Vitamin B₁₂ neuropathy in the absence of anaemia

A case report

J. G. PEROLD

Summary

A case of severe vitamin B₁₂ neuropathy (subacute combined degeneration of the spinal cord) in the absence of anaemia is described and a short review of the literature presented.


Case report

The patient, a 35-year-old White woman, presented with a 1-month history of progressive unsteadiness of gait and a tendency to fall towards the left. She also experienced a numb feeling in both legs, more pronounced on the left side. She had no other systemic symptoms and had had no serious illnesses or operations. She had not used any medication and was born in South Africa.

Although her general physical condition was good, she was extremely unsteady, walking with a broad-based gait. She had severe ataxia of both legs, but the upper extremities were normal. The test for Romberg's sign was positive but no nystagmus was evident. Proprioception in both legs was diminished although all the tendon reflexes were normal. Muscular strength and tone remained normal as did the cranial nerves. No other systemic abnormalities could be found.

The haemoglobin value was 13.2 g/dl with a haematocrit of 38%. Apart from a mean corpuscular volume (MCV) of 96 fl, the rest of the haematological examination was negative. The fasting glucose level was 5.7 μmol/l. A rapid plasma reagin card test was negative and the cerebrospinal fluid was normal. A chest radiograph and a computed brain scan were normal.

Two weeks after the initial consultation she was even more ataxic. At this stage the deep reflexes in both legs were clearly exaggerated and bilateral positive Babinski responses were evident. Periodically, involuntary flexor spasms of her upper legs could be observed.

The combination of posterior and lateral column signs suggested the possibility of vitamin B₁₂ neuropathy. A blood sample was taken in order to ascertain the vitamin B₁₂ and folic acid levels and 1000 μg vitamin B₁₂ was administered intramuscularly. After 7 days she felt much improved and after a further 3000 μg vitamin B₁₂ was administered over the next 8 weeks she was symptom free. On re-examination the posterior column signs were found to have resolved completely, but bilateral positive Babinski signs were still in evidence. The vitamin B₁₂ level was 100 ng/ml (normal 300 - 1000 ng/ml) but the folic acid level was normal. She had achlorhydria both when fasting and after stimulation. A Schilling test confirmed an intrinsic factor deficit (vitamin B₁₂ absorption without intrinsic factor was 7% compared with a normal value in excess of 20%). After the addition of intrinsic factor the absorption increased to 47.5%.

Discussion

The earliest observation of the association existing between neurological disease and anaemia was made by Leichtenstern in 1884. In 1900 the first complete account of subacute combined degeneration of the spinal cord was published by Russell et al. They denied that this disease was intimately associated with pernicious anaemia (PA) and in fact considered that there were 'two totally distinct conditions'. It is known that the two conditions usually coexist, but one may precede the other. Up to 95% of patients with PA had neurological disease a few decades ago but only about 30% experience mild symptoms, usually presenting as paraesthesiae and vague cerebral symptoms. Conversely, vitamin B₁₂ neuropathy is usually associated with PA. In 1929 Ungley and Suzman described 61 cases of vitamin B₁₂ neuropathy, 55 of the patients having PA. The degree of nervous system involvement does not correlate with the degree of anaemia. In exceptional cases, pronounced neuropathy is found in the absence of any haematological abnormality. In one series haemoglobin levels were in excess of 9 g/dl in 30% of patients with spinal involvement.

Conclusions

Although vitamin B₁₂ neuropathy is normally accompanied by pernicious anaemia, gross neurological disease may develop without any haematological or even bone marrow abnormalities. The pointer to diagnosis in the present case was provided by the progression of neurological signs and the slightly raised MCV. The earlier the diagnosis is made, the better the prognosis for the patient. The peripheral nerves may recover completely, but the spinal cord lesion is slow to regress and residual damage may persist.

REFERENCES