Case report

One confused patient, many confused physicians: a case of delayed post-hypoxic leucoencephalopathy

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Case Report

A 28-year-old gentleman was referred to our medical department from the psychiatry department, with problems of confusion, disorientation and inappropriate behaviour. He was afebrile and physical examination was unremarkable. He denied hallucinations, delusions or mood disturbance and had no insight of his current problems. Mini-Mental State Examination (MMSE)\(^1\) score was 6/30.

Two years previously he started to use cocaine and heroin episodically. He consumed more than 40 units of alcohol each week.

Five weeks prior to his transfer, he had been admitted to another local hospital after an episode of heavy alcohol and drug consumption. His girlfriend had been unable to wake him up for a number of hours, and on arrival he was treated with naloxone, with a rapid improvement in level of consciousness.

Routine investigations were normal. A urinary toxicology screen was suggestive of recent ingestion of benzodiazepines, opiates and cocaine. A lumbar puncture revealed a normal opening pressure, glucose and protein concentrations and no white cells. CT scan of brain was reported as showing no abnormalities.

At this stage he was treated with intravenous thiamine and acyclovir. Magnetic resonance imaging (MRI) scan of brain (Figure 1) showed periventricular white matter changes with sparing of the basal ganglia and grey matter. Subsequent investigations of autoantibody screen, anti-ENA screen, vasculitis screen, anti-voltage gated K\(^+\) channel antibody, JC polyomavirus serology were all negative. Serum lactate, ammonia, copper, caeruloplasmin, Vitamin B\(_12\), very long-chain fatty acid concentrations, Anti-thyroid peroxidase antibody titre, Arylsulphatase A activity and galactocerebrosidase activity were all within reference ranges. An EEG showed no evidence of asymmetry or of epileptiform waves.

He deteriorated both physically and mentally and required a period of ventilatory and haemodynamic support in the intensive care unit during treatment for a lobar pneumonia. Given his MRI appearances and clinical deterioration a rapidly progressive neuro-degenerative condition was thought probable.

Following return to the ward he began to improve slowly. Eight weeks after admission MMSE\(^1\) score was 19/30. Now at 12 months post insult, he has regained functional independence and is living alone. Discussion with his family members suggests that he has returned to his pre-insult mental state.

Discussion

This case’s clinical course of complete recovery following a hypoxic episode, and deterioration a number of weeks later is characteristic of a diagnosis of delayed post-hypoxic leucoencephalopathy. This condition was first described in 1979,\(^2\) and only five further cases have been described.\(^3\)–\(^7\)
Neurobehavioural problems are most common; but a case with spastic quadraparesis has been reported. Prognosis is good with patients experiencing variable degrees of functional recovery. MRI findings assist in narrowing the differential diagnosis.

Cerebral tissue is susceptible to ischaemic damage via either decreased cerebral perfusion, hypoxaemia or to both processes occurring simultaneously. Damage commonly presents acutely (acute ischaemic hypoxic encephalopathy) with a delayed presentation occurring rarely. The hypoxic insult is usually mild to moderate in delayed presentations, giving rise to a different picture of cerebral tissue damage and a different clinical picture. MRI findings differ, with acute ischaemic hypoxic encephalopathy showing early grey matter changes and in delayed post-hypoxic leucoencephalopathy there are findings of periventricular white matter changes with grey matter sparing.

Grey matter is considered to be more vulnerable to ischaemic damage than white matter. However, in delayed post-hypoxic leucoencephalopathy, the grey matter is classically spared. The deep white matter is perfused by widely spaced linear arterioles with relatively few anastomoses, whilst the grey matter has a better vascular supply. In periods of moderate hypoxia grey matter maintains adequate metabolism due to its better perfusion. It has also been proposed that it is damage to glial cells in the white matter which is responsible for the demyelination observed. This may take a few days to become evident. It is likely that in severe acute hypoxic ischaemic encephalopathy there is damage to both grey and white matter, but that damage to white matter takes some time to become evident radiologically.

**Conclusion**

We present a case of delayed posthypoxic leucoencephalopathy. A delay between hypoxic insult and deterioration can cloud the diagnosis. This rare condition has a classical clinical course and good prognosis. MRI appearances assist in defining the diagnosis.

**Conflict of interest:** None declared.

**References**