CLINICOPATHOLOGICAL CASE CONFERENCE

Progressive ataxia, focal seizures, and malabsorption syndrome in a 41 year old woman

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Case presentation

The patient was a right handed woman, born in 1952 and married. She was a radio taxi controller.

In October 1993 she presented to her local district general hospital having had two short lived episodes of involuntary twitching of the left side of her face and then a third episode of left sided facial twitching which was followed by collapse and a generalised tonic clonic seizure. Nine months previously she had been investigated and treated for a right upper lobe pulmonary abscess, from which the causative organisms were not isolated. She had no other relevant medical or family history. She did not smoke. A contrast enhanced cranial CT was unremarkable. She had macrocytic anaemia and abnormal liver function. Further investigations included a liver biopsy, which showed widespread fatty change, and a jejunal biopsy which showed total villous atrophy with crypt hyperplasia. She was established on a gluten free diet and given anticonvulsant drugs, initially phenytoin but this was subsequently changed to carbamazepine after she developed a rash.

Despite anticonvulsant medication, between late 1993 and early 1994 she continued to have intermittent episodes of predominantly left sided facial twitching but no further generalised seizures. Her speech became slurred and her gait increasingly unsteady. In March 1994 examination showed a thin, pale woman. A smooth non-tender liver edge was felt 2 cm below the costal margin. Neurological examination showed repeated episodes of rapid

twitching of the left side of the face and a staccato dysarthria which was exacerbated by the twitching movements of her mouth. There was mild incoordination of the lower limbs but with full power on formal testing, and no sensory abnormalities, normal tendon reflexes, and bilateral flexor plantar responses. She was able to walk unaided but unsteadily.

Her haemoglobin content was 11.1 g/dl with a moderate macrocytosis and a mean corpuscular volume of 104 fl (normal 78-98 fl); a raised platelet count at $609 \times 10^9/l$ (normal $150-400 \times 10^9/l$); and a white cell count of $5.4 \times 10^9/l$ (normal $4-11 \times 10^9/l$). The differential white count was essentially unremarkable although some neutrophils showed toxic granulation and occasional myelocytes were seen on a blood film. Erythrocyte sedimentation rate was 20 mm/hour. The urea, electrolytes, including calcium and magnesium, and plasma glucose were normal. Her liver function tests were deranged with an ALT of 108 U/l (normal 5-59 U/l), alkaline phosphatase 233 U/l (normal 30-140 U/l); γ -GT 159 U/l (normal 6-31 U/l). Thyroid function tests were normal. Serum angiotensin converting enzyme was 106 U/l (normal 37-75 U/l). A T₂ weighted MRI showed multiple areas of high signal scattered diffusely throughout the cerebral white matter (fig 1). There were also abnormal areas of high signal in the midbrain (fig 2). Lumbar puncture showed an opening pressure of 11 cm CSF. No red cells were seen and there was 1 lymphocyte/mm³. Protein in CSF was raised at 0.7 g/l (normal 0.15-0.45 g/l). The CSF glu-

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Figure 1 T2 weighted cranial MRI showing multiple areas of high signal scattered diffusely throughout the white matter.

Figure 2 T2 weighted MRI section showing abnormal area of high signal in the left midbrain.

cose was 3.4 mmol/l (simultaneous plasma glucose 6.1 mmol/l). Cytological analysis of CSF was unremarkable. A chest radiograph suggested scarring in the right hemithorax and thoracic CT confirmed this area of scarring situated posteriorly in the right upper lobe. There was no evidence of cavitation or generalised pulmonary parenchymal disease. Two EEGs were similar and showed "frequent non-convulsive runs of focal seizure discharges". A further jejunal biopsy showed "severe partial villous atrophy" after she had been on a gluten free diet for eight weeks.

The carbamazepine was gradually replaced with clonazepam with reduced facial twitching. She was discharged on a combination of clonazepam and multivitamins including vitamin E, thiamine, and folate.

Although she had seemed initially to respond to clonazepam she required readmission at the end of April 1994 with increasingly frequent episodes of facial twitching, which was now bilateral, and worsening unsteadiness. Examination showed bilateral facial twitching which had become continuous. Her speech was very jerky. She had a symmetric postural tremor of both hands but strength and sensation were preserved. There were mild bilateral cerebellar signs with finger to nose and heel to shin ataxia, plus gait ataxia. The reflexes remained unremarkable with flexor plantars. Two further EEGs were performed which remained abnormal with frequent runs of bilateral high voltage delta wave activity. Clobazam was introduced instead of clonazepam with improvement of the twitching.

However, her clinical condition continued to deteriorate and in June 1994 she looked thin and unwell. She had oral candidiasis. Continuous bilateral facial twitching persisted. She seemed to have a gaze preference to the right. There was mild global weakness of all four limbs. Reflexes and plantars remained unchanged and there was persistent cerebellar ataxia. Haematological investigations were largely unchanged from previous analyses. With the patient's consent an HIV test was performed and this was negative. The CSF was re-examined and on this occasion the results were all normal with unremarkable cytology. She had further CT of the thorax this time including the upper abdomen, and this was normal. A four vessel cerebral angiogram was normal.

Despite further attempts at treatment with anticonvulsants, broad spectrum antibiotics, and intravenous methyl prednisolone, she entered a rapid decline such that by the end of August 1994 she was effectively confined to bed. The findings on examination had changed. She had become dysphasic. She had bilateral sustained nystagmus, most pronounced on right lateral gaze. Palatal movement was poor. She had developed an asymmetric spastic tetraparesis more pronounced on the right. She had pathologically brisk reflexes with bilateral extensor plantars. She had frequent generalised seizures. She developed a chest infection and despite full supportive measures continued a rapid deteriora-

tion and died on 18 October 1994. A post-mortem examination was performed.

Discussion

DR N A FLETCHER

Nine months before the onset of her neurological illness this woman had developed a right upper lobe lung abscess, from which she recovered. The abscess could have been due to several causes including tuberculosis, although the recovery was very rapid for that condition; other possibilities include a *Klebsiella pneumoniae* and other infectious agents which would question whether her immune status was normal. However, she improved and then developed two episodes of left sided facial twitching, and then another event which was followed by a secondarily generalised seizure. This tells us that there was a discharging cortical focus, because a subcortical abnormality would be unlikely to be followed by a generalised seizure. She was treated appropriately but did not improve. It then became clear that she was developing a progressive neurological illness, because between October 1994 and March 1995, a period of only five months, she developed progressive twitching of the left side of her face, slurred speech, and limb and gait ataxia. Her liver was enlarged. The slurred speech may have been due to cerebellar dysfunction or myoclonus. The best clinical clue to differentiate these is the eye movements, because if myoclonus is severe, the speech may become jerky. Although we are not told about the eye movements here, I suspect that her speech problem was mainly cerebellar, because she had additional limb and gait ataxia. Clearly there was a systemic illness as well.

She deteriorated further, becoming more unsteady, with bilateral facial twitching, cerebellar signs, and a postural tremor. Tremor in patients like this can be confusing because patients with myoclonic disorders may have fine myoclonus of the fingers, mimicking a postural tremor. She was switched to clobazam, but became worse over the next two months, developing oral candidiasis, more pronounced bilateral facial twitching and a gaze preference to the right, global weakness, and cerebellar signs. She was clearly very unwell, so the presence of oral candida does not necessarily point to disturbed immune function. The gaze preference to the right could indicate a brain stem or cerebral lesion, but she seems to have had a multifocal neurological illness.

Between June 1994 and August 1994 she became confined to bed and dysphasic, indicating some left sided cortical or subcortical involvement. Lateral gaze nystagmus and palatal weakness point to involvement of the brainstem, and she had a spastic tetraparesis.

At this point the key question was to decide on the nature of the facial twitching. (Dr Fletcher had available a video of the facial movements.) The twitching involved the whole of the side of her face, was fine, irregular, and worse when she spoke. These movements

could be given several names—for example, focal motor seizures or even *epilepsia partialis continua*. A specialist in movement disorders might call them focal cortical myoclonus, but this is due to a paroxysmal discharge of cortical neurons, and therefore is a fragment of epilepsy. My interpretation of these movements is bilateral (multifocal) cortical myoclonus. This is not a form of subcortical focal myoclonus, which is more commonly regular and rhythmical, such as palatal myoclonus. I would mention oculomasticatory myoarrhythmia, which is a peculiar movement disorder with possible relevance to this case, as the patient had gastrointestinal symptoms; this is pathognomonic of cerebral Whipple's disease,¹ and is a rhythmic oscillation of eye and jaw movements at around 1 Hz, associated with impaired conscious level, vertical eye movements, and sometimes a complete ophthalmoplegia, and the condition reflects a rostral midbrain lesion. This woman's facial appearance did not have the features of that condition.

In summary, the clinical features showed a rapidly progressive multifocal neurological illness over 10 months, with cortical, brainstem, and cerebellar involvement, leading to ataxia, prominent facial twitching, seizures and the later development of corticospinal tract signs. The clinical differential diagnosis must include the Ramsay-Hunt syndrome,² which is a heterogeneous disorder comprising progressive cerebellar ataxia as a prominent feature, myoclonic twitching, and seizures, possibly with the development of other neurological signs later on; it presents as a myoclonic ataxia. At times the seizures and the myoclonus are much more prominent than the cerebellar features, and this is sometimes referred to as progressive myoclonic epilepsy. If we accept that this patient had progressive ataxia with myoclonus and seizures, and accept this as a form of the Ramsay-Hunt syndrome, then among the conditions to consider are spinocerebellar degenerations, such as Unverricht-Lundborg disease (a recessively inherited early onset ataxia sometimes called Baltic or Mediterranean myoclonus),^{3,4} and dentatorubropallidoluysian atrophy,⁵ which is autosomal dominant and may present in adult life with myoclonus and ataxia. A clear family history would be expected with this disorder. Mitochondrial disease should be considered, especially myoclonic epilepsy with ragged red fibres (MERRF),⁶ but the progression seems too rapid for this or a spinocerebellar degeneration. It has recently been emphasised that coeliac disease can cause this syndrome,⁷ and it has been suggested that any patient presenting with the Ramsay-Hunt syndrome—that is, progressive ataxia and myoclonus—should have a jejunal biopsy specifically to look for coeliac disease. Whipple's disease has already been mentioned as a condition that may present in this way, sometimes diagnosed by intestinal biopsy, but sometimes only on post-mortem examination of the brain. There are several rarer conditions to mention. Lafora body disease⁸ would develop at a much earlier

stage than in this case. Kuf's disease (neuronal ceroid lipofuscinosis)⁸ is an adult onset disorder which may be sporadic or familial, with characteristic lipofuscin deposits in the nervous system which may be diagnosed by liver or rectal biopsies. The condition is similar to some of the forms of Batten's disease affecting children. Sialidosis type 1⁹ can be excluded, because patients have a cherry-red spot at the macula, not present in this case. Biotin responsive encephalopathy,⁸ due to biotinidase deficiency, usually occurs in much younger patients, although there are a few reports of adults who develop acquired biotin deficiency as a result of intestinal malabsorption, and get a myoclonic ataxic syndrome. Gaucher's disease⁸ and the rare "action-myoclonus/renal failure" syndrome⁸ are both very unlikely here.

Moving on to the investigations: the CT was normal, she was anaemic, and liver function was deranged, with a biopsy showing fatty change. This last feature tends not to occur in malabsorption syndrome and makes me suspect that she consumed a large quantity of alcohol. The jejunal biopsy showed total villous atrophy, a severe lesion, with crypt hyperplasia, entirely consistent with coeliac disease. The macrocytic anaemia would be consistent with both folate deficiency and alcohol excess. I doubt whether this is vitamin B12 deficiency, because people with coeliac disease tend to absorb B12 relatively well. There were myelocytes and toxic granulation on the blood film, and the chest radiograph was normal with no evidence of any active lung disease. The liver function was non-specifically deranged with raised γ -glutamyl transferase, perhaps due to alcohol, although abnormal liver function tests are occasionally seen in coeliac disease. The CSF protein was raised, which is difficult to interpret, and the EEG was highly abnormal with frequent runs of high amplitude sharp and slow wave activity which looked like seizure discharges, and intervening periods where the background rhythm only looked mildly slow. The repeat jejunal biopsy, after only eight weeks of a gluten free diet showed severe partial villous atrophy, probably not very dissimilar to the initial biopsy. A delay of several months or perhaps a year would be needed to see a real improvement in such a biopsy, even after dietary gluten exclusion in coeliac disease.

Magnetic resonance imaging showed multifocal lesions scattered through the cerebral hemispheres with a predominant subcortical distribution, with many lesions in the brainstem and thalamus. There were lesions in both white and grey matter, so this was not a purely white matter disease, and there was prominent brainstem and diencephalic involvement. There are a few other relevant tests: we know she was HIV negative, CT of chest and abdomen were uninformative and the cerebral angiogram, I imagine looking for the "beading" of vasculitis was unremarkable, although that would not entirely exclude the condition.

There was a curious MRI appearance with multifocal lesions mainly in the brainstem,

cerebellum, and diencephalon, but we know that there is cortical involvement in this case because of the epilepsy and the dysphasia. There was the recent lung abscess, intestinal malabsorption which appears to be due to coeliac disease, the megaloblastic anaemia, fatty infiltration of the liver, raised angiotensin converting enzyme activity which may have numerous causes apart from just sarcoidosis, but this should perhaps be considered here. The MRI might fit with sarcoid, but the clinical presentation would be very unusual, and CSF abnormalities and meningeal involvement would be expected in neurosarcoidosis that was bad enough to have prominent epileptic activity and to be fatal. The neutrophilic changes are non-specific, and would fit with either inflammation or infection, and the raised platelet count can be attributed to hyposplenism which commonly accompanies intestinal malabsorption.

In summary, this is progressive ataxia with myoclonus and epilepsy—that is, a “Ramsay-Hunt” or progressive myoclonic epilepsy type neurological syndrome with unusual facial movements which seem to be myoclonic. The crux of the case hinges on whether the intestinal biopsy findings are considered to be relevant to the case or not. I imagine that they must be relevant and should therefore be taken seriously, especially as there is an association between coeliac disease and this type of neurological illness. We should consider an immunodeficiency state, as this can also cause small bowel atrophy (acquired hypogammaglobulinaemia or variable immunodeficiency may cause recurrent infections and a bowel appearance similar to coeliac disease). Obviously a small bowel lymphoma associated with coeliac disease could have spread to the nervous system, but this would be an unusual presentation. Infections and infestations of the bowel, and tropical sprue could give a similar histological appearance, but it would be difficult to link these to this neurological presentation. Accordingly this neurological illness is probably due to the coeliac disease.

Turning to the reported neurological complications of coeliac disease, there have been reports of a fatal neurological illness associated with coeliac disease, characterised by progressive cerebellar ataxia, myoclonus, epilepsy, peripheral neuropathy, pyramidal tract signs, tremor, and brain stem features,^{10–12} and the key point is that this neurological syndrome did not respond to the withdrawal of dietary gluten. The mucosal lesion in the small bowel did improve, but the neurological features did not. Pathologically there was reported to be extensive gliosis and neuronal loss in a multifocal and patchy way, involving the cerebellum, deep grey matter, and brainstem nuclei, with focal cortical lesions as well. The malabsorption may be very mild and only detectable on biopsy.

Therefore I suspect this neurological illness was secondary to coeliac disease, and that this patient had progressive myoclonic ataxia associated with coeliac disease (Ramsay-Hunt syndrome).

Figure 3 Small bowel biopsy showing subtotal villous atrophy with crypt hyperplasia. An increased number of chronic inflammatory cells are present in the lamina propria, with numerous intraepithelial lymphocytes. The appearance is consistent with a diagnosis of coeliac disease.

DR J W IRONSIDE

The first small bowel biopsy from this patient confirmed an abnormal mucosal pattern with subtotal villous atrophy with crypt hyperplasia, increased inflammatory cells in the lamina propria, and intraepithelial lymphocytes, findings consistent with coeliac disease (fig 3). These features were largely unchanged on the second biopsy.

The necropsy was performed shortly after death, and the most striking findings were in the abdomen with abnormal enlarged rounded lymph nodes scattered within the mesentery. When I saw this I wondered about lymphoma or infection, but sections showed cavitation within the lymph nodes which were full of pale sterile acellular proteinaceous material, an unusual finding called the “mesenteric lymph node cavitation syndrome.”¹³ The lymphatic vessels around the small bowel were also full of this proteinaceous fluid as part of the same syndrome. There was evidence of extensive coeliac disease in the upper small bowel with no sign of tumour, infection, or Whipple’s disease. There was the expected gross splenic atrophy, the organ weighing only 20 g. In the lungs there was florid bronchopneumonia (the immediate cause of death), and scarring from the old abscess, but no sign of tuberculosis, calcification, or tumour.

The brain weighed 1270 g, a little less than expected for a patient of this age. At first glance, there did not seem to be much wrong, either internally or on cross section, but closer examination showed a rather flecked appearance of the white matter, with small areas of discolouration at the junction of the grey and white matter in the cerebral cortex (fig 4).

Figure 4 Coronal section of the cerebral hemispheres at the level of the pulvinar shows small areas of cavitation at the grey/white matter junction, most evident in the region of the left parietal convexity. There are no signs of haemorrhage or evidence of malignancy.

Figure 5 Section from the abnormal region of the left parietal convexity photographed at low power shows the features of a necrotising leucoencephalopathy, with cavitation in the subcortical white matter. No sign of haemorrhage is present (haematoxylin and eosin).

This was more than just perivascular rarefaction, and in places there was cavitation between the junction of the grey and white matter. In the cerebellum there were no external lesions, but a similar discoloured appearance was noted in the white matter on cross section, with a brownish colour around some blood vessels suggesting previous haemorrhage. There were similar findings in the brainstem with discolouration around the blood vessels.

Low power histological view of the cerebellum (fig 5) showed good preservation of the cortical mantle, the main site of damage being the white matter, which was cavitated and had a sponge-like appearance; the adjacent blood vessels were prominent and thick walled. On high power (fig 6) there were multiple areas of cavitation, and the blood vessels showed perivascular haemorrhage in both the grey and white matter. These vessels had cuffs of mononuclear cells, which infiltrated the white matter, and in several areas there was frank vasculitis and surrounding infarction.

Figure 6 Abnormal subcortical white matter contains numerous thickened blood vessels which are infiltrated by lymphocytes, macrophages, and occasional polymorphs. No sign of recent haemorrhage is noted but there is extensive cavitation and gliosis in the adjacent white matter (haematoxylin and eosin $\times 40$).

Immunostains for glial fibrillary acid protein, an astrocytic marker, confirmed gliosis at the grey-white matter interface. Immunostaining for macrophages showed an accumulation of these cells in the white matter around blood vessels and areas of infarction could be seen. No granulomata were identified; the lymphocytes around the blood vessels comprised a mixed population of B and T cells. Stains for amyloid, bacteria, fungi, and parasites were all negative.

The deep grey matter structures, hypothalamus, and cranial nerves were spared, but in the cerebellum there were numerous thick walled blood vessels with active vasculitis and a cavitating leucoencephalopathy. In the spinal cord the vessels within the white matter were the most severely affected by the same pathological process, but there was also evidence of less severe grey matter involvement.

In summary, the necropsy findings were a necrotising leucoencephalopathy with vasculitis, and gliosis involving astrocytes and microglia. The patient did have coeliac disease with splenic atrophy, and the incidental but very unusual finding of mesenteric lymph node cavitation. There was no convincing evidence of alcoholic liver damage.

Conclusions

Dr N A Flecher's diagnosis:

- Ramsay-Hunt syndrome associated with coeliac disease (progressive myoclonic ataxia)

Pathological diagnosis:

- Necrotising leucoencephalopathy with vasculitis (associated with coeliac disease)
- Mesenteric lymph node cavitation
- Coeliac disease
- Acute bronchopneumonia

Comment

The histological features of the small bowel biopsies in this patient, along with the necropsy findings in the jejunum and ileum, confirm the diagnosis of coeliac disease. No evidence of Whipple's disease was noted and there was no evidence of lymphoma. The unusual findings in the mesenteric lymph nodes have previously been reported in coeliac disease.¹³ The pathogenesis of this abnormality is unknown, and unlike other features of coeliac disease, the lymph node changes apparently do not respond to a gluten free diet. The presence of a necrotising leucoencephalopathy associated with vasculitis throughout the CNS has previously been reported in coeliac disease.¹⁰⁻¹² The mechanism of this abnormality is unknown; patients with coeliac disease have been reported to have a wide range of immunological abnormalities, and vasculitis has been described in the skin and in other sites in this disorder. Immune complex deposition has been suggested as the pathogenetic mechanism for the vasculitis, but this remains unproved. The histological features of the CNS vasculitis did not exhibit the features associated with granulomatous angiitis, amyloid angiopathy, sarcoido-

sis, Wegener's granulomatosis, giant cell arteritis, or polyarteritis nodosa. The distribution of the vasculitic lesions in the CNS was more extensive at necropsy than the clinical symptomatology had suggested, although the striking involvement of the brain stem, cerebellum, subcortical white matter, and spinal cord correlates well with the major neurological abnormalities in this patient.

The association of coeliac disease and a progressive neurological disease of this nature is not unique. Presentation of adult coeliac disease as an isolated cerebellar syndrome has been reported,¹⁴ and Kinney *et al*¹⁵ reviewed the cases of 10 patients with coeliac disease who exhibited a progressive CNS degeneration. In a review of the diverse causes of the Ramsay-Hunt syndrome, which was defined as the triad of severe myoclonus, progressive ataxia, and epilepsy with cognitive change, Marsden and Obeso² identified two patients in a series of 10 who had coeliac disease and malabsorption as part of the overall clinical picture.

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Addendum

The case was presented in this form as a clinicopathological conference at the Edinburgh Advanced Neurology Course, 30 March 1995.

The patient had a cerebral biopsy performed before death and was treated intensively with steroids and cyclophosphamide without clinical benefit. These clinical details

were not included in the clinicopathological case conference presentation.

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