negative biopsies month course of progression and reflexes and Rhythmic elevations by masticatory was of adduction or proximal jejunal Erythrocyte range (12-30%), teric examination showed ment found, confirming and were but the the the of the minimental status from bed. nuclear oculomotor paresis, confirming and unexplained supranuclear oculomotor palsy. Despite two definite reports of reversible CNS involvement, however, the treatment of CNS Whipple’s disease has so far been disappointing. The mild improvement in neurological states on pefoxacin was important in our patient but needs further confirmation. Pefoxacin is a quinolone which readily diffuses across the blood–brain barrier (BBB) and is effective against intracellular micro-organisms such as those supposed to be the cause of Whipple’s disease. Nevertheless, taking into account the specificity of OMA in the diagnosis of Whipple’s disease and the possibility that “abnormality” were not justified in performing a laparotomy to confirm the diagnosis? We think that this condition needs to be formally recognised and confirmed before embarking on such a long and difficult treatment. Riggs has suggested that incidental use of antibiotics may eradicate gastrointestinal involvement in presymptomatic Whipple’s disease but the risk of relapse because of partial 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, nor reproduction of the disease in laboratory animals, and no convincing 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 of macrophages. The periventricular and periaqueductal distribution of the CNS involvement in Whipple’s disease is consistent with macrophage infiltration and subsequent inflammatory “tumoral” involvement may explain why antibiotics with good BBB diffusion are not effective in this CNS disease.

Binswanger's clinical and neuropathological criteria for “Binswanger’s disease”

Bennett et al1 have made a timely and critical attempt to standardise the diagnostic criteria for “Binswanger’s disease”. They have based their suggestions on current English and French literature. It might be of interest to compare these modern criteria to Binswanger’s original description, which had only been available in English in a grossly truncated form.3

<table>
<thead>
<tr>
<th>Table Binswanger’s clinical and neuropathological criteria for the diagnosis of “Encephalitis subcorticalis chronica progressiva”</th>
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</thead>
<tbody>
<tr>
<td>Clinical criteria: (verbalism from reference 2, page 1184)</td>
</tr>
<tr>
<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 ultimate destruction of cortico–subcortical areas; most frequently observed are aphasic disturbances (as in the present case), hemiamblyopia or hemianopia, hemiparesis with loss of the sense of position, 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 decerebrate laboratory animals.</td>
</tr>
<tr>
<td>Neuropathological criteria: (from reference 2, page 1137)</td>
</tr>
<tr>
<td>we find a pronounced atrophy of the hemispheric white matter, either restricted to one or more gyri in one brain area or of several hemispheric regions affected with variable severity; these changes are most clearly found in the area of the occipital and temporal lobes, so that the temporal and occipital horn are widened into bag-like cavities, while the anterior portion of the lateral ventricle shows relatively little enlargement and the frontal white matter is relatively preserved. The cortex does not show any remarkable macroscopic change apart from a slight narrowing. Invariably, these cases show severe atrophy of the cerebral arteries.</td>
</tr>
</tbody>
</table>

![Image](http://jnnp.bmj.com/)

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and the neuropathological findings which Binswanger considered essential to a disease which he called "atro-vascular dementia" or "chronica progressiva". These "reliable criteria" were introduced to differentiate "encephalitis subcorticalis" from "arteriosclerotic brain degeneration" (which also affects the white matter) and the "general paralysis of the insane" and from senile dementia, which he knew could also be accompanied by white matter changes.1

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 lobes, whereas radiological changes are most commonly found in the frontal lobes. According to Binswanger, "encephalitis subcorticalis" slowly and relentlessly progressed to a state of deprecation, 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 first described in the subcorticalis) extensively. 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 histopathological changes in his case. This was left to Alzheimer, who first used the term "Binswanger's disease",2 and to Nissl.3 Inconsistencies in Binswanger's original description may support the speculation that he eventually regarded the differentiation of such vascular dementias as too difficult or too unrewarding.4

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Pseudotumour cerebri and chronic benzene hexachloride (lindane) exposure

Pseudotumour cerebri, the syndrome of idiopathic intracranial hypertension and papilloedema, may be present as a tumour, an obstructive hydrocephalus, or 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 an idiopathic intracranial hypertension. A 45 year old man (weighing 80 kg) who kept hounds noted fleeting episodes of blurred vision in his right eye usually related to changes in posture. The blurring became persistent after three months and then he developed headaches 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 also used a 20% solution to 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 results except for visual findings: his 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. Intraocular pressures were 21 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 white matter lesions; venous sinus thrombosis was not seen. A spinal tap showed an opening pressure of 450 mm CSF with one monocyte per cu mm, protein 0.34 Gm/l, glucose 2 mmol/l, and no infection. Other laboratory values were not only for elevated cholesterol and triglyceride concentrations and mildly abnormal results of liver function tests. Thyroid function tests were normal. Benzene, lead, mercury, and other environmental concentrations were negative. Toxic screens for lead, mercury, and arsenic were negative. Management included dietary advice (weight loss), diuretics, and prednisone, but he subsequently had Right nerve sheath meningitis 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 hexachloro- cyclohexane used as a pesticide and an ectoparasiticide, is metabolised by the liver and distributed and stored in depot fat and other lipophilic tissues. Lindane is commonly prescribed topically as a 1% solution for scabies but is available in concentrations of 0.5%, 5%-, 99%; our patient used a 20% veterinary concentration for his dogs. Lindane is a powerful CNS suppressant commonly known to cause headache, nausea, vomiting, diaphoresis, 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 visual field pressure were not reported. The mechanism of lindane toxicity is unknown, although it is highly lipophilic solvent and may act as a gamma-amino- butyric acid (GABA-A) receptor antagonist to produce convulsions and interference with the production and utilisation of free ammonia in the brain.6 Chlorozone, a cyclohexene 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 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 lin dane 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 prolonged chronic alcohol consumption or exposure to lindane is unclear. The relation with lindane exposure may not be coincidental because other pesticides have been linked to pseudotumour cerebri in the past.2 The use of lindane should be discontinued when patients have unexplained raised intracranial pressure.

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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 epidermis or spinal metastasis. We report a rare case of radiation myelopathy presenting as a cervical motor neuron syndrome which developed three years after local radiotherapy in which spinal cord magnetic resonance imaging (MRI) showed a cervical spinal cord lesion. A 44 year old man without relevant history presented with dyspnoea 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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