sitivity. We present a case of OMM without symptoms of Whipple's disease and with negative peroral jejunal biopsies, which indicates the usefulness of laparotomy for jejunal biopsies as an alternative to brain biopsy to confirm Whipple's disease.

A 67-year-old woman was admitted in May 1988 in a depressed state. In June 1987 she had noted progressive visual disturbance. Rhythmic elevations of her right upper lip appeared, and later, paroxysmal hypersomnolence and an unacceptable weight gain (10 kg). A nine-month course of treatment for depression was ineffective and her symptoms and signs progressively deteriorated. Examination on admission showed complete vertical and horizontal nystagmus without sparing of optokinetic reflexes (which appeared spontaneously, giving the doll's eyes phenomenon with all movements of the head). Her eyes, face, and proximal limbs were of normal appearance, and akinesia, difficulty in speaking, and postural rigidity were noted. She had a generalised hypokinetic dystonia with a facial grimace, which was a constant finding. The possibility of brain involvement without systemic manifestation in Whipple's disease should be kept in mind. This necessitates a search for the disease by endoscopic and peroral jejunal biopsies in all patients with unexplained supranuclear opthalmoplegia. Despite two single reports of reversible CNS involvement, however, the treatment of CNS Whipple's disease has so far been disappointing. The build improvement in neurological states on pefloxacin was important in our patient but needs further confirmation. Pefloxacin is a quinolone which readily diffuses through the blood-brain barrier (BBB) and may eradicate intracellular micro-organisms such as those supposed to be the cause of Whipple's disease. Nevertheless, taking into account the specificity of OMM in the diagnosis of Whipple's disease and the relatively weak results of other antibiotics, we were justified in performing a laparotomy to confirm the disease? We think that this condition needs to be formally recognised and confirmed before embarking on such a long and difficult treatment. Ligs has suggested that incidental use of antibiotics may eradicate gastrointestinal involvement in presumptively Whipple's disease but not the CNS involvement because of the BBB diffusion. This may explain the apparently isolated and late CNS involvement. Such a hypothesis considers only the infectious mechanism and does not explain the poor effect of some antibiotics which are well able to cross the BBB. Unlike most infectious diseases, there are no established cases of direct transmission of Whipple's disease from one patient to another, no re-progression of the disease, laboratory animals, and no convincingly specific organisms isolated by culture in vitro. Whipple's disease is associated with immunodeficiency. Thus intestinal wall macrophages are ineffective in phagocytosing intracellular gram positive bacilli, resulting in inability to eliminate chronic infection. This suggests that Whipple's disease may be considered as a disease with immune deficiency. The periventricular and periaqueductal distribution of the CNS involvement in Whipple's disease consists of macrophagic infiltration and subependymal nodules. The "tumoral" involvement may explain why antibiotics with good BBB diffusion are not effective in this CNS disease.

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Correspondence to: Dr Amarenco, Service de Neurologie, Hôpital Saint-Antoine.


Table Binswanger's clinical and neuropathological criteria for the diagnosis of "Encephalitis subcorticalis chronica progressiva"

<table>
<thead>
<tr>
<th>Clinical criteria: (verbatim from reference 2, page 1184)</th>
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<td>the disease begins at the onset of senility (early in the fifties) or in advanced old age (early in the sixties); slow impairment of intellectual capabilities manifesting primarily by the progressive impairment and ultimately the inability to carry out any mental activity; the patient's mental activity is characterised by a slow deterioration; most frequently observed are aphasic disturbances (as in the present case), hemi-Lamberts or hemiaesthesia, hemiparesis with loss of sense of position, pressure or touch; these circumscribed deficits are of a stable character during the fully developed disease and they are combined with the slow and relentless deterioration of intellectual performance; (… until) the patients resemble deacrebrate laboratory animals.</td>
</tr>
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Neuropathological criteria: (from reference 2, page 137)

Table 1 Binswanger's clinical and neuropathological criteria for the diagnosis of "Encephalitis subcorticalis chronica progressiva"
and the neuropathological findings whichBinswanger considered essential to a disease which he had named "progressiv chronica progressiva". These "reliable criteria" were introduced to differentiate "encephalitis subcorticalis" from "arteriosclerotic brain degeneration" (which also affects the cortex) from the "general paralysis of the insane" and from senile dementia, which he knew could also be accompanied by white matter changes.

The similarities between Bennett's and Binswanger's criteria are obvious. Nevertheless, several striking discrepancies appear noteworthy. The white matter atrophy in Binswanger's patients was most pronounced in the occipital and temporal regions, whereas radiological changes are most commonly found in the frontal lobes. According to Binswanger, "encephalitis subcorticalis" slowly and relentlessly progressed to a state of decerebration, whereas Bennett et al excluded patients with severe dementia. Binswanger assumed that arteriosclerosis was the cause of disease and mentioned the invariable presence of cerebral arteriosclerosis (which was not described in the subcortical region). He did not describe hypertension or other evidence of systemic vascular disease.

It has already been pointed out that the relationship between Binswanger's findings and the modern "Binswanger's disease" remains open to question.

Binswanger did not present a full account of the histopathologic findings. This was left to Alzheimer, who first used the epithet "Binswanger's disease", and to Nissl. Inconsistencies in Binswanger's original description may support the speculation that he eventually regarded the differentiation of such vascular dementia as too difficult or too unwarranted.

HANS FORSTL
ROBERT HOWARD
RAYMOND LEVY

Section of Old Age Institute of Psychiatry, London SE1 8AF, UK


Pseudotumour cerebri and chronic benzene hexachloride (lindane) exposure

Pseudotumour cerebri, the syndrome of idiopathic intracranial hypertension and papilloedema, is characterized by a tumor-like enlargement of the optic nerve sheath with obstructive hydrocephalus, may be associated with exposure to drugs or toxins.1 2 We report a patient, repeatedly exposed to the pesticide benzene hexachloride (lindane), who developed signs of intracranial hypertension.

A 45 year old man (weighing 80 kg) who kept hounds noted fleeting episodes of blurring vision in his right eye usually related to changes in posture. The blurring became persistent after three months and then he developed blurred vision in his left eye. Shortly after he noticed early morning occipital headaches and tinnitus. He had used benzene hexachloride at least twice a month for about 30 years to rid his beagle hounds of fleas and ticks. He had used a 20% lindane concentrate to make dip and spray applications but wore a mask and appropriate protective clothing. He was well built but not obese. His neurological examination yielded normal resposable factor findings: his visual field was best corrected visual acuity was 6/36 OD and 6/9 OS. He had a right relative afferent pupillary defect. Ocular motility and slit lamp examinations were normal. Intracranial pressures were 28 and 23 mmHg respectively. Ophthalmoscopic examination showed distinct swollen optic discs with small cups, loss of the nerve fibre layer in the right eye, and a small pseudodrusen in the left eye, typical of chronic papilloedema. Goldmann perimetry showed visual field loss characteristic of chronic papilloedema.

MRI of the head was normal except for a few small areas of white matter lesions; venous sinus thrombosis was not seen. A spinal tap showed an opening pressure of 400 mm CSF with one monocyte per cu mm, protein 0.34 Gm/l, glucose 2 mmol/l, and no cells or infection. Other biochemical laboratory values were not only for elevated cholesterol and triglyceride concentrations and mildly abnormal results of liver function tests. Thyroid function tests were normal, and antinuclear antibodies were negative. Toxins screens for lead, mercury, and arsenic were negative. Management included dietary advice (weight loss), diuretics, and prednisone, but he subsequently had 21 visual field changes and nasal congestion and tinnitus because of progressive visual field loss. Ten months after diagnosis his field defects were stable, but his visual acuity remained impaired.

Lindane, a gamma isomer of hexachlorocyclohexane used as a pesticide and an ectoparasiticide, is metabolised by the liver and distributed and stored in depot fat and other lipophilic tissues.1 2 Lindane is commonly prescribed topically as a 1% solution for scabies but is available in concentrations of 0.5%-99%; our patient used a 20% veterinary concentration for his dogs. Lindane is a powerful CNS depressant, known to cause headaches, nausea, vomiting, diarrhea, convulsions, muscle spasms, respiratory failure with cyanosis, coma, and death.3 4 Optic neuritis after "improper use" of lindane powder has also been reported.5 Heuser and Heuser briefly described "pseudotumour cerebri" in a farmer with localised brain oedema, after "prolonged professional inhalation" of lindane; but the appearance of the optic discs and differential pressure were not reported. The mechanism of lindane toxicity is unknown, although it is highly lipophilic and may act as a gamma-amino butyric acid (GABA)-A receptor antagonist to produce convulsive effects and interference with the production and utilisation of free ammonia in the brain.5 6 Chlordecone, a cyclodiene insecticide which also induces seizures, has been implicated in causing pseudotumour cerebri by inhibition of ATPase activity, resulting in impaired resorption of cerebrospinal fluid across the arachnoid villi.7 Lindane may have similar effects on the arachnoid villi as they are both lipid soluble, neurotoxic chlorinated hydrocarbons.

Our patient stopped using lindane when the association of pseudotumour cerebri and lindane was brought to his attention; this was coincidentally reinforced when a neighbour's puppies convulsed and died after exposure to a 20% solution. Despite discontinuation of the pesticide the patient's intracranial pressure remained elevated and his headaches continued 11 months later when a lumbo-peritoneal shunt was inserted. Removal of the toxin should result in alleviation of increased intracranial pressure. Perhaps the lindane caused permanent or prolonged alteration of the arachnoid villi. Alternatively, lindane may be present in fat cells for an extended period and have a long lasting effect on CSF absorption. Whether the patient's liver damage was caused by previous chronic alcohol consumption or exposure to lindane is unclear. The relation with lindane exposure may not be coincidental because other pesticids have been linked to pseudotumour cerebri in the past.2 The use of lindane should be discontinued when patients have unexplained raised intracranial pressure.

LISA VERDERBER
PATRICK LAVIN
RALPH WESLEY

Departments of Neurology and Ophthalmology, Vanderbilt University Medical Center, Nashville, TN 37212, USA.

Correspondence to: Dr Lavin, M.D., 2101 Pierce Ave., Nashville, TN 37212, USA.


9 St Omer V. Investigation into mechanisms responsible for seizures induced by chlorinated hydrocarbon pesticides. J Neurol 1971;18:365-74.

Motor neuron syndrome in the arms after radiation treatment

Radiation myelopathy is a rare but well established complication of radiotherapy leading to diagnostic difficulties with neurological complications of the primary neoplasm, like epidermitis or spinal metastasis. We report a rare case of radiation motor neuron syndrome that developed three years after local radiotherapy in which spinal cord magnetic resonance imaging (MRI) showed a cervical spinal syrinx and lesion.

A 44 year old man without relevant history presented with dysphonia and a rapidly growing cervical anterior mass. We found a mal-
Binswanger's clinical and neuropathological criteria for "Binswanger's disease".

H Förstl, R Howard and R Levy

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