negative. Antinuclear antibodies (1:128) revealed a diffuse homogeneous pattern and LE cell preparations were positive. The titre of serum complement (C3) and IgG were normal. Electromyography and nerve conduction studies of the upper/lower limbs were normal. Urodynamic assessment was carried out by cystometry at a filling rate of 50 ml/min. Intravesical pressure during filling did not exceed 15 cmH2O. Bladder capacity was 420 ml. Although the patient had normal sensation of bladder distension, she could not initiate micturition even with the use of the Crédé manoeuvre. A coaxial needle inserted percutaneously into the striated muscle of the anal sphincter showed a continuous hyperactivity, without relaxation during the effort of voluntary micturition, denoting a detrusor-sphincteral dyssynergia. A trial with prednisone 80 mg qd and pyridostigmine 30 mg tid for 2 weeks was unsuccessful. Prednisone was lowered to 20 mg at alternating days. She was maintained with vesical catheterisation and laxatives. After 5 weeks her condition slowly improved and she could void with the use of suprapubic manual compression. We reviewed the patient 4 months later and she was well, dryness of mouth and eyes and intestinal constipation remained unchanged.

We could find only one report of a previous case of acute autonomic neuropathy in association with SLE, in a 21 year-old-female, who showed a rapid clinical response to steroids, which was not observed in our patient. The pathogenesis of this condition is unknown, although both humoral and cell-mediated immune mechanism may be implicated. In some respects this condition resembles experimental autonomic neuropathy. A prospective study of the prevalence of autonomic dysfunction in SLE patients is suggested.

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References


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Korsakoff's psychosis in the presence of multiple sclerosis: an unusual cognitive state

Sirc: The typical Korsakoff’s psychosis, an amnesic syndrome following the acute confusional state of Wernicke’s encephaIopathy, is well known and easy to recognize in chronic alcoholics. This classical picture is however uncommon, and in practice the clinician is often confronted with patients in whom memory deficits of variable severity are accompanied by other cognitive abnormalities and in whom the history of Wernicke’s encephalopathy is often missing. In such cases it is tempting to explain the symptoms as a result of dual pathology caused by thiamine deficiency and by the direct toxic effects of alcohol on the brain. The subacute onset, often seen in such cases, could be interpreted as the result of the overlapping effects of these different lesions, which need to reach a certain threshold to manifest clinically. This explanation, likely to be accurate in most cases, carries the danger of preventing clinicians from looking for other aetiological factors which could be responsible for these atypical combinations of symptoms.

We describe here a patient in whom multiple sclerosis may have contributed to an atypical picture of Korsakoff’s psychosis.

The patient was a 50 year old, twice divorced man who had been admitted for further assessment. He had been referred initially to his local psychiatric hospital by the social services as they had become concerned by his rapid social decline and seeming indifference to it. Prior to his admission the patient had been unemployed and at times living as a vagrant.

Background information was limited by the lack of informants and the patient’s poor memory. His early development was seemingly normal and he had left school with 3 A levels to study engineering at London University. After completing his training he had worked for a number of large engineering companies, but had lost his job on frequent occasions because of drink. His two marriages had failed for the same reason and he had lost contact with his 2 children from his first marriage. He admitted to drinking over half a bottle of spirits a day for several years and was able to recall an admission to a private clinic for detoxification some years previously. His mother and one of his half brothers were said to have abused alcohol.

His previous physical health appeared to have been good except for a transient period of double vision accompanied by an unusual sensation in his feet, “as though walking on cobblestones” which had taken place perhaps two years previously while working in Bahrain. On his return to the UK he remembered being admitted to hospital and receiving ACTH injections with subsequent improvement of his symptoms. The diagnosis of multiple sclerosis was mentioned at the time. He denied further neurological symptoms.

On admission the patient appeared surprised at being in hospital and had few spontaneous complaints. On questioning he admitted to having a poor memory for dates and past personal events. Physical examination revealed a dishevelled, unkempt man who had neglected his personal hygiene. A pale optic disc on the left and an afferent pupillary defect on the same side were the only other signs. There were no stigmas of alcoholic liver disease. At interview he appeared relaxed and cooperative, but rapport was difficult to establish. Answers were rapidly given, inconsistent and often contradictory. He was unconcerned about his problems and denied being depressed. No abnormal beliefs or perceptions were elicited. He was disoriented in time and short term memory was severely impaired, both on verbal and non-verbal testing. He did not confabulate spontaneously, but did so occasionally when provoked by questioning, these answers being inaccurate rather
than “fantastic” in nature. Formal psychological testing revealed a 35 point discrepancy between verbal and performance IQ (vIQ = 131, pIQ = 96). Premorbid IQ (NART) was estimated at 121, thus indicating a decline in general intellectual ability. Recognition memory for words (39/50) and for faces (30/50) was considerably impaired. Performance on an easy recognition memory test for pictures was weak (Camden A 26/30) while on a similarly difficult test performance was at a chance level (Camden B 12/30). There was some evidence of a nominal dysphasia (McKenna 19/30).

Laboratory investigations showed a normal full blood count (with a MCV of 88), red cell transketolase, U + E + creat., LFTs and gamma GTs, B12 and folate, Iron studies, TFTs and VDLR. CSF analysis revealed 17 white blood cells (99% lymphocytes) with a protein of 0.57 mg/l. Oligoclonal bands were present. An EEG showed a low amplitude record with no other abnormal features, and CT of the brain was consistent with a moderate degree of generalised cerebral atrophy. MRI of the brain demonstrated extensive periventricular high signal change on T2 weighted sequences. Multiple high signal foci were seen in the deep white matter of both hemispheres and the overall picture was that of demyelination. Visual evoked responses were abnormal on the left, showing an anterior visual pathway lesion also compatible with demyelination. Sensory evoked potentials were likewise abnormal.

A dual diagnosis of alcohol related cognitive impairment and definite multiple sclerosis was made and a course of IV Parentovite given. The patient was transferred to his local psychiatric hospital with the recommendation that abstinence from alcohol should be enforced as far as possible.

The patient presented with a profound memory deficit together with a more widespread decline in cognitive abilities as evidenced by a substantial drop in IQ and mild nominal dysphasia. In addition, the history of neurological disability and the results of the investigations made it possible to diagnose multiple sclerosis.

The cognitive abnormalities found in this patient are in many ways similar to those described by Cutting in patients with slow onset Korsakoff’s or alcoholic dementia, in whom memory difficulties occur on a background of more widespread cognitive deterioration. Alcohol abuse and associated nutritional deficiencies are considered to be the cause of this picture without invoking the presence of additional aetiological factors. Recently Lishman has put forward a hypothesis that the Wernicke-Korsakoff pathology may also be responsible for a primary alcoholic dementia by its involvement with certain key forebrain nuclei, (the nucleus basalis of Meynert), thus compromising acetylcholine production and producing a clinical picture similar to Alzheimer’s disease.

Our patient, on the other hand, had evidence of dysphasia and a very large verbal-performance discrepancy (WAIS) not often found in Korsakoff’s syndrome. Similar findings emerged in a recent study by Jacobson and Lishman in which only two out of 38 Korsakoff patients had both the severe memory and intellectual decline exhibited by our patient, and whose verbal-performance discrepancies on IQ (namely 31 and 33 respectively) matched those of our patient (Jacobson, Lishman, personal communication). Thus the possibility remains that some of the cognitive deficits present in our patient were the result of demyelination rather than alcohol related pathology. When compared with normal subjects matched for age and education, multiple sclerosis patients have been found to have significantly lower IQ’s. Grant et al in a study of 43 patients with multiple sclerosis, most of whom were in the early or middle phases of the disease, found memory to be impaired in a sizeable proportion of cases, and came to the conclusion that memory loss may be prevalent early on in the natural history of the disease. In both Korsakoff’s syndrome and multiple sclerosis verbal IQs decline more than performance IQs and it could be argued that the combination of multiple sclerosis and Korsakoff’s syndrome in the same patient may have contributed to the large discrepancy between the two IQ subsets, and to the unusual cognitive findings in general. However, even if multiple sclerosis is likely to have been significant, it is unlikely to explain the bulk of the deficits, as it is uncommon for such a profound degree of abnormality to be present in multiple sclerosis patients without severe neurological impairment, although cases of rapidly progressive dementia have been described.

In summary, this case presented with a number of interesting features. Theoretically, alcohol alone could have accounted for the clinical and psychiatric findings. However, the atypical findings and the patient’s patchy recall of a previously suspicious neurological deficit, prompted careful clinical and laboratory investigation that unearthed a second significant pathology. This highlights the importance of looking for other pathologies in patients with alcohol related brain damage (and perhaps in multiple sclerosis also), if the presentation or clinical picture is in any way unusual.

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References

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Cerebral infarction after excessive use of nasal decongestants

Sir: Sympathomimetic agents are frequently used as anorectics, nasal decongestants and cold preparations. Neurological side effects have included seizures,1,2 cerebral haemorrhage and neuropsychiatric symptoms.3 Two cases of cerebral infarction following prolonged use of oral phenylpropanolamine (PPA) and one case after a single dose of PPA have been reported. We report two patients with stroke presumably due to prolonged use of nasal decongestants.

A 35 year old white man presented to the emergency department complaining of sudden speech disturbances and weakness of the right arm and leg. His past history revealed a chronic allergic rhinitis. He had used a total of 15 mg of oxymethazoline every 3 days in nasal decongestants for 20 years. He was taking no other medication, and had no