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

A 77 year old woman was admitted in May 1988 in a depressed state. In June 1987 she had noted progressive visual disturbance. Rhythmic elevations of adduction of type pseudobulbar in sign as Erythrocyte sedimentation rate (35%) and normal CSF. Her eyes, face, and proximal movements were affected by myopathy, which consisted of masticatory movements with synchronous adduction of her eyes (convergence) and limbs at about 1 cm. Her soft palate was not involved but also occurred during sleep. Gait disturbance and a slight facial akinesia was noted but no rigidity. Her mental state was normal. There was a pseudobulbar type dysphagia but no dysarthria. No other systemic sign or symptom was recorded; there was no diarrhea. Her CSF was normal with no PAS positive cells present. EEG, CT, and T1-weighted (DTPA-Gadolium enhanced) and T2-weighted MRI both showed no focal lesions. Also, MRIs yielded normal results, as did endoscopy and 30 peroral distal and proximal jejunal and ileal biopsies. Erythrocyte sedimentation rate was 35 mm per hour. Schilling’s test was 5% (normal range is 5%), suggesting distal ileal involvement.

A laparotomy was performed and surgical biopsy specimens of the jejunum and mesenteric lymph nodes were taken. Pathological examination showed no jejunal abnormality but PAS positive cells with positive Gram stain in the mesenteric lymph nodes were found, confirming Whipple’s disease. Treatment with 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 Chloramphenicol had to be withdrawn and was replaced by pefloxacin.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 symptoms. 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. Unsteadiness of gait, probably due to the supranuclear oculomotor palsy, persisted. Her mental state was largely unaffected (miniminal status was 30/30). Her full scale intellectual quotient on the WAIS was 92 to 86, an increase to 96 in 1990. An uncontrolled new synchronous movement of flexion extension of the right thigh appeared when she relaxed in the supine position of hyperextension 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 degeneration of unknown aetiology and with no known treatment. A supranuclear oculomotor paresis for downward gaze is one of the main characteristics of this syndrome, together with axial rigidity and pseudobulbar palsy. The association of supranuclear gaze 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 under 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 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, 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 across the blood–brain barrier (BBB) and is active against 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 role of antibiotics in the treatment 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 reproduction 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 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 consists of macrophagic infiltration and subependymal nodules. Such a “tumoral” involvement may explain why antibiotics with good BBB diffusion are not effective in this CNS disease.
and the neuropathological findings which Binswanger considered essential to a disease with "latent hypertensive arteriosclerosis or senile arteriosclerosis", although he noted the subacute or chronic progression. These "reliable criteria" were introduced to differentiate "encephalitis subcorticalis" from "arterio-sclerotic brain degeneration" (which also affects the cortex) and "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 de cerebration, 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, however, not always pathologically described extensively). He did not describe hypertension or other evidence of systemic vascular disease.

It has already been pointed out that the relationships between Binswanger's findings and the modern "Binswanger's disease" remains open to question. Binswanger did not present a full account of the histopathological his findings. This left to Alzheimer, who first used the term "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 dementias as too difficult or too unrewarding.

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1 Bennett DA, Wilson RS, Gilley DW, Fox JH. Clinical diagnosis of Binswanger's disease. J Neurol Neurosurg Psychiatry 1990;53:


4 Alzheimer A. Die Seelenstörungen auf arteriosklerotischer Grundlage. Allg Z Psych 1902;54:695-701. (Transl. Mental dis-


Pseudotumour cerebri and chronic senile benzene hexachloride (lindane) exposure

Pseudotumour cerebri, the syndrome of idiopathic intracranial hypertension and papilloedema, involves a tumefaction of the meninges, obstruction of hydrocephalus, may be associated with exposure to drugs or toxins. 4, 5 We report a patient, repeatedly exposed to the pesticide benzene hexachloride (lindane), who developed 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 developed during sustained periods in the 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 his eye drops 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; findings included a right sided pseudo-onion papilla, right temporal bone robotic movement, and a balloon swelling of the right side, atypical of pseudopapilli in the left eye, typical of chronic pseudotumour cerebri. Goldmann perimetery showed visual field loss characteristic of chronic papillodema.

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 400 mm CSF with one monocyte per cu mm, protein 0.34 Gm/l, glucose 2 mmol/l, and no abnormal spinal fluid findings. Our laboratory values were notable only for elevated cholesterol and triglyceride concentrations and mildly abnormal results of liver function tests. Thyroid function tests were normal; routine tests for antinuclear and antiphospholipid antibodies were negative. Toxic screens for lead, mercury, and arsenic were negative. Management included dietary advice (weight loss), diuretics, and prednisone, but he subsequently had both optic nerve sheath oedema and persistent ocular hypertension. Goldmann perimetry showed visual field loss characteristic of chronic papillodema.

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 pres- sure changes were progressive. MRI showed that 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 pseudotumour cerebri in the past 2. The use of lindane should be discontinued when patients have unexplained raised intracranial pressure.

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Motor 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, such as ependymitis or spinal metastasis. We report a rare case of radiation myelopathy presenting as a cervical motor neuron syndrome that developed three years after local radiotherapy in which spinal cord magnetic resonance imaging (MRI) showed a cervico-thoracic cervical cord lesion.

A 44 year old man without relevant history presented with dysphonia and a rapidly growing cervical anterior mass. We found a mal-