Acta Neurochir (Wien) (2002) 144: 389–393

Acta Neurochirurgica

Surgical Treatment of a Mixed Pineocytoma/Pineoblastoma in a 72-Year-Old Patient

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Summary

Background. Although pineal parenchymal tumours are very rare in elderly patients, we recently successfully treated a 72-year-old male patient. Interestingly, the histology of his pineal parenchymal tumour was mixed pineocytoma/pineoblastoma, which is reported to be extremely rare in aged patients. We present his clinical manifestations, follow-up MRI, surgical treatment, pathological findings, and review the literature.

Clinical Material. This 72-year-old man had a mass in the pineal region detected 3 years previously on MRI in February 1996 following symptoms of headache and vertigo. Two years later, he experienced gait disturbance and disorientation. CT scans disclosed obstructive hydrocephalus, and ventriculo-peritoneal shunt placement was then performed. The tumour mass began to enlarge in July 1999 (at age 72). On October 13, 1999, total removal of the pineal region tumour was performed through an occipital transtentorial approach. The postoperative course was uneventful. The pathological diagnosis of the tumour was mixed pineocytoma/pineoblastoma.

Conclusion. Pineal parenchymal tumours are uncommon in elderly patients, and mixed pineocytoma/pineoblastomas are particularly rare. We followed this patient closely for more than 3.5 years and finally performed total surgical removal of the tumour, with excellent outcome. The present case suggests that a mixed pineocytoma/pineoblastoma tumour is controllable even in elderly patients through careful evaluation and management.

Keywords: Pineal parenchymal tumour; pineocytoma; mixed pineocytoma/pineoblastoma; pineoblastoma.

Introduction

Among pineal parenchymal tumours, pineocytomas and pineoblastomas are rare, accounting for only 0.2% and 0.1% of all intracranial neoplasms, respectively. Pathologically, pineal parenchymal tumours are now divided into three groups; pineocytomas, pineoblastomas, and mixed pineocytoma/pineoblastomas [7, 16]. The latter exhibit elements of both pineocytoma and pineoblastoma, and often occur in young males [11]. Diagnosis of patients after the age of 55 is reported to be extremely rare [5, 15]. We report here a patient with a pineal parenchymal tumour which suddenly enlarged after a 3.5-year follow-up period. We discuss the clinical course and treatment of this rare pineal region tumour.

Case Report

A 72-year-old man with a 3.5-year history of a pineal region mass visited our hospital in October 1999 due to sudden enlargement of the tumour mass detected during follow-up. At the age of 69, he had presented with symptoms of vertigo and headache and was diagnosed with pineal region tumour on MRI examination (Fig. 1A). He had since been regularly followed in our outpatient department. In January 1998, dementia and episodes of unconsciousness occurred (Fig. 1B). Under the diagnosis of obstructive hydrocephalus, ventriculo-peritoneal shunt placement was performed. The postoperative course was uneventful until September 1999, when follow-up MRI studies disclosed sudden tumour enlargement (Fig. 2A). He was re-admitted to our department.

On admission, physical and neurological examinations were normal except for pulsating headache. Tumour markers including α-fetoprotein and β-subunit of human chorionic gonadotropin were negative in both serum and cerebrospinal fluid. T1-weighted MRI revealed an iso-intense tumour which was enhanced heterogeneously by gadolinium-diethylenetriaminepenta-acetic acid, while T2-weighted images revealed a high-intensity mass with cyst formation (Fig. 2A,B,C).

On October 13, 1999, the pineal tumour was totally removed through an occipital transtentorial approach. Surgical procedure was carried out as described in our previous paper using bipolar coagulation and suction, taking much care not to injure deep veins and surrounding brain tissues [13]. The cyst content was mucinous and xanthochromic fluid. Following aspiration of the cyst content, a whitish, elastically soft, solid tumour was excised. Tumour invading the thalamus bilaterally was also totally resected. Total removal could be clearly confirmed under a microscopic view. Histologically, the tumour exhibited diffuse pattern of growth with an indefinite lobular structure. Tumour cells were composed mainly of compactly

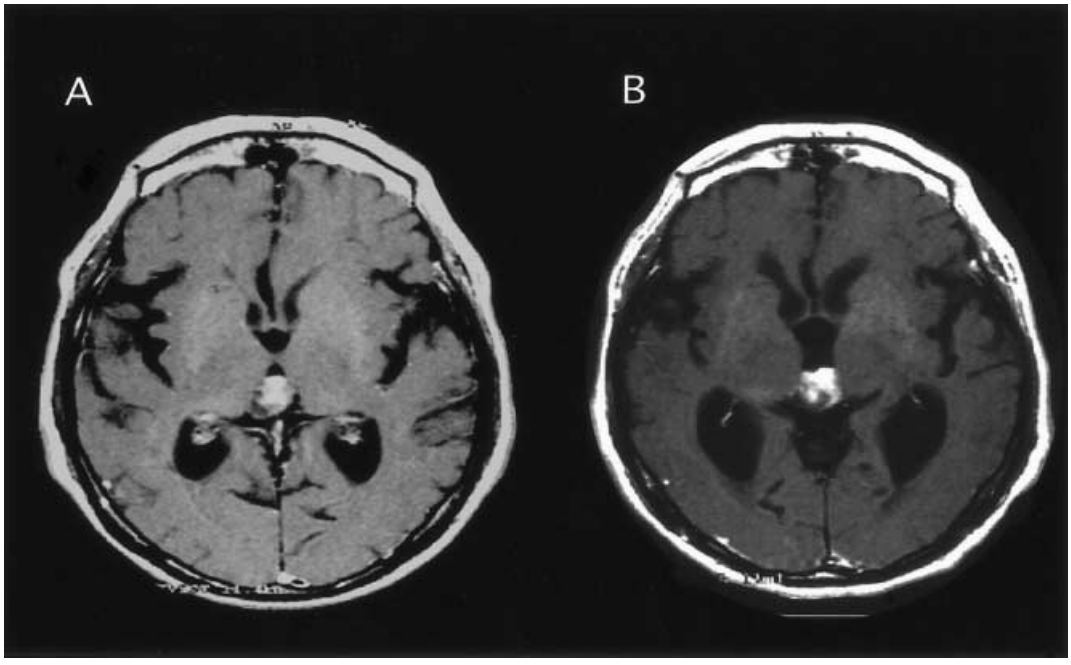

Fig. 1. Magnetic resonance images in February 1996 at the age of 69 (A) and in January 1998 when the obstructive hydrocephalus was noted (B). However, the size of the tumour remained almost unchanged

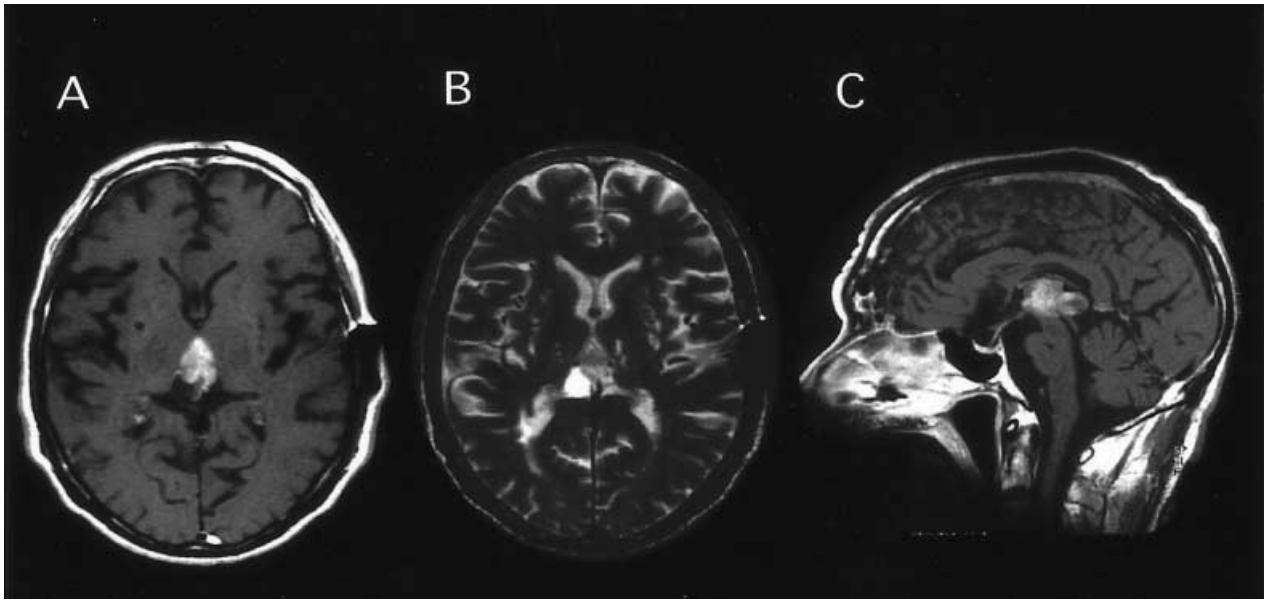

Fig. 2. Figures A, B, and C are Magnetic resonance images on admission. T1-weighted images with gadolinium-diethylenetriaminepenta-acetic acid administration, the axial view (A) and the sagittal view (C), the tumour shows heterogenous enhancement estimated as 3 cm in the length. T2-weighted image (B) shows a high intensity area indicating a cystic component of the tumour

arranged small cells with hyperchromatic oval nuclei, although foci of cells with large nuclei and coarse chromatin pattern were observed. Scattered intercellular fine fibrous anucleic fields were present. Small tumour cells surrounding the anucleic fields exhibited Homer-Wright rosettes, while incomplete pineocytomatous rosettes were also found (Fig. 3). Immunohistochemically, staining for neuron-specific enolase was positive but that for glial fibrillary acidic protein was negative. Additionally, approximately 15% of the tumour cells revealed positive staining for Ki-67. The final diagnosis was therefore mixed pineocytoma/pineoblastoma.

He was discharged 3 weeks after the operation. MRI study performed on February 16, 2001, 16 months after surgery revealed no

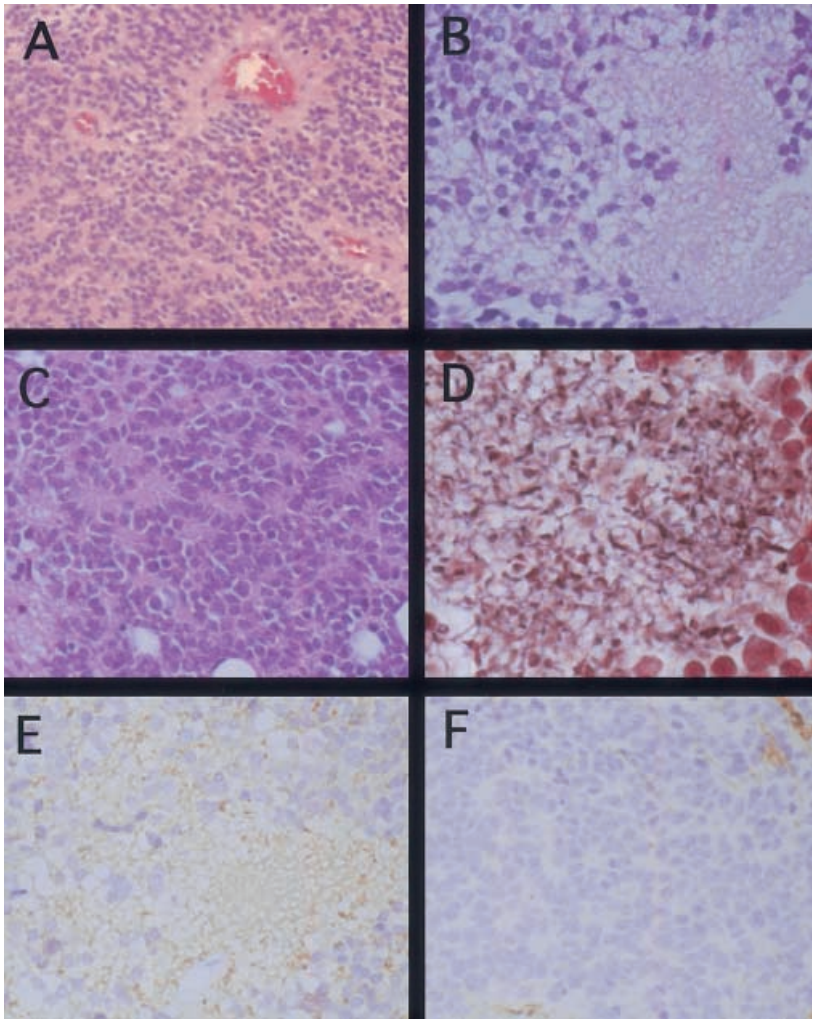

Fig. 3. Figure 3A, 3B and 3C show the Hematoxylin and Eosin stain of the resected tumour. Pineocytomatous rosette (Fig. 3B) and Homer-Wright rosette (Fig. 3C) can be observed. Figure 3D is the Bodian stain showing the cell processes with club-shaped terminal expansion. Figure 3E is immunohistochemical stain of neurofilament protein (NFP), showing positive stain at the cell processes. Figure 3F is immunohistochemical stain of glial fibrillary acidic protein (GFAP), showing the negative stain of the tumour. Original magnification $\times 100$ (Fig. 3A, 3B, 3C, 3E, 3F) and $\times 200$ (Fig. 3D)

recurrence (Fig. 4). Until now June, 2001, the patient is in good condition with no neurological deficits or abnormal CT findings for more than one and a half years after the tumour resection.

Discussion

Before 1983, pineal parenchymal tumours were divided into only two types; pineocytomas and pineoblastomas. However, since Hassoun reported the existence of a type intermediate between the above two, mixed pineocytoma/pineoblastoma has been widely accepted as disease entity in the literature [1, 4]. In the recently published edition of WHO's histological classification of central nervous system tumours, which we used for diagnosis of this case in October 1999, parenchymal tumours were newly divided into three types; pineocytoma, pineoblastoma, and mixed pineocytoma/pineoblastoma [7]. Our patient had the histological findings intermediate in type between pineocytoma and pineoblastoma. To our knowledge, this is the oldest patient to be reported to have a mixed pineocytoma/pineoblastoma [2, 6]. The newest version of WHO classification of 2000, however changed the name mixed pineocytoma/pineoblastoma to be pineal parenchymal tumour of intermediate differentiation [8].

Parenchymal tumours in the pineal region account for less than 1% of all intracranial neoplasms [3, 10]. Of these, 22% of pineocytomas and 42% of pineoblastomas are diagnosed in paediatric patients, while almost all other such tumours are found in young adults [15]. Pineal parenchymal tumours in older patients (over 70 years old), as in the present case, are very uncommon [2, 6]. Although the treatment evaluated by a long-term follow-up study has been nearly established for young adult and paediatric patients,

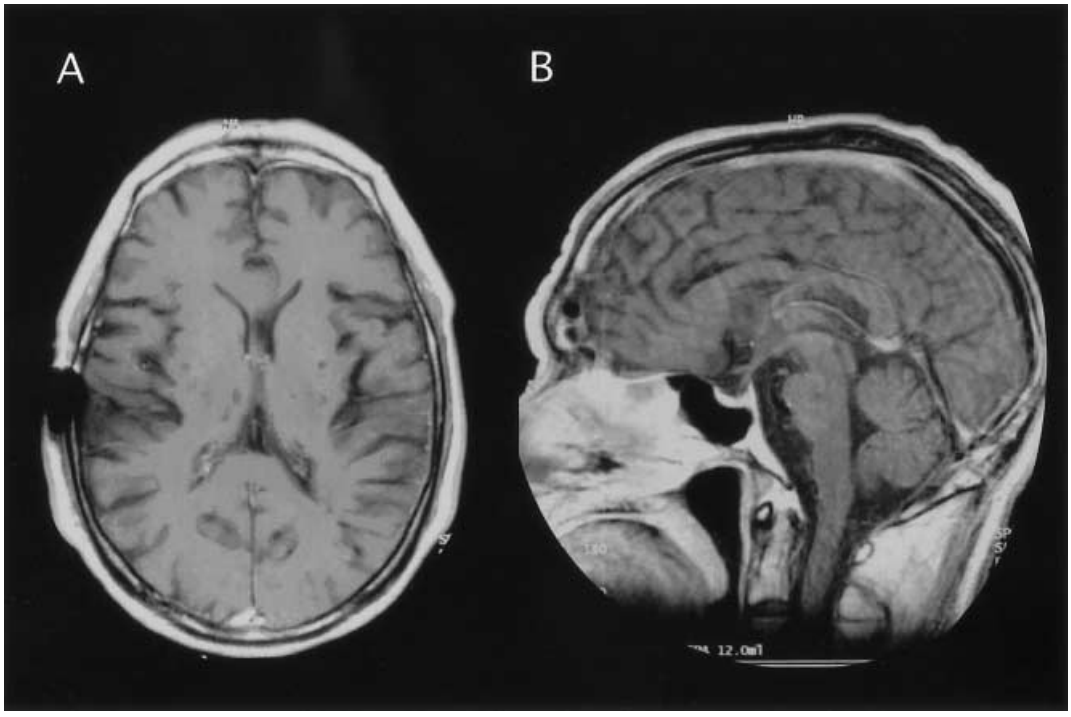

Fig. 4. T1-weighted images with gadolinium-diethylenetriaminepenta-acetic acid administration at 16 months after the operation, the axial view (A) and the sagittal view (B). No recurrent tumour mass can be seen

little is known concerning the treatment of pineal region tumours in elderly patients [9, 12]. During the past decade, surgery has become the most important modality in the treatment of pineal region tumours, particularly in young adults and paediatric patients, since only surgery and histological examination of surgical specimens can confirm the diagnosis of pineal region tumours and enable further decisions regarding treatment with irradiation and/or chemotherapy [13, 14]. Nevertheless, no reports have indicated the ideal treatment in the elderly with pineal region tumours. In the present case, given the patient's age, direct surgical intervention was attempted to completely remove the tumour by surgery alone without postoperative adjuvant therapy using radiotherapy or chemotherapy. Although it is difficult to determine therapeutic strategy for the treatment of pineal region tumours in elderly patients, the present case suggests that keen observation, regular follow-up and total excision of pineal parenchymal tumour is a reasonable approach to treatment. For symptomatic, aged patients, the present findings also suggest that mixed pineocytoma/pineoblastoma is controllable if thorough study and total removal are performed by an able surgical team.

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