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Clinical Study

A retrospective study of primary cerebellar glioblastoma multiforme in adults

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ABSTRACT

Primary cerebellar glioblastoma multiforme (GBM) is a rare tumour in adults that accounts for less than 1% of all patients with GBM. In view of their rarity, the pathogenesis and prognosis of cerebellar GBM are not yet completely understood. The aim of this study was to retrospectively analyse patients with primary cerebellar GBM treated in our institute over a period of 10 years. Data from the case records of five adult patients with cerebellar GBM was evaluated and their outcome was assessed. We observed local failure in patients who reported back with recurrence. The presence of brainstem infiltration was a significant factor influencing progression-free survival. The overall prognosis was worse than for patients with supratentorial GBM. In view of their rarity, a meta-analysis is required to assess the pathogenesis and prognostic factors affecting overall survival in patients with cerebellar GBM.

1. Introduction

Glioblastoma multiforme (GBM) is the most common primary central nervous system tumour in adults, comprising approximately 50% of all primary intracranial tumours. Primary cerebellar GBM, however, is rare in adults and accounts for less than 1% of all patients with GBM.[1–8] The reason for its rarity is not completely understood. Many reported studies have included a few patients (less than eight) from single institutions. Well-defined protocols and prognostic factors have been defined for supratentorial GBM. Clinical data regarding primary cerebellar GBM are scarce, and to our knowledge the largest series reported is a multi-institutional report of 45 patients.[9] As a result, the outcome of these patients remains somewhat unclear. Some authors have reported a poorer prognosis for patients with cerebellar GBM, with a survival of 3 months to 7 months,[10,11] whereas others have observed a similar survival to patients with supratentorial tumours.[12,13]

We retrospectively analysed patients with histologically proven primary cerebellar GBM from a single institution from March 2001 to March 2011 and discuss the pathogenesis, radiology, pathology, and treatment.

2. Materials and methods

We reviewed patients with cerebellar GBM who had undergone surgery during the period of March 2001 to March 2011. The inclusion criterion was histologically proven GBM (World Health Organization Grade 4) in an adult patient (aged >18 years). Those with supratentorial or brainstem GBM were excluded. Five patients satisfied our criteria and were included for analysis. Using a standardized data sheet, clinical, radiological and surgical data were collected from medical records. Preoperative and postoperative MRI and/or CT scans were reviewed to ascertain the imaging diagnosis and determine the extent of tumor resection.

Descriptive statistics were used to summarize the patient demographic and clinical characteristics. Overall survival (OS) was defined as the time from the date of diagnosis to the date of death or last follow-up. Progression-free survival (PFS) was defined as the time from the date of diagnosis to the date of progression, death or last follow-up.

3. Results

Patient characteristics are detailed in Table 1. All patients presented with gait ataxia. Other symptoms included headache, vertigo and vomiting. Neurological examination revealed papilledema in three (60%) patients and cerebellar dysfunction in four (80%) patients. The radiologic work-up included a brain MRI in all five patients, which revealed the tumour characteristics. In four (80%) patients the tumour was localised to the cerebellar hemisphere and in one (20%), it was vermian in origin. No synchronous regional or distant metastasis was observed. There was no evidence of leptomeningeal spread. Brainstem invasion (with the main tumour within the cerebellum) was observed in one (20%) patient.

All patients underwent gross total resection of the tumour and were referred postoperatively to another centre for adjuvant radiotherapy and chemotherapy. All patients received steroids during

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Table 1
Patient characteristics, clinical data and treatment details of five patients with cerebellar glioblastoma multiforme (GBM)

Parameter	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age (years)	60	56	48	23	61
Sex	M	M	M	F	M
KPS score	80	90	100	90	100
Symptoms	GA, vertigo, headache	GA, headache	GA, vertigo	GA, headache	GA
Signs	Papilledema, cerebellar signs	Papilledema	Cerebellar signs	Papilledema, cerebellar signs	Cerebellar signs
Tumour location	Cerebellar hemisphere	Cerebellar hemisphere	Cerebellar hemisphere	Vermis	Cerebellar hemisphere
Brainstem infiltration	Yes	No	No	No	No
CRT (with adjuvant CT)	No	No	No	Yes	Yes
Adjuvant CT after RT	Yes	Yes	Yes	No	No
Survival (months)	9	18	15	On follow-up	On follow-up
PFS (months)	8	15	12	9	8

CRT = concurrent chemoradiation therapy, CT = chemotherapy, GA = gait ataxia, KPS = Karnofsky performance scale score, PFS = progression-free survival, RT = radiation therapy.

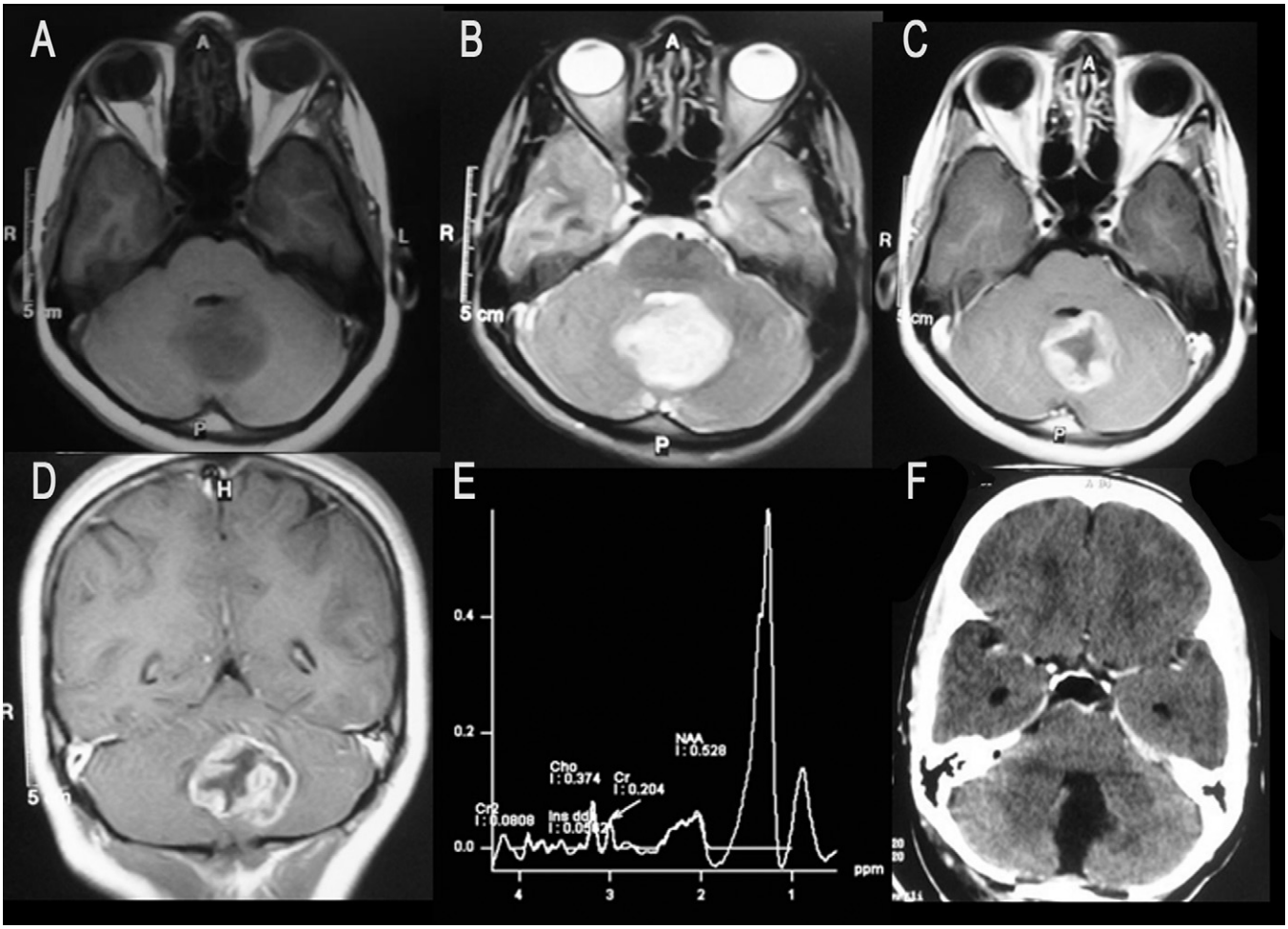

Figure 1. (A) Axial T1-weighted MRI showing a hypointense glioblastoma multiforme (GBM) involving the vermis with minimal compression of the fourth ventricle. (B) The lesion appears uniformly hyperintense on axial T2-weighted MRI without any significant peritumoural edema. (C) Axial post-contrast and (D) coronal post-contrast T1-weighted MRI showing enhancement of the tumour wall with a central non-enhancing necrotic component. (E) Magnetic resonance spectroscopy (MRS) demonstrating the characteristic high choline, low N-acetylaspartate and high lactate peak of GBM. (F) Postoperative post-contrast axial CT scan showing gross total removal of the tumour.

irradiation. No craniospinal prophylactic irradiation was delivered. Two patients (40%) received concurrent chemoradiation therapy (CRT) as per the Stupp protocol[14] and the remaining three (60%) received radiation therapy followed by chemotherapy. Chemotherapy given concurrently was temozolomide (TMZ). Adjuvant agents included procarbazine, carboplatin and vincristine (PCV) and TMZ-containing regimens.

The median follow-up time was 9 months (range 6–15 months). Three patients (Patients 1, 2 and 3) progressed and opted for hospice in view of poor Karnofsky Performance Scale scores or non-willingness to undergo any further treatment. Patient 1 had the shortest time to recurrence (8 months) and showed evidence of brainstem infiltration at the time of surgery. On contacting the relatives of these patients as a part of this study, we found out that