Subacute Combined Degeneration of the Spinal Cord and Air Encephalography

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SUMMARY

Subacute combined degeneration of the spinal cord is a rare complication of vitamin $B_12$ deficiency and is seldom encountered today. A case of Addisonian pernicious anaemia is reported in which the classical signs of subacute combined degeneration developed suddenly after air encephalography had been performed. The patient made a complete recovery.


With recent advances in the diagnosis and treatment of vitamin $B_12$ deficiency, subacute combined degeneration of the spinal cord is seldom encountered as a complication of this deficiency.

Intensification of spinal cord signs in patients with pretreated, or previously treated subacute combined degeneration, has been noted during acute intercurrent illness. Such cases have occasionally been termed cases in 'relapse'. This, however, is not the case, and treatment of the intercurrent illness leads to full neurological recovery.

We report a case of Addisonian pernicious anaemia in which the classical signs of subacute combined degeneration of the spinal cord developed suddenly after air encephalography had been carried out.

CASE REPORT

A 47-year-old businessman with a 3-month history of irritability, insomnia, loss of memory and depression was seen by us. He had received psychiatric treatment for depression 10 years previously, but had since remained well. His diet had been good at all times, he was not receiving any medication, and was a teetotaller and non-smoker. All members of his family were well. Physical examination failed to reveal any evidence of organic disease. His blood count was normal at the time of presentation, except for a haemoglobin concentration of 14.0 g/100 ml (normal range at Johannesburg altitude of 2000 metres is 14.5 - 19.2 g/100 ml), a raised mean cell volume of 104 $\mu m^2$ and Westergren erythrocyte sedimentation rate of 30 mm/h.

During the following 2 weeks the patient noted progressive weakness, paraesthesia and cramp-like pains in the lower extremities. Again, clinical examination was normal. In view of the mild anaemia and the raised mean cell volume, serum folate and vitamin $B_6$ assays were performed and the patient was referred for a neurological consultation. At this time he also complained of low backache, and had a fine tremor of the right leg. Neurological examination revealed no other abnormality.

X-ray films of the chest, skull and lumbar spine were normal. Lumbar puncture showed a normal cerebrospinal fluid pressure of 110 mm. An air encephalogram revealed mild cerebral cortical atrophy, with no evidence of a space-occupying lesion. On the following day, the serum vitamin $B_12$ level of 143 pg/ml became known (normal range 400 - 1020 pg/ml using a radio-isotopic dilution technique).

Clinical examination one day later (less than 48 hours after the air encephalogram was performed) revealed the typical features of subacute combined degeneration of the spinal cord. The patient was very depressed and mentation was slow. Examination of the upper limbs revealed only mild hypertonia and hyperreflexia, but the lower limbs showed marked hypertonia with sustained clonus, and marked hyperreflexia. There was pronounced muscle weakness, especially of the quadriceps group. The plantar responses were extensor. Vibration sense was markedly reduced to the level of the iliac crests, and there was significant proprioceptive sensory loss. The neurological deficit was symmetrical. There was marked Rombergism, and the patient developed urinary incontinence. A diagnosis of vitamin $B_12$ deficiency with cerebral cortical atrophy and subacute combined degeneration of the spinal cord was made. The haemoglobin concentration was 12.8 g/100 ml, mean cell volume 106 $\mu m^2$ and mean cell haemoglobin 37.5 pg. The leucocyte count was 3400/$\mu l$ (neutrophils 46%) and the platelet count 325 000/$\mu l$. Serum folate was 28 ng/ml, red cell folate 363 ng/ml and serum lactic dehydrogenase 298 units. Blocking antibodies to intrinsic factor were not detected, but the patient's lymphocytes underwent blastic transformation when exposed to intrinsic factor. Marrow aspiration revealed moderate megaloblastic changes of the erythroid and myeloid series. Since the patient had developed urinary incontinence, a Schilling test was not

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performed. Instead, the absorption of an orally administered dose of 1 µg of 12Co-cyanocobalamin with and without intrinsic factor was measured in a whole-body counter. In the absence of intrinsic factor, 5% of the 12Co-cyanocobalamin was retained after 7 days. When 12Co-cyanocobalamin was given with intrinsic factor, there was 32% retention after 7 days. There was gastric achyia and pentagastrin failed to induce any acid secretion. The diagnosis of pernicious anaemia was thus confirmed.

The patient was treated with hydroxycobalamin and made a dramatic recovery. Within 3 months of the commencement of therapy, during which time he received 6,000 µg hydroxycobalamin, he was mentally normal and there was no residual neurological deficit.

After remission, vitamin B12 turnover was measured in a whole-body counter after intravenous injection of 12Co-cyanocobalamin, and was found to be normal.

**DISCUSSION**

The exact relationship between vitamin B12 deficiency and cerebral cortical atrophy is unclear. Although atrophic changes in the cerebral cortex are well described in patients with vitamin B12 deficiency, cerebral cortical atrophy is not an uncommon finding in the pernicious anaemia age group, and its presence, as revealed by air encephalography, could be coincidental.

The onset and progression of subacute combined degeneration of the spinal cord are generally gradual and evolve over a period of several months. Rarely, and inexplicably, a more acute onset (several weeks) may be encountered. Our patient with pernicious anaemia developed marked spastic ataxia within 48 hours of the procedure of air encephalography, although he had complained of progressive weakness and paraesthesia for 2 weeks previously. It is possible that the classic myelopathy of subacute combined degeneration which developed in our patient might have been precipitated by the procedure of air encephalography. A wide range of side effects has been observed after air encephalography. Those most commonly encountered are headache, vomiting, pyrexia, tachycardia, alterations in blood pressure, neck stiffness, mental confusion and electro-encephalographic changes. On rare occasions, varied non-specific neurological signs may occur temporarily in the first 24 hours after the procedure. We suggest that air encephalography (or even possibly lumbar puncture) could precipitate, or make more evident, a specific neurological syndrome such as that observed in our patient, if there is an underlying organic condition which predisposes to the development of that syndrome, but which otherwise might not manifest, or would take longer to become evident.

Although there is no biological explanation for this phenomenon, the mechanism in our patient would appear to be similar to that which occurs during intercurrent illness. Such cases of 'acute' combined degeneration of the spinal cord have a good prognosis, and complete neurological recovery appears to be the rule, despite severe neurological disturbances during the episode.

**REFERENCES**