Case report

A treatable form of amnesia and rapid cognitive decline

A. AHMAD1 and M. DORAN2

From the 1Care of the Elderly, Glan Clwyd Hospital, Rhyl, North Wales and 2The Walton Centre for Neurology & Neurosurgery, Fazakerley, Liverpool, UK

Case

A 66-year-old gentleman presented with an episode of generalized tonic clonic seizure. His wife informed that his problems began 7 months earlier when he lost consciousness and began to have dizzy spells. He had several faint like episodes in the next few weeks. He showed behaviour changes and became very anxious and emotionally labile. In the next 2 months, he developed marked problems with his memory immediate, short term and long term. Subsequently, he presented to the accident and emergency department with an episode of generalized tonic clonic seizure. He had further two episodes of generalized tonic clonic seizure in the following 2 months prior to this admission. He also complained of visual hallucinations around this time. There was no significant past medical history apart from hypertension for which he was taking atenolol. He was also on Levitiracetam, which was started after he was diagnosed to be suffering from complex partial seizures during the course of his illness. On examination, he was confused and unable to give any coherent history. There was spontaneous muscle twitching of the calf and thigh muscles bilaterally. Reflexes were present, but generally reduced. Both plantars were down-going. There was no focal wasting or weakness. Sensations, gait and cranial nerves were all normal. Blood tests were unremarkable apart from serum sodium levels of 128 mmol/l. He underwent a magnetic resonance imaging (MRI) scan, which showed a swelling of hippocampi gyri bilaterally with subtle increase in T2 signal (Figure 1). Electroencephalogram revealed occasional theta range slow waves over both frontotemporal regions, at times generalized. There was no epileptic discharge. LP showed a raised protein at 0.85 g/l. There were no oligoclonal bands. PCR for Herpes simplex was negative. The serum and urine osmolarities were consistent with syndrome of inappropriate antidiuretic hormone (SIADH) secretion. The voltage gated potassium channel antibody (VGKC antibody) titre was markedly raised at 621 pmol/l. The following tests were all normal: postcontrast CT of thorax and mediastanum, ultrasound of the testes (to look for occult malignancies), onconeural antibody screen, tumour marker screen, ganglioside antibodies, thyroid antibodies, serum immunoglobulins, anti-neutrophilic antibodies and cardiolipin antibodies.

A diagnosis of autoimmune limbic encephalitis was made on the basis of MRI scan presentation, presence of VGKC antibodies and negative screen for tumours causing paraneoplastic encephalitis.

Plasma exchange (5 l × 5 times a day), IVlg (total dose 2 g/kg), IV methyl prednisolone 1 g/day for 3 days followed by oral prednisolone 1 mg/kg/day. His higher functions improved slowly. By discharge, his behaviour was normal, he had no hallucinations, he had no fits and was self-caring. Pre-treatment addenbrooke’s cognitive exam score was 74/100 and improves to 93/100 after aggressive immunomodulatory therapy was started. Post-plasma exchange serum VGKC antibody titre was

Address correspondence to Dr A. Ahmad, 4 Rhodfa Conway, Dyserth, Rhyl, North Wales LL18 6LS, UK. email: dramirahmad@yahoo.co.uk

*Written consent was obtained for publication of the patient’s details and publication of the image in this report.

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Hyponatremia is also common usually secondary to the SIADH secretion. The presence of voltage-gated potassium channel antibodies confirms the diagnosis. Serum levels of the VGKC antibodies correspond to the degree of neurologic dysfunction. The titres decrease with treatment and clinical improvement. MRI shows bilateral increased signals in the limbic areas (mesial temporal lobe) on T2 images. Unlike paraneoplastic limbic encephalitis, VGKC-associated limbic encephalitis is reversible with prompt administration of immunosuppressant treatment and the prognosis is good. Because of this potentially reversible nature of this condition on timely treatment, it is important to recognize this disease early and hence the need to spread awareness among clinicians.

Conclusion

The learning point from this case is to think of limbic encephalitis in any patient presenting with a combination of subacute cognitive impairment especially loss of memory, behavioural changes and seizures. A blood test (VGKC antibody) and a MRI scan is all that may be needed to confirm the diagnosis and patients can lead normal lives following timely treatment.

References