

Our case presented with the clinicopathological picture of acute necrotising herpetic encephalitis. Serological evidence indicated the virus type involved to be HSV II.

This report probably represents a previously unrecognised entity, namely acute necrotising encephalitis due to HSV Type II in a non-immunocompromised adult.

MK BAKER
MA SANDLER
RD BAYNES
S MILLER

Division of Neurology,
Department of Internal Medicine,
H F Verwoerd Hospital,
Pretoria,
Transvaal,
South Africa

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Adult coeliac disease and brainstem encephalitis

Sir: Various neurological disorders have been reported in association with adult coeliac disease.¹⁻⁴ Vitamin E deficiency, gluten toxicity and the HLA-A8-antigen are frequently related to coeliac disease. Their possible influence on the development of neurological disorders has often been stated but has never been clarified in detail.³⁻⁶ The neuropathological findings most commonly included degenerative changes whereas inflammatory lesions were rare.^{1,7,8} In our case the diagnosis of brainstem encephalitis was established by clinical and CSF investigations. Neuropathological analysis revealed an atypical subacute panencephalitis, an uncommon finding in adult coeliac disease.

The 45 year old male patient in whom coeliac disease had been diagnosed from a jejunal surgical specimen in summer 1985, noticed a marked decrease in vitality in November 1985, 2 weeks after he had returned from a trip to Georgia (USSR).

Subfebrile temperature was measured there and in early December 1985 rhythmic jerking of the left arm, the left half of the mouth, the tongue and the soft palate occurred and led to his admission to the neurological clinic on 17 December. Arrhythmic myoclonic movements of the left arm, the tongue and the soft palate as well as slight dysarthria and mild unsteadiness in Romberg's test were found on examination. CSF investigation revealed 20 cells/ml, mostly lymphocytes, and an increase of total protein to 82 mg/dl. Thus, the diagnosis of a brainstem encephalitis was established. However, neither serum nor further CSF investigation could determine viral, bacterial or fungal microorganisms as causative agents. In addition, antibodies against gliadin and a vitamin E concentration below 0.1 mg% were detected in serum. Schilling's test was abnormal indicating a reduced resorption of vitamin B 12. The stools were bulky and foul smelling. Our patient developed right sided ataxia and showed a strong tendency to fall backwards at the beginning of January 1986. Auditory brainstem potentials suggested a pontomesencephalic lesion on the right and nuclear magnetic resonance imaging revealed pontine lesions in the dorsal, medial and right paramedian areas.

In the middle of January, the patient additionally developed peripheral facial palsy on the right, a horizontal gaze paralysis to both sides and exaggerated deep tendon reflexes at the upper and lower extremities. Babinski's sign was positive on both sides. In February 1986, the neurological abnormalities worsened, bulbar disturbances occurred and progressed quickly. Soon after, the patient died from

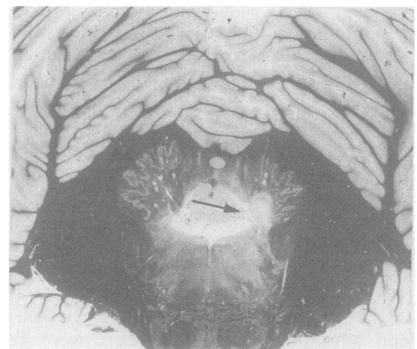


Fig 1 Section of brain stem. (Luxol fast blue for myelin counterstained with nuclear fast red.) Note the partially necrotic irregular periventricular lesions on the right side (arrows) also involving the hilus of the right dentate nucleus.

severe pulmonary embolism. Post mortem investigation grossly showed brain oedema. Irregular periventricular lesions, adjacent to the 4th ventricle extended to the hilus of the right dentate nucleus (fig 1). These lesions consisted of inflammatory necrosis and/or myelin loss. As a consequence of the lesion of the right dentate nucleus the left inferior olive was hypertrophic (fig 2). Additional findings were a fibrous gliosis in the inferior vermis and a prominent lymphocytic infiltration in the mesencephalon and the rostral pons. The neuropathological data indicated an atypical subacute panencephalitis. Again, special stainings for bacterial or fungal microorganisms and immunocytochemical investigations for viral inclusion bodies failed to clarify the aetiology of the panencephalitis.

Considering the clinical and laboratory findings in adult coeliac disease and associated neurological complications the following speculations about the pathogenesis seem to be reasonable. First, might vitamin E deficiency which was also found in our case cause encephalopathy?⁶ Second, might malnutrition which is often found as a result of coeliac disease cause or promote cerebellar disturbances, myelopathy and neuropathy?^{1-3,7,8} Third, is coeliac disease known to be frequently associated with a genetically determined alteration in immune functions?^{2,8} This might be suggested by a high coincidence of coeliac disease and lymphoma, carcinoma and multifocal leucoencephalopathy.⁹ A high susceptibility of the patients for jejunally mediated infection might be due to a deficiency of mucosal IgA antibodies.

The myoclonic movement disorder of our patient is explained by the neuropathological changes in structures that are known as Guillain-Mollaret's triangle.¹⁰

In conclusion, our case of adult coeliac disease does not provide any further clues to

the pathogenesis of the brainstem encephalitis. Our report merely documents an uncommon relationship between coeliac disease and an atypical subacute encephalitis.

THOMAS BRÜCKE*
 HARALD KOLLEGER*
 MANFRED SCHMIDBAUER†
 CHRISTIAN MÜLLER*
 IVO PODREKA*
 LÜDER DEECKE*

Neurologische Universitätsklinik Wien,*
 Lazarettgasse 14, 1097 Wien, Austria.
 Neurologisches Institut der Universität

Wien,†
 Schwarzspanierstraße 17, 1090 Wien,
 Austria.

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Brain stem glioma mimicking progressive basilar artery thrombosis

Sir: Vascular insufficiency in the posterior cerebral circulation often presents with

visual disturbances, ataxia, dysequilibrium, and alternating motor and sensory signs.¹⁻³ The anatomical origin of these symptoms and signs can be confirmed by neurophysiological testing such as brainstem auditory evoked responses (BAERs), and high resolution cranial computed tomography (CT) or magnetic resonance imaging (MRI) of the brain.⁴ The aetiology of the dysfunction can be confirmed by demonstrating diminution of vertebral-basilar flow via angiography.⁵⁻⁷ Diminution or absence of flow, however, is only presumptive evidence as flow may also be impeded by non-atherosclerotic disorders such as extravascular compression due to mass lesions. We report such a case.

A 70-year-old male initially presented with a 3 month history of diplopia, dysarthria, and gait ataxia. Although of rapid onset, the symptoms were progressive. He appeared sallow, but otherwise had a normal general physical examination. His mental status and speech were normal except for a mild scanning dysarthria. Cranial nerve examination showed right medial and bilateral upward gaze palsies. Extraocular movements and gait were ataxic. No other neurological abnormalities were recorded.

Initial CT suggested a sagittal sinus thrombosis and revealed a small basilar artery (fig). Intravenous digital subtraction angiography (DSA) excluded the sinus thrombosis but detail of the posterior fossa circulation was not obtained. Cerebrospinal fluid (CSF) studies were normal. He was discharged with a presumptive diagnosis of brainstem ischaemia after investigation had not shown a systemic malignancy.

Over the next month, his gait ataxia and dysarthria became worse. New problems developed including dysphagia, inability to sit unsupported, and urinary incontinence. On examination, he appeared dehydrated and confused. His speech was slow and ataxic. Gaze palsies persisted as above. Deep tendon reflexes were brisk throughout, with sustained ankle clonus. Bilateral extensor plantar responses were present with upper motor neuron distribution weakness of the right body parts. The patient had marked dysmetria and could not sit unsupported. Repeat CT revealed lucencies in the basal ganglia. BAERs showed prolonged latencies in the left brain stem. Repeat CSF studies were normal. Repeat DSA during aortic arch injection of contrast was consistent with basilar artery thrombosis because of nonfilling. He was unable to cooperate for a cranial MRI study.

The patient developed nausea, vomiting and intractable hiccups, increasing right-sided motor weakness, and somnolence.



Fig 2 Section through inferior olivary nucleus (left). (Bodian's silver method, $\times 775$) showing some prominent dentritic proliferation in neurons.