and the neuropathological findings which Binswanger considered essential to a disease which he called “chronica progressiva”. These “reliable criteria” were introduced to differentiate “encephalitis subcorticallis” from “arteriosclerotic brain degeneration” (which also affects the cortex) and 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 subcortical and temporal lobes, whereas radiological changes are most commonly found in the frontal lobes. According to Binswanger, “encephalitis subcorticallis” slowly and relentlessly progressed to a state of deacrebration, 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 he later called “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 unwarranted.

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Pseudotumour cerebri and chronic benzene hexachloride (lindane) exposure

Pseudotumour cerebri, the syndrome of idiopathic intracranial hypertension and papilloedema, is a result of a tumour or 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 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 intermittent blurring 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 a normal creatinine clearance 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. MRI findings were the 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 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 400 mm CSF with one monocyte per cu mm, protein 0.34 Gm/l, glucose 2 mmol/l, and no other abnormality. Other 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 subsequently had bid nocturnal, severe, fleeting episodes of retrobulbar ophthalmoplegia 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. Lindane is said to be prodrived 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 stimulant known to cause headache, nausea, vomiting, diarrhoea, 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 Heuer and Heuer 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 was not reported. The mechanism of lindane toxicity is unknown, although it is highly lipid soluble 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.6 Chlorodecone, a cyclodienes insecticide which also induces myelination abnormalities in castrated 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; 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 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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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, such as epidermis or spinal metastasis. We report a rare case of radiation induced motor neuron syndrome that developed three years after local radiotherapy in which spinal cord magnetic resonance imaging (MRI) showed a cervicomedullary lesion.

A 44 year old man without relevant history presented with dysphonia and a rapidly growing cervical anterior mass. We found a mal-
pigmented carcinoma invading the thyroid and the right wall of the trachea. He was treated with ablation of the right thyroid lobe, tracheotomy, and three courses of chemotherapy (CCDFP, 5FU) followed by radiation therapy (70 Gy) and upper mediastinum (45 Gy) with a 20 Gy dose to the sub-glottic area. The calculated total radiation dose received on the spinal cord was 48 Gy at the C6 level (maximal dose at 2 cm high), 44 Gy at the spinal cord received at 42 Gy at the cervical region (including C7) and 41 Gy for T1. There was no development of the mass on regular CT scans. The only treatment was thyroid hormones after surgery.

Three years after finishing radiation therapy and after transient parasthesia of the hands and a Lhermitte sign (lasting less than two weeks), the patient developed progressive wasting and weakness of both hands and forearm extensors. He had no symptoms in the legs nor sphincter disturbance. We found no sensory abnormality (except a questionable reduction of tactile perception on the involved hand) nor motor dysfunction, and tendon reflexes were present except supinator and triceps jerks on both sides. Neurological examination otherwise was normal.

Electromyographic examination showed fibrillation, giant potentials and some fasciculations in right and left hand muscles, and reduced recruitment in forearm extensors. There was no palsy of the arm or weakness of the face and legs were normal. Motor conduction velocities, distal motor latencies, and amplitude of sensory potentials were normal in all limbs. Values of somatosensory evoked potentials were in the normal range after median and posterior tibial stimulation. Haematological, biochemical, and hepatic routine tests, thyroid hormones, lipopigment, syphilitic serology, viral serology, serum and urine lead concentrations were normal. Cerebrospinal fluid was normal for cellular count, biochemistry and protein electrophoresis.

Cervical MRI was performed five months after the radiation, in the site of the arm and muscles of the face and legs were normal. Motor conduction velocities, distal motor latencies, and amplitude of sensory potentials were normal in all limbs. Values of somatosensory evoked potentials were in the normal range after median and posterior tibial stimulation. Haematological, biochemical, and hepatic routine tests, thyroid hormones, lipopigment, syphilitic serology, viral serology, serum and urine lead concentrations were normal. Cerebrospinal fluid was normal for cellular count, biochemistry and protein electrophoresis.

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