sivity. We present a case of OM without symptoms of Whipple’s disease and with negative peroral jejunoileal biopsies, which indicates the uselessness of laparotomy for jejunoileal 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 hypersomnia and very weak 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 ophthalmoplegia with sparing of oculoocephalic reflexes (which appeared spontaneously, giving the doll’s eyes phenomenon with all movements of the head). Her eyes, face, and proximal areas were all spared. Pathological examination showed no neuronal abnormality but PAS positive cells with positive Gram stain in the mesenteric lymph nodes were found, confirming Whipple’s disease. Treatment with trimethoprim-sulfamethoxazole and tetracycline for two months stabilised the disease. Trimethoprim-sulfamethoxazole and chloramphenicol were then given for a further two months. The results of Schilling’s test returned to normal, but signs and symptoms did not improve. Chloramphenicol had to be withdrawn and was replaced by pefloxacin. Two and a half years after the onset of treatment there was mild improvement of signs. Masticatory movements remained the same, but the adduction of the eyes had disappeared; the eyes could be displaced to the left and right to 50% and slightly downward, with oculogyric crises. Unsteadiness of gait, probably due to the supranuclear oculomotor palsy, persisted. Her mental status was largely unaffected (miniminal status was 30/30). Her full scale intellectual quotient on the WAIS-E-II slightly declined from 92 to 86 then increased to 96 in 1990. An uncontrolled new synchronous movement of flexion extension of the right thigh appeared when she relaxed in bed. Worsening of hypersomnia and appearance of cataplexy required high doses of imipramine which gave partial relief. Progressive supranuclear palsy is a feature of Steele Richardson Olszewski disease, a description of a disorder of unknown aetiopathology and with no known treatment. A supranuclear oculomotor paresis for downgaze is one of the main characteristics of this syndrome, together with axial rigidity and pseudobulbar palsy. The association of supranuclear gait paresis with oculomotor myopathy, however, seems to be pathognomonic of the effects of Whipple’s disease on the CNS. This led us to perform surgical jejunal and mesenteric lymph node biopsies rather than a brain biopsy despite negative endoscopic and numerous peroral distal jejunal and ileal biopsies. The normality of the CT and MRI scans also supported this decision. The possibility of brain involvement without overt systemic manifestation in Whipple’s disease should be kept in mind. This necessitates a search for the disease by endoscopic and peroral jejunoileal biopsies in all patients with unexplained supranuclear oculomotor palsy. Despite two single reports of reversible CNS involvement, treatment of CNS Whipple’s disease has so far been disappointing. The morphological changes in the CNS in patients with Whipple’s disease are consistent with a neurodegenerative disease with gliosis and mononuclear and multinuclear macrophages. Brain biopsy showed granulomas in 4 of 18 patients with symptoms of CNS involvement. The histological changes of biopsies taken from 9 patients showed a mixed inflammatory infiltrate of mononuclear and polymorphonuclear leucocytes with microorganisms such as those supposed to be the cause of Whipple’s disease. Nevertheless, taking into account the specificity of OM in the diagnosis of Whipple’s disease and the morphological changes, the diagnostic criteria were 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 protocol. Riggs has suggested that incidental use of antibiotics may eradicate gastrointestinal involvement in presymptomatic Whipple’s disease but not the more proximal involvement because of 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 reproductibility of the disease in 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 intracerebral gran positive bacilli, resulting in inability to eliminate chronic infection. This suggests that Whipple’s disease may be considered as a form of macrophagocytosis. The periventricular and periarterial distribution of the CNS involvement in Whipple’s disease consists of macrophagic infiltration and atrophy of subependymal zones. The brain formation in Whipple’s disease shows no tumoural” involvement which may explain antibodies with good BBB diffusion are not effective in this CNS disease.

BINSWANGER'S CLINICAL AND NEUROPATHOLOGICAL CRITERIA FOR "BINSWANGER'S DISEASE" 

Bennett et al. 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. The table shows the clinical hallmarks

**Table Binswanger’s clinical and neurological criteria for the diagnosis of “Encephalitis subcorticalis chronica progressiva”**

<table>
<thead>
<tr>
<th>Criteria</th>
<th>1184</th>
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<tbody>
<tr>
<td>Clinical criteria: (from reference 2, page 1184)</td>
<td></td>
</tr>
<tr>
<td>is the disease begins at the onset of senility (early in the fifties) or</td>
<td></td>
</tr>
<tr>
<td>as usual, in a person of advanced age (early in the sixties)</td>
<td></td>
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<tr>
<td>slow impairment of intellectual capabilities manifesting primarily by</td>
<td></td>
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<tr>
<td>the progressive impairment of voluntary movements of the upper extremities</td>
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<tr>
<td>most frequently observed are aphasic disturbances (as in the present case,</td>
<td></td>
</tr>
<tr>
<td>hyperrhythmia or hemianopia, hemiparesis with loss of the sense of pressure,</td>
<td></td>
</tr>
<tr>
<td>these circumscribed deficits of are of a stable character during the</td>
<td></td>
</tr>
<tr>
<td>and are combined with the slow and relentless deterioration of intellectual</td>
<td></td>
</tr>
<tr>
<td>(… until the patients resemble decrebrate laboratory animals.</td>
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</tbody>
</table>

**Neurophysiological criteria: (from reference 2, page 1137)**

We find a pronounced atrophy of the hippocampal formation on one or both sides, especially on the right. This is associated with an abnormal activity and a decrease in the number of neurons and glia. These circumscribed deficits of are of a stable character during the fully developed disease and they are combined with the slow and relentless deterioration of intellectual performance;
and the neuropathological findings whichBinswanger considered essential to a disease which he called "chronica progressiva". These "reliable criteria" were introduced to differentiate "encephalitis subcorticalis" from "arteriosclerotic brain degeneration" (which also affects the cortex) from "Binswanger's original 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 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 decerebration, whereas patients with Binswanger's disease did not present a full account of the histopathological picture. This was left to Alzheimer, who first used the term "Binswanger's disease", and to Niissl. Inconsistencies in Binswanger's original description may support the speculation that he eventually regarded the differentiation of such vascular demetias as too difficult or too unrewarding.

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RAYMOND LEVY
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3 Olzewski I. Subcortical arteriosclerotic encephalopathy, a clinical description of a new concept, or called Binswanger's disease and presentation of 2 cases. World Neurol 1962;3:59-75.

Pseudotumour cerebri and chronic benzeno hexachloride (lindane) exposure

Pseudotumour cerebri, the syndrome of idiopathic intracranial hypertension and papilloedema, is usually a consequence of a tumour, obstructive hydrocephalus, or may be associated with exposure to drugs or toxins.1,4 We report a patient, repeatedly exposed to the pesticide benzene hexachloride (lindane), who developed intracranial hypertension.

A 45 year old man (weighting 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 intermittent blurring in his left eye. Shortly after he noticed early morning ocipital 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 then used a 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 and findings showed the best corrected visual acuity was 6/36 OD and 6/9 OS. He had a right relative afferent pupillary defect. Ocular mobility 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, mildly indurated 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 evidence of infection. Of the laboratory values were notable only for elevated cholesterol and triglyceride concentrations and mildly abnormal results of liver function tests. Thyroid function tests were normal; rheumatoid factor and antinuclear antibodies were negative. Toxic screens for lead, mercury, and arsenic were negative. Management included dietary advice (weight loss), diuretics, and prednisone, but he subse- quently had both optic nerve dysfunction and cerebral oedema; further investigation showed a state 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 hexachlorocy-clohexane used as a pesticide and an ectoparasiticide, is metabolised by the liver and distributed and stored in depot fat and skeletal muscle. A lipophilic toxicant, it 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 GABA-A agonist known to cause optic atrophy, headache, nausea, vomiting, diarrhoea, convulsions, muscle spasms, respiratory failure with cyanosis, coma, and death.4,5 Optic neuritis after "improper use" of lindane powder has also been reported.6 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 papilloedema were not reported. The mechanism of lindane toxicity is unknown, although it is highly lipophilic and may act as a gamma-aminobutyric acid (GABA)-A receptor agonist to produce convulsive effects and inter- ference with the production and utilisation of free ammonia in the brain.7 Chlordecone, a cytochrome inactivator which also induces apoptosis, has been implicated in causing pseudotumour cerebri by inhibition of ATPase activity, resulting in impaired resorption of cerebrospinal fluid across the arachnoid villi.8 Lindane has also been shown to cause 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 cerebi 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 pres- sure remained elevated (44 mm Csf) and headaches continued 11 months later when a lumbo-peritoneal shunt was inserted. Removal of the toxin should result in allevia- tion of increased intracranial pressure. Per- haps the lindane caused permanent or long- inged alteration of the arachnoid villi. Alter- natively, 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 pesticides have been linked to pseudotumour cerebri in the past.7 The use of lindane should be discontinued when patients have uneexplained raised intracranial pressure.

9 St Omer V. Investigation into mechanisms res- ponsible for seizures induced by chlorinated hydrocarbon pesticides. J Neurol 1971;183:365-74.

Motor syndrome in the arms after radiation treatment

Radiation myelopathy is a rare but well described complication of radiotherapy leading to diagnostic difficulties with neurological complications of the primary neoplasm, like epiduritis or spinal metas- tasis. We report a rare case of radiation myelopathy resembling a cervical motor neuron syndrome that 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-