and the neuropathological findings which Binswanger considered essential to a disease which he called "encephalitis subcorticalis 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.²

The similarities between Bennett's¹ and Binswanger's own 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 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, very mild in the only case he described 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.³ Binswanger did not present a full account of the histopathology in his cases. This was left to Alzheimer, who first used the eponym "Binswanger's disease",⁴ and to Nissl.⁵ 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.²

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Pseudotumour cerebri and chronic benzene hexachloride (lindane) exposure

Pseudotumour cerebri, the syndrome of idiopathic intracranial hypertension and papilloedema in the absence of a tumour or obstructive hydrocephalus, 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 chronic 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 said he used a 20% concentrate to make dip and spraying 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 mm Hg 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 periventricular 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. 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 bilateral optic nerve sheath fenestration 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. 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 CNS stimulant known to cause headache, nausea, vomiting, diarrhoea, convulsions, muscle spasms, respiratory failure with cyanosis, coma, and death.⁵⁻⁸ "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,⁷ but the appearance of the optic discs and the spinal fluid pressure were not reported. The mechanism of lindane toxicity is unknown, although it is highly lipid soluble and may act as a gamma-aminobutyric acid-A (GABA-A) receptor antagonist to produce convulsive effects and interference with the production and utilisation of free ammonia in the brain.^{8,9} 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.² Lindane and chlordecone 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 (440 mm CSF) 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 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, like epiduritis or spinal metastasis.¹ 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 centropinal cystic lesion.

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

pighian 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 (CCDP, 5FU) followed by radiation therapy on the larynx (70 Gy) and upper mediastinum (45 Gy) with a 20 Gy dose on 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). Other parts of the spinal cord received 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 inner side of the right hand) nor autonomic dysfunction, and tendon reflexes were present except supinator and triceps jerks on both sides. Neurological examination elsewhere was unremarkable.

Electromyographic examination showed fibrillation, giant potentials and some fasciculations in right and left hand muscles, and reduced recruitment in forearm extensors. The more proximal muscles of the arms 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, lipidogram, syphilitic serology, viral serologies, serum lead 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 first neurological symptoms on a 1.5 Tesla superconducting magnet (Magnetom Siemens). Two excitations and a display matrix of 256×256 were used; the section thickness was 4 mm. Sagittal and axial studies showed a cystic lesion with well defined borders and a signal isointense with respect to CSF on T1 weighted images (TR = 500 ms, TE = 15 ms) and T2 weighted images (FIS P a = 10° , TR = 100 ms, TE = 27 ms) within the spinal cord from C4 to C6 and a smaller cavity behind the body of the atlas (figure). There was no enhancement of this image after IV injection of gadolinium and the cerebellar tonsils were not in an ectopic position.² The fatty replacement of spinal bone marrow due to radiation therapy accounted for the high signal intensity of the cervical spine on the same level.³ The thoracic MRI was normal. There was no change in neurological condition after eight months.

This case fulfils the strict criteria established for an accurate diagnosis of radiation myelopathy⁴: spinal cord included in the area of radiation; principal neurological manifestations compatible with the irradiated portion of the cord; other possible aetiologies such as compression due to metastases excluded by neurological investigations; and a latent period of at least nine months before the development of symptoms. Radiation myelopathies may be divided into five main

Figure Sagittal T1 weighted SE (500 ms/15 ms) image showing cystic hypointense cavity affecting spinal cord from C4 to C6 without ectopic tonsils. Note fatty replacement of cervical spine bone marrow from C2 to C6, responsible for hypersignal on T1 weighted images.

types:^{1,4} transient myelopathy (the more common and generally marked only by a Lhermitte sign); chronic progressive myelopathy; arrested radiation myelopathy; selective anterior horn cell injury or amyotrophy; and disseminated demyelination of the central nervous system. Our patient was in the fourth category as there was weakness and amyotrophy without significant sensory change, sphincter disturbance, or clinical involvement of corticospinal tract. There was electrophysiological evidence of an involvement of the anterior horn: giant potentials and normal motor and sensory conduction velocities. MRI data agree with this location. A coincidence, however, cannot be excluded but the only alternative diagnosis would be idiopathic syringomyelia for which the clinical picture, without a considerable sensory problem, the rapid progression of symptoms, the absence of Chiari or kyphoscoliosis would be atypical. There was no evidence for a syrinx secondary to an intraspinal tumour (thoracic MRI normal). Furthermore, before the wasting of the hands, a Lhermitte sign occurred, which is a common symptom of radiation myelopathies.

Nevertheless, to our knowledge such a clinical presentation has not previously been described. All reports of delayed lower motor neuron syndrome after radiation concern the legs, generally after irradiation for lymphomas or testicular malignant tumours.⁷⁻¹¹ These motor neuron syndromes usually present as a progressive weakness of the legs without sensory or sphincter disturbance. Development stops after some months or years and so far the prognosis is better than in other forms of myelopathy. Signs suggestive of a cervical anterior horn involvement (muscle wasting, fasciculations, tendon reflex loss) have been mentioned in some cases¹⁵ but they were associated with corticospinal and long sensory tract lesions (mainly Brown-Sequard syndrome).

The pathogenesis of radiation myelopathies remains controversial, but vascular changes are almost constant and are considered as the primary damage rather than neuronal or immunological disorders. As the grey matter is classically considered less vulnerable to radiation than white matter, the anterior horn is presumed to be less or not involved, although in several anatomical cases of radiation myelopathies an extension of lesions towards the anterior horns has been reported.^{5,6} There are no well documented anatomical reports of motor neuron syndromes after radiation and some controversy exists as to the site of lesions (anterior horn,

motor anterior roots, or plexus), but clinical and electrophysiological data from most cases suggest an involvement of the anterior horns.⁷ We know of only one report of MRI data from a patient with motor neuron syndrome of the legs and the thoracolumbar spinal cord images were normal.⁸ In a case reported by Lechevalier *et al* a patient had Brown-Sequard syndrome after radiation with proximal wasting and weakness of the left arm. Histological examination of the cervical spinal cord showed a centropinal necrosis with complete neuronal loss of the left anterior horn. Perhaps in our case the spinal cavity on MRI was secondary to necrosis involving the cervical anterior horns.

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The incidence of muscle cramp

In true muscle cramp there is sudden involuntary and painful shortening of muscle and visible or palpable knotting of muscle often with abnormal posture of the affected joint which is relieved by stretching or massage.¹ Epidemiological data on muscle cramp are sparse and based upon empiricism. Wilder in 1940 found that 19% of young soldiers, 33% of workers in a silk mill, and 67% of medical staff questioned had experienced muscle cramp. The incidence in a group of internal, neurological, and psychiatric patients was 40%.² In a study in 1947 of 100 men and 100 women aged 15-80 years, Hall found that 56% had suffered from cramp; of those aged 50 years or more, 70% had suffered.³ Of 121 college students questioned in 1956, Norris found that 115 had experienced spontaneous muscle cramp at least once, 18