

Case Report

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Atypical magnetic resonance imaging findings of craniopharyngioma

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SUMMARY

Three cases of craniopharyngiomas with atypical MRI findings are reported. The first patient had a nasopharyngeal craniopharyngioma. Its unusual location made diagnosis difficult. The second patient had a massive craniopharyngioma with extensive cystic expansion, involving the anterior, middle and posterior cranial fossae, and extending into the foramen magnum. The tumour of the third patient involved the suprasellar region with a large extension into the third ventricle, and demonstrated a predominantly high signal intensity on all T1-weighted, proton-weighted and T2-weighted images. These patients further stressed the complexity of MRI findings in craniopharyngiomas.

Key words: *craniopharyngioma; magnetic resonance; sella turcica.*

INTRODUCTION

Craniopharyngiomas are unusual neoplasms, accounting for 2–3% of all brain tumours and 14% of supratentorial tumours.¹ A bimodal distribution by age has been noted with peak incidence rates in children (aged 5–14 years) and among older adults (aged 50–74 years).² Due to its multiplanar capabilities, MRI has been proven to be invaluable for evaluating the extent of a craniopharyngioma and its relationship to neighbouring structures.³ It has been suggested that MRI should be the primary radiological investigation in such patients.⁴ At Rui Jin Hospital (Shanghai), MRI (plain scans, enhanced scans and MR angiography when necessary) and X-ray tomography of the cranial skeletal structure are routinely used for pre-operative neuroradiological work up of craniopharyngiomas.

Owing to their complex histological components, craniopharyngiomas demonstrate a wide range of MR appearances. Although MRI is accurate for delineating a tumour's extension, histopathological diagnosis by MRI is not always easy. Here we report three cases of surgical histopathologically proven craniopharyngiomas with unusual MRI appearances.

CASE REPORTS

Case 1

A 9-year-old girl presented with a clinical history of nasal obstruction, exophthalmos and an irritable personality for

2 years. She also had ophthalmodynia and visual problems for 1 year. Magnetic resonance imaging demonstrated a large mass, measuring 46 × 52 × 67 mm, in the nasopharyngeal region, which had invaded the sphenoid bone and ethmoid bones (Fig. 1). The tumour contained both solid and cystic components. The cystic component showed low signal intensity on T1-weighted (T1W) images and high signal intensity on T2-weighted (T2W) images. The solid component showed moderate signal intensity on T1W images and heterogeneous signal on T2W images. The solid component was enhanced markedly after injection of gadolinium-chelate. The sella turcica was elevated, but the morphology of the pituitary gland, suprasellar cistern and chiasm were unremarkable. Surgical pathology showed that the mass lesion was an ectopic craniopharyngioma.

Case 2

A 9-year-old girl was referred for examination due to a deterioration in hearing in her right ear for 1 week. Magnetic resonance imaging demonstrated a massive intracranial tumour with a lobulated configuration, which involved the anterior, middle and posterior cranial fossae, and extended into the foramen magnum (Fig. 2). The brainstem and the fourth ventricle were compressed and displaced posteriorly. The tumour involved both the right and left sides, but was predominantly located on the right side and

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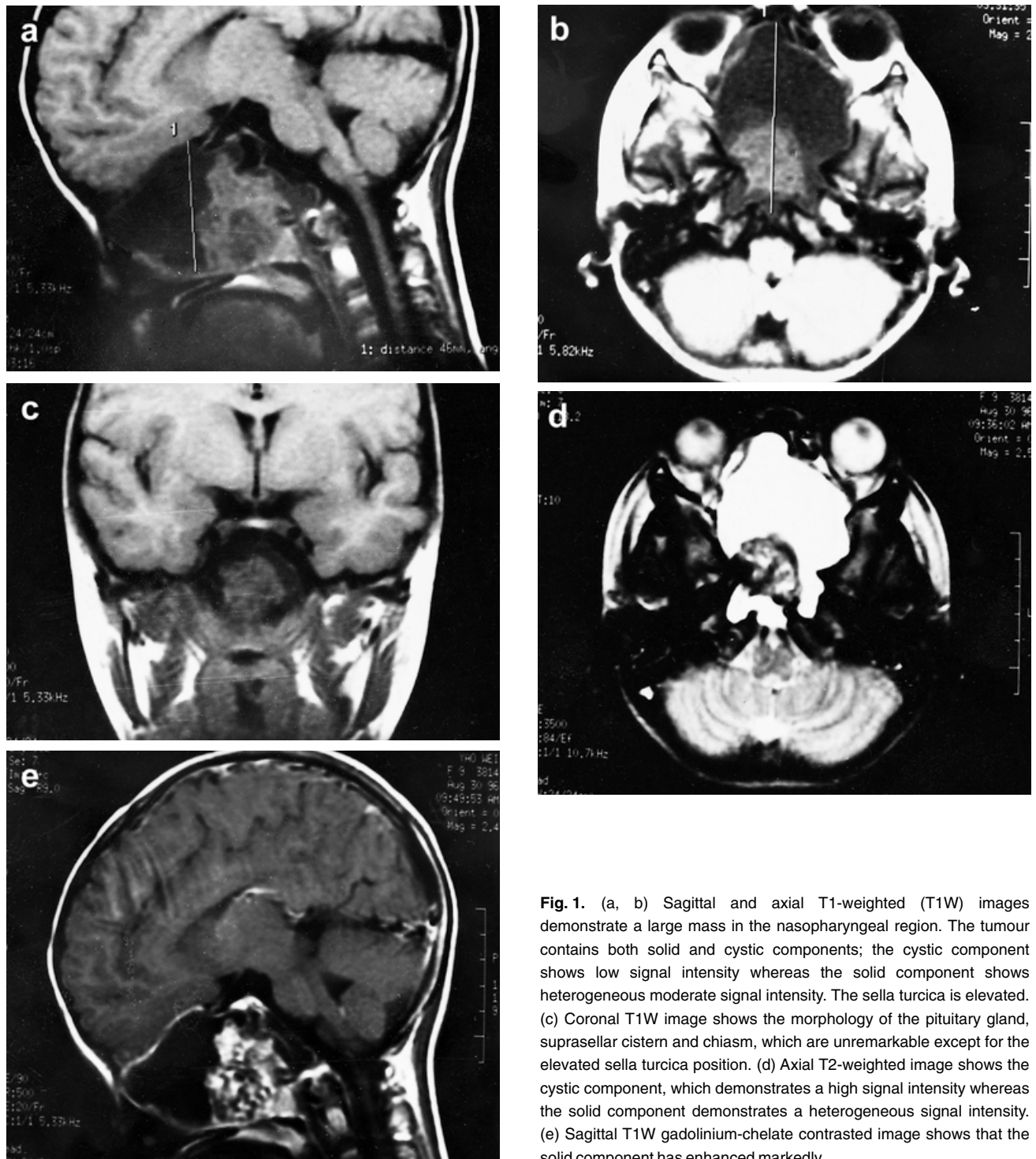


Fig. 1. (a, b) Sagittal and axial T1-weighted (T1W) images demonstrate a large mass in the nasopharyngeal region. The tumour contains both solid and cystic components; the cystic component shows low signal intensity whereas the solid component shows heterogeneous moderate signal intensity. The sella turcica is elevated. (c) Coronal T1W image shows the morphology of the pituitary gland, suprasellar cistern and chiasm, which are unremarkable except for the elevated sella turcica position. (d) Axial T2-weighted image shows the cystic component, which demonstrates a high signal intensity whereas the solid component demonstrates a heterogeneous signal intensity. (e) Sagittal T1W gadolinium-chelate contrasted image shows that the solid component has enhanced markedly.

centred in the sellar and suprasellar region. The midline structures were shifted slightly to the left side. Encased blood vessels could be found within the tumour. The tumour was isointense to cerebral parenchyma on T1W images and had homogeneously high signal intensity on T2W images. After injection of gadolinium-chelate, moderate peripheral rim enhancement was demonstrated. Surgery revealed a predominantly cystic craniopharyngioma with extensive expansion.

Case 3

A 53-year-old woman presented with staggering gait, urinary incontinence and dementia for 1 month. Magnetic resonance imaging demonstrated a $20 \times 25 \times 28$ mm mass located in the suprasellar cistern and third ventricle, with the main volume of the lesion located within the third ventricle (Fig. 3). The tumour appeared to be of predominantly high signal intensity on all T1W, proton-weighted, and T2W images, but the suprasellar part of the

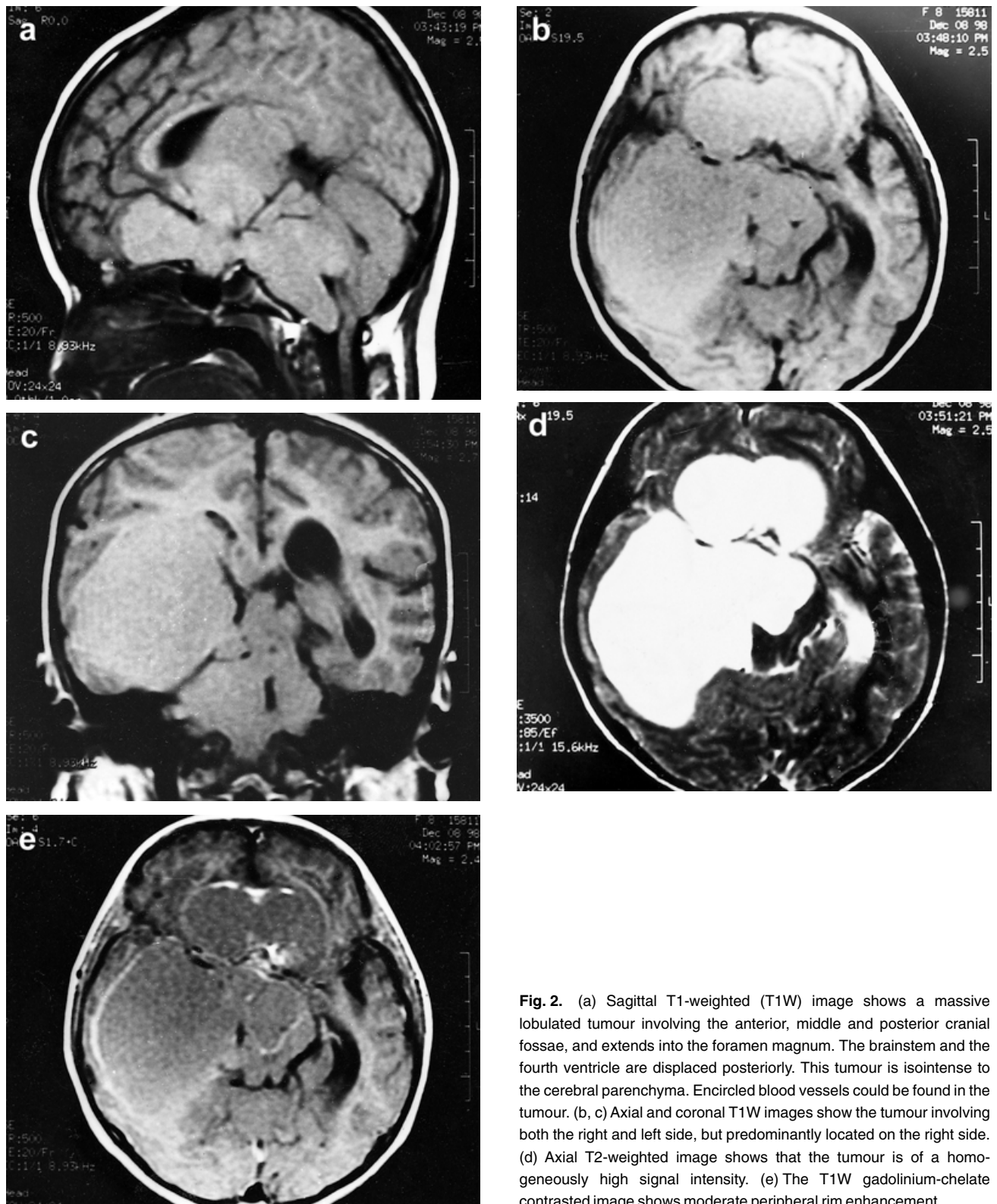


Fig. 2. (a) Sagittal T1-weighted (T1W) image shows a massive lobulated tumour involving the anterior, middle and posterior cranial fossae, and extends into the foramen magnum. The brainstem and the fourth ventricle are displaced posteriorly. This tumour is isointense to the cerebral parenchyma. Encircled blood vessels could be found in the tumour. (b, c) Axial and coronal T1W images show the tumour involving both the right and left side, but predominantly located on the right side. (d) Axial T2-weighted image shows that the tumour is of a homogeneously high signal intensity. (e) The T1W gadolinium-chelate contrasted image shows moderate peripheral rim enhancement.

lesion had heterogeneous signal. In axial images, the lesion in the anterior third ventricle did not cause the collapse of the posterior third ventricle. Obstructive hydrocephalus of both lateral ventricles and abnormal peri-ventricular zones consistent

with interstitial oedema were also demonstrated. Surgical pathology demonstrated that this craniopharyngioma had a large cystic component with cystic fluid that was rich in cholesterol crystals and blood degradation products.

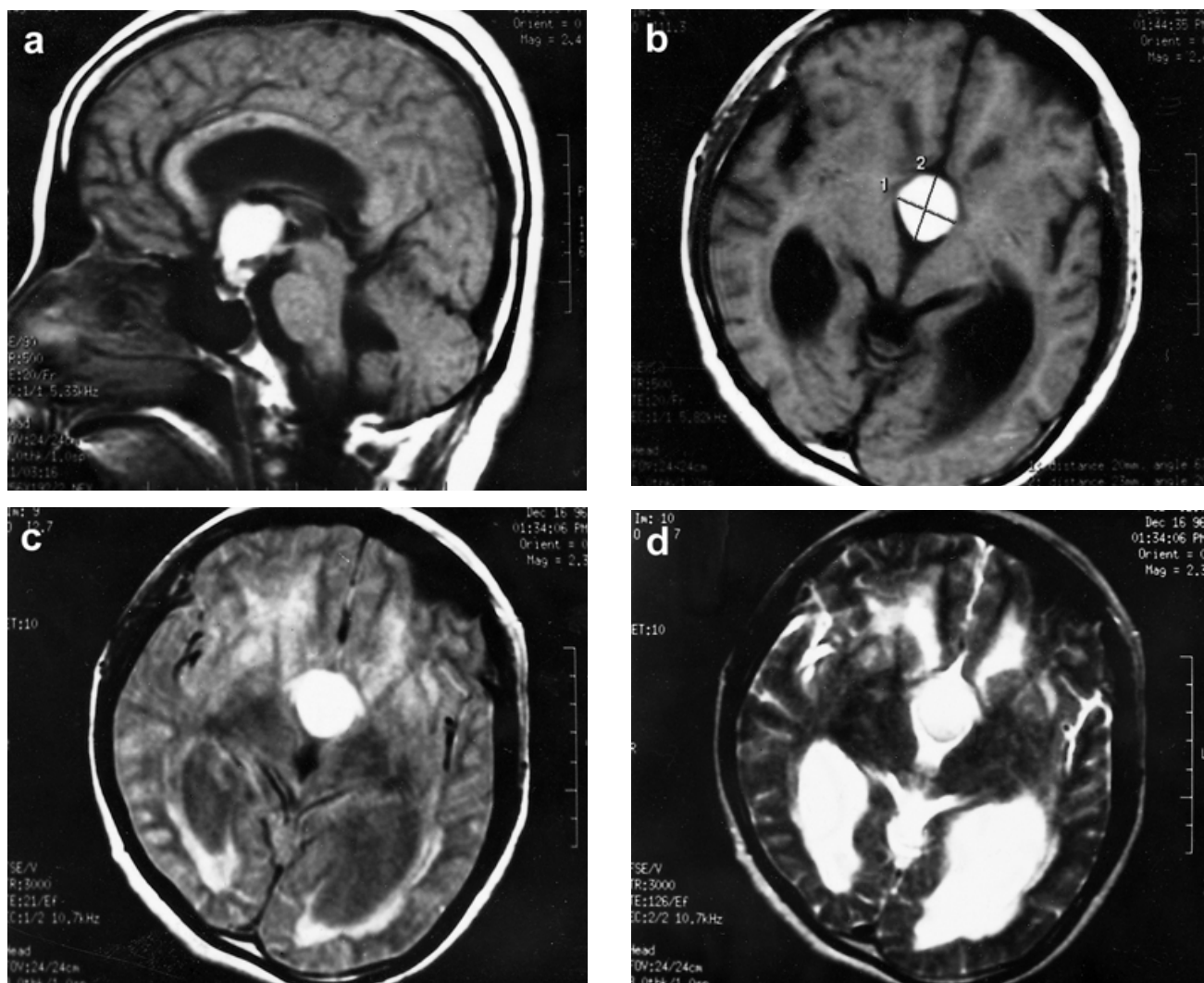


Fig. 3. (a) Sagittal T1-weighted (T1W) image shows a predominantly high signal mass located in the suprasellar cistern and third ventricle, with the main volume of the lesion lying in the third ventricle. The suprasellar cistern is of a heterogeneous intensity. (b–d) Axial T1W, proton-weighted, and T2-weighted (T2W) images show the tumour demonstrating high signal intensity on all spin echo sequences. Obstructive hydrocephalus of both lateral ventricles and with interstitial oedema in the periventricular zones (low signal intensity on T1W images and high signal intensity on T2W images) is also demonstrated.

DISCUSSION

Craniopharyngiomas are thought to arise from the path of the craniopharyngeal duct, which is the route taken by Rathke's pouch from the oropharynx to the floor of the third ventricle. Although they are histologically benign, they are capable of causing substantial morbidity and mortality because of their propensity to invade surrounding structures, including the optic chiasm, pituitary gland and hypothalamus.⁵ The most common signs and symptoms are visual deficits, endocrine dysfunction, increased intracranial pressure and cranial nerve deficits. Aggressive treatment is indicated, which is currently centred on surgical strategies and radiation therapy.^{6,7}

Craniopharyngiomas tend to have a suprasellar location (90%); however, 18% of craniopharyngiomas extend into the sella, and 5% of these tumours are purely intrasella.^{8,9} These

tumours can also extend to the anterior, middle and posterior cranial fossae. Rarely craniopharyngiomas arise primarily in unusual locations, such as the nasopharynx,^{10–12} sphenoid bone,¹³ third ventricle,¹⁴ the cerebellopontine angle¹⁵ and within the optic chiasm.¹⁶

Craniopharyngiomas may be solid or cystic or, more commonly, mixed. Calcification, cyst formation and intravenous contrast media enhancement in a suprasellar and/or intrasellar mass are the hallmarks of craniopharyngiomas on CT.¹⁷ Computed tomography is superior to MR in detecting calcifications, but MR is preferred in the evaluation of a tumour's extent for therapeutic planning.^{3,18} Owing to their complex histological components, craniopharyngiomas demonstrate a wide range of MR appearances.^{19–22} The cystic component can be hyperintense on all spin echo sequences because of cholesterol,

haemorrhage or proteinaceous fluid. If keratin, calcium or haemosiderin predominate in the cyst, MR images show hypointensity. A mixture of different chemical substances can cause isointensity to brain parenchyma on all MR sequences. Solid components of the tumour are more likely to show to intermediate intensity on T1W images. On T2W images craniopharyngiomas show a predominantly high signal of variable homogeneity. Non-calcified solid components of the tumour usually show enhancement on MR images obtained after administration of gadolinium-chelate.

The main difficulty encountered in diagnosing the first patient was caused by the tumour's unusual location. In other respects the MRI appearance was typical for craniopharyngioma. Craniopharyngiomas arising primarily beyond the sellar and suprasellar region are extremely rare. The first case of ectopic craniopharyngioma was reported by Drummond in 1938.¹⁰ The pertinent embryology for the pathogenesis of ectopic craniopharyngiomas has been described by Benitez and colleagues.¹¹ Clinically, in patients with inferiorly extending craniopharyngiomas, the most common complaints are visual disturbances; whereas in patients with craniopharyngiomas arising from the sphenoid bone or nasopharynx, nasal obstruction is the most common symptom. Radiologically, an enlarged or destroyed sella can be demonstrated in tumours located in the sella and in those with infrasellar extension. With tumours arising from the sphenoid bone and nasopharynx, the sella is usually normal in size.

The MRI findings in the second patient are atypical in terms of the extensive expansion involving the anterior, middle and posterior cranial fossae, even extending into the foramen magnum, although extension into one or other of the cranial fossae is common.²³

The MRI appearance of the craniopharyngioma of patient 2 is similar to that of epidermoid tumour. Epidermoid tumours are benign lesions, consisting of a mixture of cellular debris, keratin and cholesterol. Epidermoid tumours are well-defined, lobulated masses.²⁴ On T1W MR images, epidermoid tumours show low or moderate signal intensity, and on T2W MR images the tumours show very high signal intensity. Calcifications have also been reported.^{24,25} In a series of 40 cases of epidermoid tumours reported by Shen and colleagues, the para- and suprasellar regions were relatively common sites.²⁴ These tumours tend to insinuate themselves around delicate structures, such as cranial nerves, blood vessels and the brainstem, rather than to displace them. While the case mimicked the MRI appearance of an epidermoid tumour, peripheral rim enhancement of epidermoid tumours is less common.²³⁻²⁵

The third case is unusual because of the predominantly high signal on all sequences and its large extension into the third ventricle. These changes mimicked a colloid cyst.²⁶ However, there is a suprasellar part of the lesion that is heterogeneous in signal, and in axial images the lesion in the anterior third

ventricle has not caused the collapse of the posterior third ventricle. These observations suggest that a colloid cyst is unlikely. Although the most common pattern of craniopharyngioma is that of hypointensity on T1W images and high signal intensity on T2W images,¹⁹ it has been documented that hyperintensity on T1W images (so called 'machine oil cysts') is also observed.²⁰ The high signal on T1- and proton-weighted images was thought to be caused by cholesterol crystals and blood degradation products, as demonstrated on surgical pathology.

In conclusion, three cases of craniopharyngiomas with atypical MRI findings are reported: one nasopharyngeal craniopharyngioma; one massive craniopharyngioma with extensive expansion mimicking an epidermoid cyst; and one craniopharyngioma with a predominantly high signal on all spin echo images and marked extension into the third ventricle. These patients further stressed the complexity of the MRI appearances of craniopharyngiomas.

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