

This is the ABN Case Report Winner and we congratulate the authors on this achievement. This case report by Monaghan et al makes for sobering reading and highlights the point that making a diagnosis is one thing, but making sure it is properly treated is another.

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The Woman who Mistook the Past for the Present

A 'variant' presentation of an old disease: a further differential for the 'pulvinar sign'.

The case is presented of a 41-year-old female surgical patient who was transferred to our care with an amnesic syndrome associated with ataxia and ophthalmoplegia. This followed a complicated surgical course associated with abdominal pain, vomiting, adhesions, ileus and parenteral nutrition. Wernicke-Korsakoff syndrome was suspected. Certainty regarding the diagnosis was complicated by lack of information regarding the thiamine content of Total Parenteral Nutrition (TPN) administered to the patient. An alternative diagnosis of Creutzfeldt-Jakob Disease with important quarantine implications had to be considered due to neuro-radiological abnormalities demonstrated on magnetic resonance imaging affecting the medial pulvinar, as well as prominent neuro-psychiatric features common to both disorders. Creutzfeldt-Jakob Disease has previously been misdiagnosed as Wernicke-Korsakoff syndrome. Wernicke-Korsakoff syndrome is an important under-reported

differential diagnosis for the 'pulvinar sign'. Total Parenteral Nutrition is a dangerous misnomer as not all preparations contain total requirements of vitamin supplementation.

Case history

The case is presented of a 41-year-old woman who was well until her wedding day three months earlier. She was transferred to our hospital for investigation and management of grandiose delusions, gaze palsy, nystagmus, ataxia and memory impairment. Subsequently she developed seizures.

She had been investigated for epigastric abdominal pain that developed during her honeymoon in Central America. While abroad she had been vomiting and had reduced oral intake throughout that time. This occurred soon after the onset of bilateral leg oedema following multiple insect bites.

On her return to Ireland, she underwent an OGD

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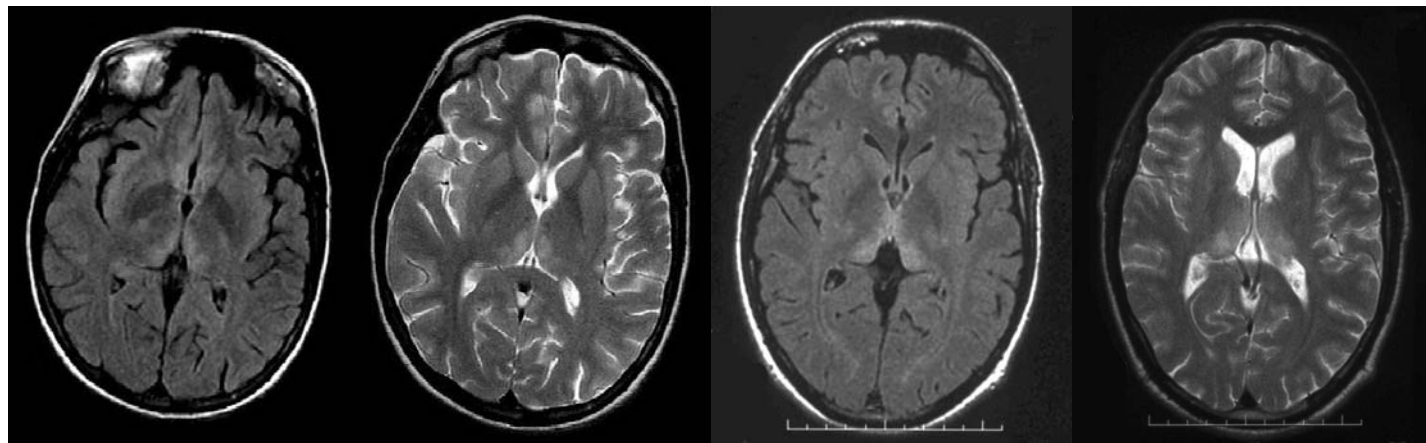


Figure 1a (left) and Figure 1b (right): The inversion recovery image on the left and the T2 image on the right demonstrate the pulvinar sign with increased signal in the medial thalamus.

Figure 2a (left) and Figure 2b (right): Inversion recovery (left) and T2 (right) imaging provided for comparison from a 20-year-old male with variant Creutzfeldt-Jakob disease imaged on the same machine also illustrating the pulvinar sign.

and colonoscopy which were normal.

A laparoscopy was performed because of ongoing vomiting with abdominal pain and multiple abdominal adhesions were surgically lysed at laparotomy. She remained unwell after the procedure with episodes of constipation and vomiting and had poor oral intake. Recovery was delayed over 10 days before the passage of flatus.

The patient was readmitted after recurrence of severe vomiting and constipation. A second laparotomy was performed with resection of 32cms of ischaemic small bowel and further adhesion lysis one month before transfer. Recovery was once again complicated by paralytic ileus and she received TPN for twenty-five days.

Three months into her illness, she then began to demonstrate social withdrawal and became somewhat apathetic. She was referred to a psychiatrist and depression was diagnosed initially. Then confusion and agitation developed. It became apparent that her memory had deteriorated. She was seen by a neurologist.

Clinically she had the following findings: Her affect was flat with features of 'la belle indifference'. She had bilateral ptosis, square wave jerks and ophthalmoplegia to approximately 50% of the normal range. There was nystagmus in the horizontal and vertical planes. There were florid confabulations and she was unable to remember her recent marriage. There was no memory of her new husband as being more than a good friend. She was unable to retain any new information presented. The gait was broad-based and ataxic but there were no signs of peripheral neuropathy.

Investigations

Standard haematological investigations were all normal. Computed tomography of the brain and an electroencephalogram were normal aside from bilateral slow waves ascribed to phenothiazines newly administered for the control of anxiety. A lumbar puncture was performed and all indices were within the normal range.

Magnetic resonance imaging of the brain

revealed high signal intensity lesions in the pulvinar bilaterally (Figure 1a and 1b). This is a well-described sign in variant Creutzfeldt-Jakob disease (vCJD) which has received considerable media and medical attention with the diagnosis of the first two indigenous cases in the Republic of Ireland. Our patient had worked regularly in the United Kingdom during the 1990s and had undergone surgery there.

Management

Clinically Wernicke-Korsakoff syndrome was suspected and therapy with intravenous thiamine was commenced immediately. However on the basis of the imaging and clinical presentation, it was deemed a necessary precaution to quarantine temporarily the surgical instruments used in the four procedures detailed above. This quarantine was removed after re-analysis of the lymphoid tissue of her resected small bowel failed to demonstrate prion protein. An absence of 14-3-3 prion protein in the cerebrospinal fluid, along with clinical improvement in the ophthalmological signs after intravenous thiamine, were supportive of the diagnosis of Wernicke-Korsakoff syndrome. Careful examination of the TPN regimen that was used revealed that it did not contain thiamine.

Discussion

Magnetic resonance imaging with high signal uptake in the pulvinar has been described in vCJD¹. This sign has been demonstrated to have a high sensitivity for vCJD in the appropriate clinical context². It is also reported in sporadic CJD³. Bilateral thalamic infarcts, perinatal ischaemia, iron deposition, copper deposition and neoplastic infiltration have all been associated with the pulvinar sign⁴. These changes are also reported in Fabry disease⁵. More recently this sign has also been described in paraneoplastic limbic encephalitis⁶. Wernicke-Korsakoff syndrome has been reported as part of the clinical differential diagnosis of CJD in one report⁷. Magnetic Resonance pulvinar abnormalities have been described rarely in Wernicke-Korsakoff syndrome^{8,9,10}.

'Total Parenteral Nutrition' regimens which do not contain supplemental vitamins may be more commonly available in pharmacy stocks because these products have a longer shelf life than compounds containing Recommended Daily Amount (RDA) requirements. We propose that TPN regimes which do not contain supplemental vitamins be termed 'Limited Parenteral Nutrition' (LPN) as an alert to avoid the risks of Wernicke-Korsakoff syndrome in nutritionally compromised patients who may receive parenteral nutrition for prolonged periods.

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