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LETTER TO THE EDITORS

Gliomatosis cerebri presenting as a recurrent cervical myelopathy

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Dear Sirs,

Gliomatosis cerebri (GC) is a rare diffuse neoplastic infiltration of the brain involving more than two lobes and, occasionally, the brainstem and spinal cord [1]. Due to the preservation of surrounding anatomical architecture and nonspecific clinical presentation, GC can be confused with central nervous system (CNS) inflammatory diseases, stroke and leukoencephalopathy. We describe here a patient with GC who presented with a recurrent cervical myelopathy.

A 36-year-old woman was admitted due to a 2-month history of progressive tetraparesis, urinary retention, constipation and dysphagia. Her medical history revealed a previous episode of acute tetraplegia 14 months before, after a fall at home. At that time, a brain and spinal cord MRI scan showed a lesion hyperintense on T2-weighted and FLAIR sequences extending from the midbrain down to the C5 level that was considered to be secondary to the trauma. No contrast enhancement was observed. She was admitted to another hospital, treated with high-dose steroids and gradually improved. Ten months later, at the time of recurrence of the symptoms, she was able to walk unassisted and do the housework. There were no other systemic complaints. The neurological examination revealed a downbeat nystagmus, bilateral facial hypoesthesia, palatal weakness and an asymmetric spastic MRC III/IV tetraparesis with increased deep tendon reflexes and extensor plantar responses. A new MRI scan showed a T1-weighted isointense and T2-weighted and FLAIR hyperintense non-enhancing lesion at the thalamus, extending to the brainstem and the spinal cord at the C5 level. Smaller lesions were also observed in the cerebellar vermis and left hemisphere (Fig. 1). CSF analysis was normal. She was initially diagnosed with a recurrent myelitis and received high-dose methyl prednisolone. A week later, she became tetraplegic and developed respiratory failure, and was transferred to the intensive care unit. A work-up for infectious and autoimmune diseases, including antiaquaporin 4 antibodies did not reveal any abnormalities. Plasmapheresis was performed without improvement. Nineteen days after admission, a brain biopsy revealed a grade II astrocytoma (Fig. 2). She received radiation therapy with rapid improvement of weakness and weaning from ventilation and was discharged 2 months later for clinical follow-up.

Before modern brain imaging, GC was diagnosed only at autopsy. It was only in 1987 that the first antemortem diagnosis was described based on the MRI appearance and biopsy [2]. Regardless of the advances in imaging techniques, GC remains a challenging diagnosis due to its low frequency and heterogeneous presentation. GC can be divided into primary (when there is diffuse infiltration at time of presentation) and secondary (when there is a focal lesion confined to one lobe initially with spreading to contiguous structures on follow-up) [3]. Several histological types of GC have been observed. In a large series of 179 patients, 108 patients had an astrocytic tumor, 54 had

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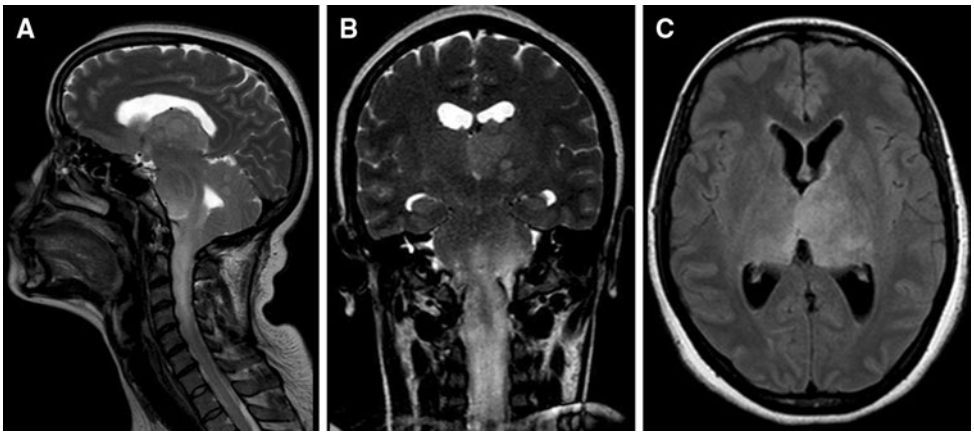

Fig. 1 MR images. Sagittal (**a**) and coronal (**b**) T2-weighted images show a hyperintense infiltrative lesion extending from the thalamus to C5. Smaller lesions are observed in the cerebellar hemisphere. **c** FLAIR image shows bilateral thalamic involvement

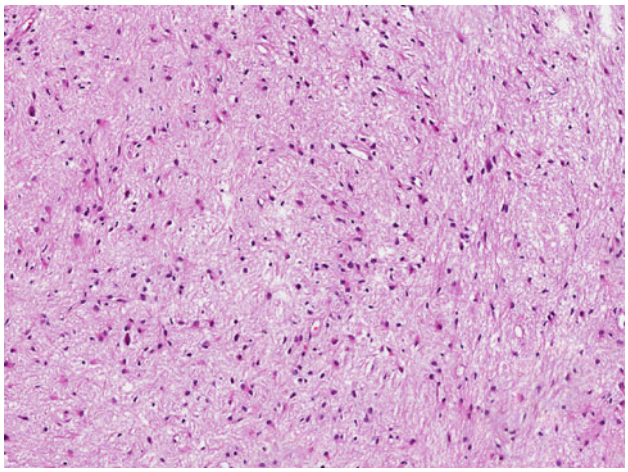

Fig. 2 Histological section showing mild cellular proliferation and atypia suggestive of grade II glioma (hematoxylin and eosin, ×100)

an oligodendrocytic tumor and 17 had a mixed oligoastrocytic tumor. Tumors were considered grade II in 50% of patients, grade III in 40.5% and grade IV in 9.5% [1]. Lesions are found predominantly in the cerebral hemispheres (76%), followed by the midbrain and pons (52%), thalamus (43%) and basal ganglia (34%). The spinal cord is affected in less than 10% of patients [4].

Due to its infiltrative nature, the presentation of GC is usually subtle and dominated by slowly evolving focal signs, cognitive changes and seizures. Symptoms of raised intracranial pressure occur only in one-third of patients despite the extension of the lesions [4]. Since presentation is nonspecific, cases are frequently confounded with other inflammatory neurological diseases. In the series of Taillibert et al., 6.4% of the patients had a diagnosis such as CNS vasculitis, Behçet disease, encephalitis and leukoencephalopathies [1]. In our patient, the relapsing–remitting aspect of the symptoms initially suggested a recurrent cervical myelitis, but the normal CSF, the absence of antiaquaporin 4 antibodies and other autoantibodies and the failure of immunosuppressive/immunomodulatory treatment raised the suspicion of an alternative diagnosis. The MRI appearance can also be misleading, since the typical appearance is a high-signal lesion on T2-weigthed and FLAIR sequences with minimal displacement of structures, which can be observed in many other neurological diseases. Contrast enhancement is present in a minority of patients with GC, varying from 16% to 45% in different series [1, 5]. MR spectroscopy can be helpful and shows elevated Cho/Cr and Cho/NAA in addition to decreased NAA/Cr ratios [5].

Traditionally, the treatment of choice for GC has been radiation therapy, but chemotherapy regimens have been used recently both as adjuvant or upfront, with encouraging results [6, 7]. Overall, prognosis is still poor. In two series, the median overall survival was 12 and 14.5 months [1, 4].

In conclusion, despite its rarity, GC should be included in the differential diagnosis of patients with extensive CNS lesions and relapsing–remitting symptoms.

Conflict of interest None.

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