A difficult diagnosis of gliomatosis cerebri

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Abstract Gliomatosis cerebri, a rare condition, requires clinical, radiological and pathological correlation for diagnosis. Mental and personality changes are the most common presenting symptoms with or without focal neurological signs. The widespread nature of the disease is revealed by CT or MRI. The shape of the brain may be maintained and pathological gross examination may be unremarkable or show hypertrophy without evident tumour. Microscopic examination reveals infiltration of the brain by variably differentiated neoplastic glial cells. We present a patient with gliomatosis cerebri in whom we experienced difficulties with diagnosis. The literature is reviewed and the diagnostic features summarised.

Key words Gliomatosis cerebri · Computed tomography · Magnetic resonance imaging

Introduction

Gliomatosis cerebri is an uncommon condition characterised by diffuse overgrowth of the central nervous glial cells, in varying stages of differentiation, with preservation of the underlying neural structures and without a focal tumour [1]. The criterion for the diagnosis is considered to be involvement of at least two lobes of the brain by small, elongated cells, without necrosis [2–5].

Personality and mental changes are the most frequent symptoms, with or without neurological signs. CT and especially MRI typically demonstrate diffuse expansion of the central parts of the brain, with low density on CT and abnormal signal intensity on MRI, and the findings are stable in the short term [4, 6, 7].

The diagnosis requires radiological-pathological correlation, with multiple biopsies. Otherwise the diagnosis may not be appreciated, because the infiltrative nature of the lesion will not be evident. The prognosis is poor, ranging from weeks to some years after the manifestation of the symptoms. Steroids may be useful in the short term, but chemotherapy is of little value and radiotherapy of questionable benefit [7, 8].

Case report

A 12-year-old boy had been healthy, apart from febrile convulsions. One morning he was confused on waking and had an attack of unconsciousness and convulsions. On admission he was unconscious; the right eye was deviated to the right, but there were no other neurological signs. CT revealed low-density nonenhancing expansion of the thalamus and brain stem. The cerebral sulci were largely invisible and there was mild hydrocephalus due to third ventricular compression (Fig. 1). A deep cerebral infarct was suspected. The EEG was consistent with increased intracranial pressure and cerebral oedema. A ventricular drain was inserted and later a ventriculoperitoneal shunt. The intraventricular pressure was about 25 cm H₂O.

The patient developed exaggerated reflexes with a positive Babinski sign, more marked on the left. The eyes still deviated to the right. MRI 12 days after the first attack showed large pathological brain in the frontal and temporal lobes and basal ganglia; the thalamus was enlarged. These areas gave low signal on T1-
Fig. 1  CT on admission reveals diffuse low-density swelling of the central parts of the brain. The third ventricle is compressed and the sulci are invisible. The lateral ventricles are slightly enlarged.

Fig. 2a, b MRI 12 days after the first symptoms. a T2-weighted image shows high signal with expansion of the thalamus, medial frontal lobes and left insular cortex. b T1-weighted coronal image shows bilateral symmetrical low-signal expansion of the medial temporal lobes and left insular cortex.

Fig. 3 A T2-weighted image 6 weeks after the first symptoms shows the central abnormal area to be the same, but reveals a slight extension of the left insular lesion. The lateral and third ventricles and sulci are slightly larger.

Fig. 4 The last CT before death, 1 year and 9 months after the first attack. The swelling and low density around the third ventricle are less obvious, and ventricles and sulci are slightly larger than on the first CT. This might be due to irradiation or to steroids weighted images and high signal on T2-weighting and did not correspond to vascular territories (Fig. 2). CT 3 weeks after the first study showed that the lateral ventricles had decreased in size; the oedematous areas were slightly smaller.

Cerebral angiography 5 weeks after the first attack showed no evidence of vasculitis. There was no laboratory evidence of encephalitis. Single-photon emission CT 5 weeks after the onset of symptoms revealed a low-perfusion area in the left thalamus, thought to be due to a postencephalitic state. MRI 6 weeks after the first attack was unchanged, except that the abnormal area in the left insular region had enlarged slightly (Fig. 3). Neurophysiological examination showed normal peripheral nerve conduction velocities, excluding a diffuse demyelinating process. Many blood tests were normal, as were cerebrospinal fluid samples. Extensive investigation failed to identify any metabolic or infective cause, although there had been a respiratory infection of some days' duration.

The convulsions were treated with anticonvulsive drugs, antibiotics were given for bronchitis, and dexamethasone was started.