
CASE REPORT

Suprasellar Ependymoma

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ABSTRACT

Supratentorial ependymomas account for approximately 40% of intracranial ependymomas. We present an unusual case of supratentorial ependymoma, with both clinical and radiological features of a suprasellar tumour.

Key words: Computed tomography, Ependymoma, Magnetic resonance imaging, Suprasellar

INTRODUCTION

Ependymomas may occur in various locations in the brain and spinal cord. They account for approximately 2% to 8% of all primary intracranial brain tumours. About 60% of intracranial ependymomas are infratentorial, 90% of which are in the 4th ventricle. Approximately 40% are supratentorial, with extra-ventricular locations being more common.¹ We describe an unusual case presenting with clinical and radiological features of a suprasellar tumour with intrasellar extension. The tumour was excised and the diagnosis of ependymoma confirmed on pathological examination.

CASE REPORT

A 32-year-old male presented with insidious onset of disturbed sensorium, visual loss, and increased appetite of 1 year's duration. Clinical examination and laboratory investigations revealed hypopituitarism, diabetes insipidus, complete blindness in the right visual field, temporal hemianopia, and superior nasal quadrantanopia in the left visual field. Only the lower nasal quadrant in the left visual field was preserved.

Computed tomography (CT) scanning of the brain showed a large hyperdense mass in the suprasellar region (Figure 1). The tumour extended into both the

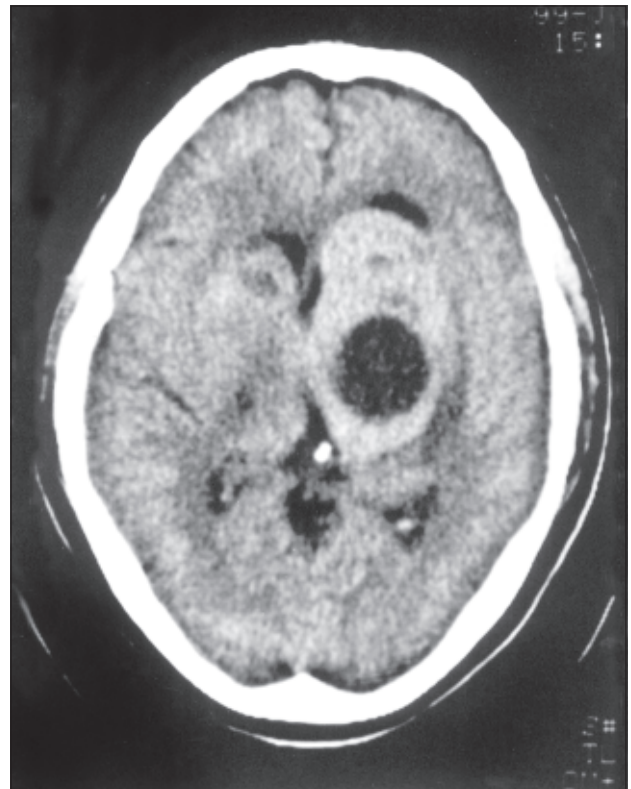


Figure 1. Non-contrast axial computed tomography showing a large suprasellar mass with internal cystic areas.

basal ganglia and the left lateral ventricle, which was enlarged. Cystic areas were noted within the lesion. No calcification was evident. Intense contrast enhancement was noted on the post-contrast scan.

Magnetic resonance imaging (MRI) of the brain showed a large lobulated suprasellar mass, with hypointense signals on T1- and hyperintense signals on T2-weighted

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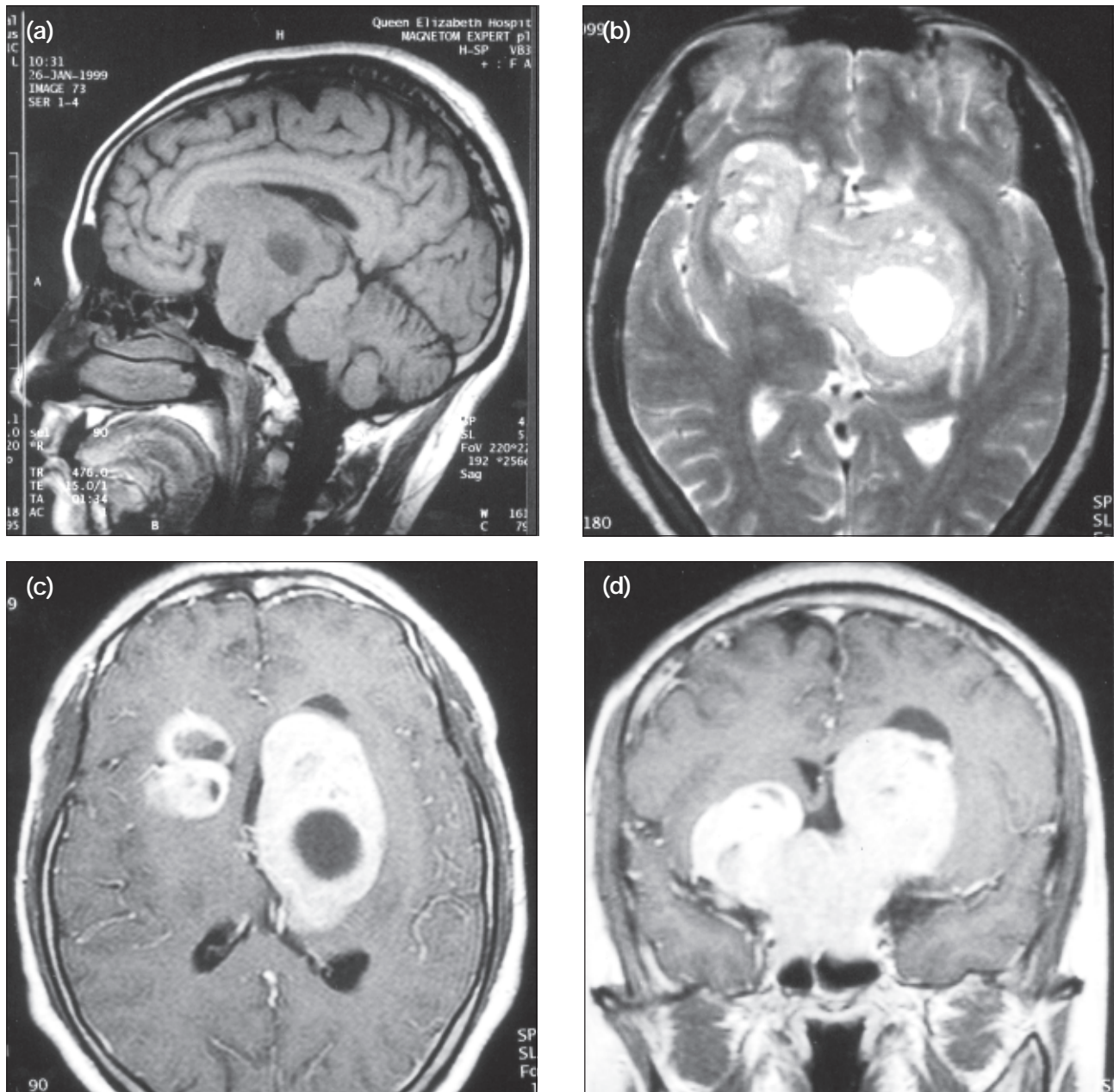


Figure 2. Sagittal (a) T1-weighted, (b) axial T2-weighted, (c) and (d) post gadolinium-enhanced axial and coronal T1-weighted magnetic resonance images, showing a large suprasellar mass with lobulated outline and internal cystic areas. Note the extension inferiorly into the pituitary fossa, and the elevated and compressed 3rd ventricle.

images (Figure 2). Cystic areas were present inside the lesion. Intense contrast enhancement in the solid area was evident. Inferiorly, the mass extended into the sella, which was enlarged and eroded. The 3rd ventricle was elevated and compressed. The basal ganglia on both sides were markedly distorted.

Craniotomy (pterional approach) and resection of the tumour were then performed. Pathological examination of the tumour specimen revealed an ependymoma (Grade II, World Health Organization²) containing sheets

of polygonal cells with a rich delicate vascular network (Figure 3). The tumour cells possessed round and regular nuclei and pale eosinophilic cytoplasm. Perivascular pseudo-rosettes were scattered throughout the tumour. Immunohistochemical findings of positive glial fibrillary acid protein (GFAP) and negative cytokeratin confirmed the glial origin of these cells.

DISCUSSION

The most frequent suprasellar mass seen is suprasellar extension of a pituitary adenoma.¹ Meningioma,

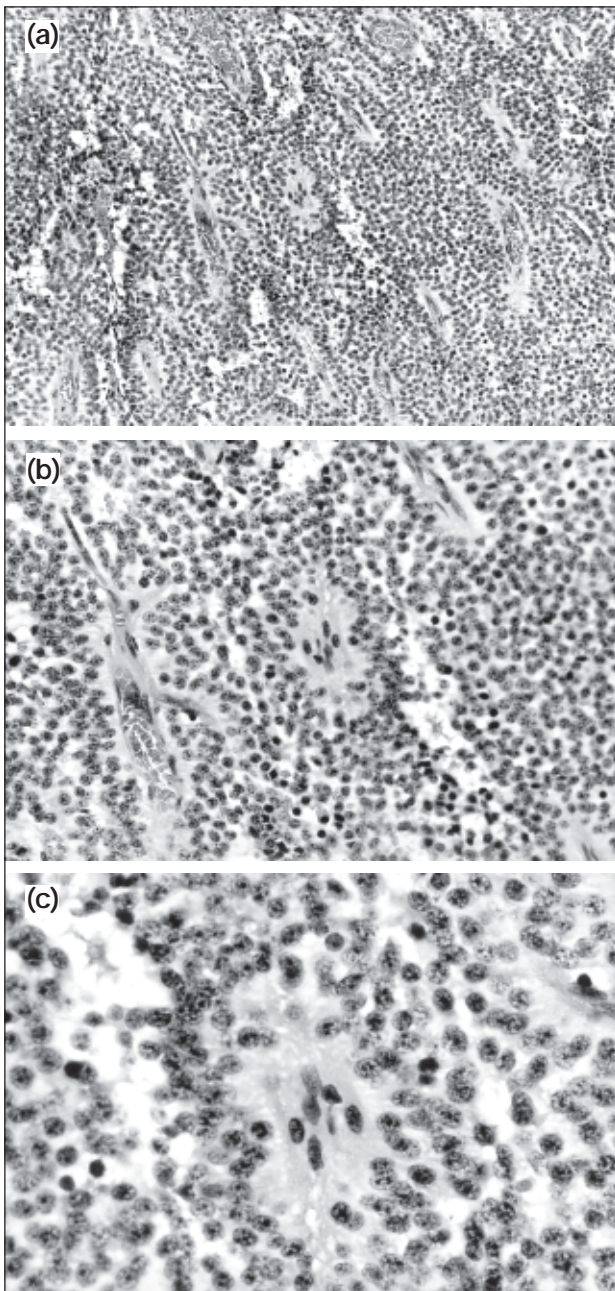


Figure 3. Haematoxylin and eosin stained sections of the ependymoma. (a) The tumour shows moderate cellularity and is punctuated by perivascular nuclear-free zones. (x 100). (b) These nuclear-free zones are so-called perivascular pseudorosettes in which tumour cell processes converge upon the blood vessels. (x 200). (c) The converging cell processes are best seen here. The uniform nuclear appearance is also appreciated. (x 400).

craniopharyngioma, hypothalamic/chiasmatic glioma, and aneurysm follow in varying order. These five lesions are responsible for more than three-quarters of all sellar and juxtaseilar masses. Other less common suprasellar lesions include germinoma, epidermoid and dermoid tumours, lymphoma, leukaemia, and metastases. In both adults and children, ependymoma occurring in this location is considered uncommon,

even though 40% of intracranial ependymomas are found above the tentorium.

Supratentorial ependymomas are typically large, cystic, calcified masses found in children and young adults. They are most commonly found in the extraventricular locations, and in the periventricular region.^{3,4} Armington et al found that supratentorial ependymomas were usually intraparenchymal, larger than 4 cm, and cystic on CT.⁵ The grade of malignancy rises with increasing distance from the ventricular level.⁶

The presentation of an ependymoma in the sella/juxtaseilar region is rare. Winer et al reported a case of intraseilar ependymoma with suprasellar extension.⁷ Donich et al described a case of extra-axial ependymoma at the cerebello-pontine angle with transtentorial extension into the right ambient cistern and cavernous sinus.⁸ Suprasellar ependymoma extending to the pituitary fossa has not been well documented in the literature.

Ependymomas are primary glial tumours presumed to arise from cells related to the ependymal lining. Typically they arise from the ependymal ventricular surface and most commonly occur in the 4th ventricle. The occurrence of ependymomas in an extraventricular region may be attributed to heterotopic ependymal cell rests.⁹ The other possibility is that the tumour may in fact arise from the wall of the ventricle and extend into the brain parenchyma. Ependymomas also typically protrude through the outlet foramina into the adjacent cisterns. This is referred to as desmoplastic development or 'plastic' growth pattern.¹⁰ When ependymomas occur in the 4th ventricle, with this 'plastic' nature, they tend to protrude through the foramen of Magendie into the cisterna magna and through the foramina of Luschka into the cerebello-pontine angle cisterns.

In this report, we described a case of an ependymoma presenting with clinical and radiological features typical of a suprasellar mass. The presence of either embryological remnants or heterotopic ependymal lining cells within the pituitary stalk or the sella may explain the unusual location of this tumour. The other possibility is that the tumour might have arisen from the wall of the 3rd ventricle, and then grown in a desmoplastic manner through the suprasellar cistern into the pituitary fossa.

Supratentorial ependymomas are hyperdense on non-contrast enhanced CT scanning and are usually mixed

lesions with the low densities suggesting cystic or necrotic portions.¹¹ Most tumours are well-demarcated and demonstrate moderate to marked enhancement after the intravenous administration of contrast material.⁵ Intratumoral calcification is present in one-third to 80% of the cases,³ while hydrocephalus and peritumoural oedema are noted in 50% of cases.^{5,11}

With multiplanar imaging capability, particularly in the sagittal and coronal planes, MRI is invaluable in assessing the location and routes of extension of ependymomas. MRI is also useful to characterise the various components of ependymomas. In the series reported by Spoto et al, cystic components were found in two of the four supratentorial ependymomas, while all of the six infratentorial tumours were completely solid.¹² Solid ependymomas and the solid components of mixed solid and cystic tumours were iso- to hypointense relative to normal white matter, and hyperintense to cerebrospinal fluid (CSF) on T1-weighted images. They were hyperintense relative to normal white matter on T2- and proton-density-weighted images. Compared with CSF, the solid tumours were hypo- to hyperintense on T2-weighted images and iso- to hyperintense on proton-density-weighted images. The signal intensity of the cystic components was similar to that of CSF on T1-weighted images and iso- to hyperintense relative to CSF on T2-weighted images. Furie and Provenzale have reported similar findings in imaging studies.³ Signal heterogeneity with scattered foci of increased or diminished signal intensity on T1- and/or T2-weighted images within the solid components could be related to intratumoural bleeding,¹² though this is infrequent in ependymomas.^{5,9}

MRI is also useful to demonstrate tumour vascularity, as well as encasement and displacement of normal vessels. On gadolinium-diethylenetriaminepentaacetic acid enhanced T1-weighted images, the tumour margins become well-delineated. This sign helps to distinguish the enhancing tumour from the non-enhancing surrounding oedema and normal brain tissue.¹² The MRI signal characteristics of ependymomas are non-specific,

however. The location and mode of spread provide more helpful information in distinguishing ependymomas from other types of gliomas. MRI is also less sensitive than CT in detecting tumour calcification.¹²

In conclusion, this case illustrates an ependymoma occurring in the suprasellar and sellar regions. Ependymoma, though unusual, has to be considered in the differential diagnosis when a suprasellar glioma-like tumour extends into the pituitary fossa.

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