Pilocytic astrocytoma mimicking craniopharyngioma: a case series

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ABSTRACT

Although they are one of the most common brain tumours in childhood, pilocytic astrocytomas are rare in adulthood. We report a series of three pilocytic astrocytomas presenting as an intra/suprasellar mass in the fourth to sixth decades of life. All three radiologically mimicked the appearance of a craniopharyngioma, thus representing a potential radiological and diagnostic pitfall. In this series all three patients underwent successful operative management and have remained well throughout follow-up.

The majority of information concerning pilocytic astrocytomas is derived from paediatric populations and so only limited and often conflicting data exist for pilocytic astrocytomas developing in adulthood. Decisions regarding treatment of pilocytic astrocytomas in adults must be made on a multidisciplinary, case-by-case basis.

KEYWORDS

Pilocytic astrocytoma – Craniopharyngioma – Trans-sphenoidal – Suprasellar

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Pilocytic astrocytomas were originally described by Harvey Cushing in 1931.¹ They are the most common glial cell neoplasm in children and the most common paediatric cerebellar tumour, accounting for up to 25% of all brain tumours seen in paediatric neurosurgical practice.^{1,2} Peak incidence is during the first two decades of life and they typically arise in deep, midline structures such as the cerebellum, brain stem, optic nerve and infundibulum, whereas lobar tumours are rare.^{1,3} In late adolescence and early adulthood they are usually found in the vicinity of the third ventricle and it is rare for them to present in adults after the third decade.¹

Pilocytic astrocytomas are notable for their indolent behaviour but can still cause symptoms, which vary depending on the tumour's location. Cross-sectional magnetic resonance imaging (MRI) may demonstrate the classical appearance of a circumscribed, enhancing mural nodule with a non-enhancing cystic component.¹ Histologically, pilocytic astrocytomas are composed of loose glial tissue punctuated by numerous vacuoles, microcysts and compacted piloid tissue forming characteristic Rosenthal fibres.¹ Malignant transformation to a high grade tumour is rare.¹

Macroscopic total surgical resection remains the treatment of choice⁴ with post-surgical reports demonstrating a five-year progression-free survival rate of 75%.^{3,5} Radiotherapy and chemotherapy are reserved for cases of progressive and recurrent tumours, with chemotherapy as the preferred alternative in younger patients. Overall, prognosis is generally good, with a 10-year and 20-year survival rate of 94% and 79% respectively.¹

Case history 1

A 39-year-old woman presented with a progressive 6-week history of short episodes (1–5 minutes) of bitemporal visual disturbance resulting in a permanent right-sided hemianopic deficit that was confirmed on formal perimetry testing. Visual acuity was 6/9 on the left and 6/18 on the right. There were no other neurological symptoms. MRI revealed an irregular, heterogeneous suprasellar lesion with irregular ring enhancement and an area of T2 hyperintensity consistent with central necrosis (Figs 1 and 2). The optic chiasm could not be identified as an isolated structure on imaging and the pituitary gland appeared normal. The images were thought initially to suggest either a craniopharyngioma or a glioma of the optic chiasm.

The tumour was approached operatively via a fronto-orbito-zygomatic craniotomy. The tumour margin was indefinable intra-operatively and complete excision was therefore not possible. Formal histopathology indicated a low grade glioma and glial fibrillary acidic protein (GFAP) staining in the perinuclear cytoplasm confirmed the diagnosis of pilocytic astrocytoma (World Health Organization [WHO] grade 1). Unfortunately, no improvement in visual impairment has occurred but serial imaging over the subsequent ten years has demonstrated no evidence of residual tumour progression.

Case history 2

A 56-year-old man presented with a 5-month history of worsening frontal headaches accompanied by dysphasic

Figure 1 Coronal T2 fluid attenuated inversion recovery image of this 39-year-old female illustrating a large suprasellar mass. In this case the lesion appears relatively homogenous with fluid attenuation.

Figure 2 Axial T2 weighted image of this 39-year-old female corroborating the large suprasellar mass with variable homogeneity, demonstrating a classic hyperintense cystic component

Figure 3 Coronal T1 weighted, contrast enhanced image of this 56-year-old male revealing a cystic ring enhancing suprasellar lesion. Close inspection indicates fluid debris in the cyst consistent with the radiological appearances of a craniopharyngioma.

episodes and non-specific visual disturbances. MRI revealed an irregular cystic mass in the suprasellar cistern, accompanied by nodular rim enhancement. The mass was separate from the pituitary gland but appeared to be in continuity with the pituitary stalk and extending superiorly into the hypothalamus as well as causing effacement of the third ventricle and mild obstructive hydrocephalus (Figs 3 and 4). A radiological diagnosis of craniopharyngioma was reached.

Debulking was performed via a trans-sphenoidal approach and tumour histopathology revealed hypercellularity, mild nuclear atypia and presence of loose reticulated areas. Positive GFAP staining and the presence of dense piloid glial tissue confirmed the diagnosis of pilocytic astrocytoma (WHO grade 1). The patient's immediate post-operative course was complicated by acute deterioration of his visual acuity secondary to a tumour bed haematoma that was successfully evacuated surgically, resulting in an improvement in his visual deficit. However, persistent diabetes insipidus ensued, requiring replacement therapy. The post-operative imaging has remained stable with no recurrence of symptoms.

Case history 3

A 45-year-old woman presented with a 5-month history of right-sided visual impairment associated with a 2-month history of mild headache, weight increase and menstrual irregularity. Ophthalmic assessment revealed a bitemporal field defect worse on the right and diplopia on left upward

Figure 4 Axial T2 weighted magnetic resonance imaging of this 56-year-old male revealing a high-intensity suprasellar mass

gaze. Ophthalmoscopy demonstrated a pale right optic disc whereas MRI displayed a 2cm suprasellar, mixed intensity mass on T1 weighted images with focal areas of enhancement, causing compression of the optic chiasm and the right optic nerve (Figs 5 and 6). The lesion contained areas of calcification and appeared to be separate from the pituitary gland. The tumour was difficult to classify radiologically and the differential diagnoses included optic nerve glioma. However, a diagnosis of craniopharyngioma was felt to be the most likely.

Trans-sphenoidal tumour resection was performed as it was felt that this would give the easiest and best access to the lesion. It resulted in complete resolution of the patient's visual disturbance. Histopathological analysis revealed GFAP positivity and mixed density piloid tissue, diagnostic of a WHO grade 1 pilocytic astrocytoma.

Discussion

We have presented three cases of pilocytic astrocytoma in adulthood, all diagnosed and treated at the same centre and by the same surgeon within five years of each other. All three patients presented later than the typical age of presentation, with one patient in each of the fourth, fifth and sixth decades. All three cases had clinical, radiological and macroscopic appearances consistent with a diagnosis of craniopharyngioma and were only confirmed to be pilocytic astrocytomas following formal histopathological review.

Figure 5 Coronal T1 weighted image with contrast of this 45-year-old female revealing a mixed intensity suprasellar mass with a high signal, particularly in the suprachiasmatic region

Figure 6 Sagittal T1 weighted, contrast enhanced image of this 45-year-old female revealing a high intensity suprasellar mass with an irregular border, compressing the optic chiasm and the right optic nerve

In all three cases the sellar/suprasellar location of the pilocytic astrocytoma contributed significantly to the confounding diagnostic difficulty. The vast majority of craniopharyngiomas (94–95%) have a topologically suprasellar

component (20–41% purely suprasellar, 53–75% both suprasellar and intrasellar) whereas only a very small proportion are found outside this vicinity.³ There are reports of polycystic astrocytomas presenting in adults in unusual locations such as the trigeminal root entry zone.^{6,7} The paucity of infrasellar craniopharyngiomas can potentially be explained by the theory of Erdheim, which states that such lesions can arise anywhere along the craniopharyngeal canal.^{4,5} Our three cases mimicked craniopharyngiomas in terms of their suprasellar location and their characteristics as space occupying lesions with variable heterogeneity demonstrating fibrotic, cystic and calcific components.

The incidence of pilocytic astrocytomas has been estimated as 4.8 per million per year,⁵ only 10–17% of which arise in patients over 30 years of age.⁸ In rare cases, these tumours have been reported in the elderly population.⁹

The majority of information concerning pilocytic astrocytomas is derived from paediatric populations and so only limited and often conflicting data exist for pilocytic astrocytomas developing in adulthood. There are examples of spontaneous regression and a small number of studies describe favourable outcomes and prognosis in adults following both total and subtotal surgical resection.^{8,10} However, alternate authors suggest a less benign course in adulthood with a far higher incidence of tumour recurrence, progression and death than previously reported.^{2,11} This variation may reflect the small patient numbers in many of these studies but it could also represent the potential misdiagnosis of malignant pilocytic astrocytomas as WHO grade 3 anaplastic astrocytomas, excluding these more aggressive tumours from series demonstrating a benign prognosis.²

Survival rates of 95.8% at 10 years after macroscopic total resection have been reported,⁵ and accurate diagnosis and staging of pilocytic astrocytomas is therefore essential in order to achieve such favourable post-operative prognoses and symptom control.⁴ Furthermore, complete resection has been shown to correlate with improved progression free and overall survival.⁴

Conclusions

Although it seems logical that complete resection remains the primary objective when planning surgical intervention, it is clear that there is a paucity of data relating to prognosis and progression in pilocytic astrocytomas of the suprasellar region. The optimal management of suprasellar pilocytic astrocytomas, particularly where they involve eloquent structures such as the hypothalamus or chiasm, remains an area of clinical judgement. Decisions regarding treatment of pilocytic astrocytomas in this unusual location must therefore be made on a multidisciplinary, case-by-case basis.

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