CASE REPORTS

Autoimmune limbic encephalitis causing fits, rapidly progressive confusion and hyponatraemia

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Abstract

A 78-year-old woman presented with a short history of confusion and seizures and was found to be hyponatraemic. Routine investigations failed to find a cause. Magnetic resonance imaging showed encephalitic changes in the temporal lobes, and voltage-gated potassium-channel antibodies were found in her blood in high titre. Although rare, this condition is important because it may respond to immunosuppression.

Keywords: encephalitis, hyponatraemia, voltage-gated potassium-channel antibodies, elderly

Case report

A previously independent 78-year-old woman was admitted to her local district general hospital with a 2-week history of mild confusion, two occurrences of fits over the preceding few days and an absence-like attack in her doctor’s surgery. She had no relevant past history, was only taking quinine, and had been a smoker. She also complained of dysuria and frequency. On examination, she was apyrexial and intermittently vacant, with an Abbreviated Mental Test Score of 7/10, but had no significant physical signs. Investigations showed a sodium of 124 mmol/l, a heavy growth of Escherichia coli, but no pus cells, in a midstream urine, a normal chest X-ray and short Synacthen test. Cranial Computed Tomography showed superficial atrophy only and EEG slight slowing, without any changes when she was confused and talking to herself. She was treated with fluid restriction and trimethoprim, but remained confused, and was discharged to the care of her daughter after a week.

She was readmitted to a geriatric unit a week later having remained confused despite a serum sodium of 129 mmol/l. Examination was again unremarkable, with an AMTS of 9/10. She had a further fit two nights after admission. Because of the recurrent fits and the possibility of complex partial status epilepticus, she was treated with sodium valproate. A further EEG showed excess slow wave activity with bitemporal high-amplitude delta waves, but no spikes or sharp waves. Other investigations included a normal cerebrospinal fluid glucose, protein, cell count, culture and cytology, negative syphilis serology and urinary porphyrin screens. Computed tomography of chest and abdomen showed no tumours.

She continued to deteriorate, with increasing confusion, hallucinations (sometimes olfactory), falls, retention of urine and consequent infections. Magnetic Resonance Imaging (Figure 1) showed increased signal in deep white matter consistent with cerebrovascular disease and increased signal in the medial temporal lobe on the right, suggesting an encephalitic process. Her valproate was changed to levotiracetam without benefit, and in view of her deterioration, she was transferred to the regional neurological unit.

On transfer, a repeat EEG showed theta and delta waves and frequent spike complexes, at times approaching partial status. She was treated with intravenous phenytoin, with improvement in the EEG. She was thought to have a paraneoplastic or limbic encephalitis. Yo, Hu and Ri antibodies were not found, but assays for voltage-gated potassium channels (VGKC) antibodies were requested. She deteriorated further with sepsis and pulmonary oedema and died. A month after her death, the VGKC antibodies were reported at 1637 pm (normal range 0–100).

At post-mortem, there was severe neuronal loss with multiple reactive astrocytes, macrophages and scattered T-cells in the right amygdaloid nucleus and adjacent hippocampus. Staining for herpes simplex virus was negative.
The diagnosis of encephalitis is based on the clinical picture of fluctuating confusion together with epileptiform changes in the EEG and an abnormal magnetic resonance imaging scan. A paraneoplastic basis to the encephalitis was suspected but no tumour found. The cause of the encephalitis was only established by the presence of antibodies against VGKCs in high titre, reported some weeks after her death. A form of autoimmune encephalitis associated with such antibodies has recently been established [1, 2]. This disorder is typically associated with hyponatraemia and a normal CSF cell count as in this case. Improvement in up to 60% of patients has been reported [2] with plasma exchange, intravenous immunoglobulin either alone or in combination and followed by oral steroids. This rare syndrome is a cause of rapidly progressive cognitive deterioration: early recognition and treatment may be beneficial.

References
