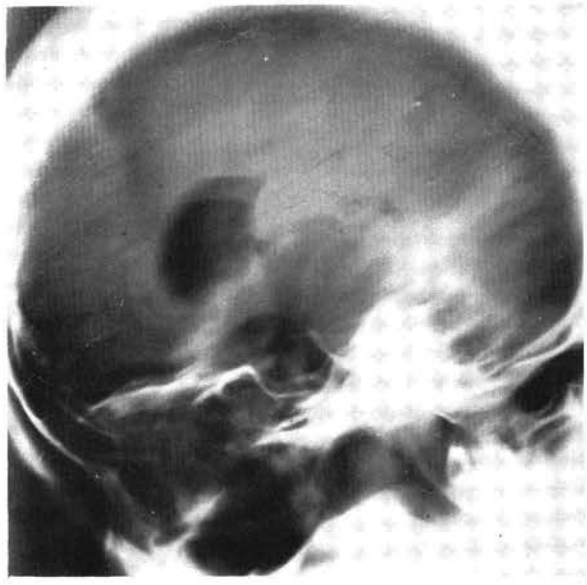

Fig. 4
Lateral projection taken in "over hanging" head position showing air in the enlarged sella.

Discussion

The aetiology and pathogenesis of the primary empty sella syndrome have not been established with certainty. Caplan & Dobben[3] reviewed the possible mechanisms and concluded that the syndrome was the end result of a number of different pathogenic mechanisms. Neelon et al[8] considered the rupture of an intra or parasellar cyst; pituitary hypertrophy and subsequent abiotrophy; infarction of sellar contents and transmission of cerebrospinal fluid pressure through a congenitally defective sellar diaphragm as possible mechanisms to explain the occurrence of the primary empty sella. They stated that the most accepted mechanism concerns the transmission of a normal or elevated cerebrospinal fluid pressure, with an incomplete sellar diaphragm as an essential pre-requisite.

Colby and Kearns[4] from the Mayo Clinic, in a review of their results of radiation therapy of 149 pituitary tumours considered an empty sella as a rare complication. Three cases reported by Lee and Adams[7] (1967) and the second patient in this report are examples of secondary empty sella syndrome. It is important to be aware of this complication because with recurrence of symptoms the patient may be subjected to an unnecessary operation.

The absence of any endocrine dysfunction in our patients is not surprising as pituitary function may remain normal even when large amounts of pituitary parenchyma has been destroyed[10]. Caplan[3] noted pituitary dysfunction in four out of six patients whilst Neelon[8] in an analysis of thirty one patients noted endocrine dysfunction in only eight of them. Brisman[2] in a study of 19 patients with empty sella noted some degree of endocrine dysfunction in eleven of them.

The prognosis in patients with primary empty sella is generally considered to be good. Neelon[8] et al in their extensive review of 31 cases considered the condition benign and the occasional instances of endocrine dysfunction to be unrelated to sellar enlargement or to the extension of the subarachnoid space into the sellar cavity. They concluded that pneumoencephalography prior to therapeutic intervention is essential in all patients suspected of having pituitary tumours based on plain skull radiographs. Radiation therapy is not warranted in patients with the empty sella syndrome. Patients without symptoms should be examined regularly since pituitary dysfunction or chiasmal pressure may develop.

Acknowledgement

We would like to thank Professor H.O. Wong and Professor T.G. Loh for their helpful criticisms, Puan Rohani for typing the manuscript and the Medical Illustration Department for the illustrations.

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