Psychiatric symptomatology in early-onset Binswanger’s disease: two case reports

R.M. Lawrence¹ and J.C. Hillam²

¹ Department of Psychogeriatric Medicine and Neuropsychiatry, Purley Hospital, 50 Pampisford Rd, Purley, Surrey CR8 2NE, and ² Department of Psychiatry, Whittington Hospital, Highgate Hill, London N19 5NF, UK

Correspondence to: R.M. Lawrence at above address

We describe two cases of Binswanger’s disease of pre-senile onset which presented with affective and psychotic symptoms well before the appearance of cognitive deterioration and neurological signs, initially evading an accurate diagnosis. Psychiatrists should be aware of white matter disease and its role in the pathogenesis of psychiatric illness. Particular attention should be given to a history of hypertension as a risk factor in the early identification of these cases.

Keywords: Binswanger’s disease – Psychopathology – Hypertension

INTRODUCTION

Binswanger’s disease (BD) is characterized by white matter demyelination with lumbar narrowing of penetrating medullary arteries and arterioles, causing ultimately a subcortical-cortical disconnection dementia. Periventricular white matter disease, radiologically identifiable as leukoaraiosis (Hachinski et al., 1987), is positively linked with both advancing age and hypertension, which are major risk factors towards the development of vascular dementias (Shimada et al., 1990; Yao et al., 1992). The prevalence of BD is as high as 5% in the over 65 year old population (Goto et al., 1981; Peterson and Summergrad 1989). Babikian and Ropper (1987) reviewed the clinical histories of 46 autopsy-confirmed cases of BD. They found an average age at onset of 57 years, with demise about 5 years later. An acute neurological event marked the onset of the disease in one-third of the cases. In the remaining two-thirds, initial changes were more insidious. Behavioural changes and amnestic problems were early findings in 65% of cases. Psychiatrists are said to recognize BD infrequently (O’Brien, 1988; Summergrad and Peterson, 1989), but BD has been associated with a series of psychiatric features, including irritability, aggressiveness, depression, hypomania and paranoia. Such psychopathology may represent early symptoms of BD in the absence of neurological disturbance. An early diagnosis can be confirmed by radiological techniques, such as computed tomography (CT) or magnetic resonance imaging (MRI) scan (Van Swieten et al., 1991).

CASE REPORTS

Case 1

Mrs ST was a 58 year old right-handed hypertensive woman with a positive family history of depression in her two sisters. She first presented to the psychiatric services in 1990 at the age of 56 with an 8 month history of low mood, anhedonia and hypochondriacal delusions. She believed that she was unable to breathe because of a blocked nose and that she would die of suffocation. She also reported an increased sensitivity and intolerance of various odours coming from perfumes, food and even her hi-fi system. Her appetite was poor and she had lost more than one stone in weight during the last 12 months. She displayed diurnal mood variation with uplifting in the evening and suffered from early morning waking.

Physical examination was unremarkable except for mildly raised blood pressure (140/100 mmHg). The haematological and biochemical screenings were normal. She was treated with tricyclic antidepressants (clomipramine 150 mg daily) and neuroleptics were
ized hyperreflexia, increased tone in the upper limbs added later (trifluoperazine up to a maximum of 10 mg daily). She also received a course of six electro-convulsive therapy (ECT) treatments.

Her condition improved slightly until, in July 1991, she started complaining of forgetfulness. There were also instances of very bizarre behaviour, e.g. bringing a baby's push chair with her to the day hospital. A psychometric assessment at this time revealed a NART score of 119 and a relative impairment in her WAIS performance score (WAIS verbal score 113, WAIS performance score 95). Chest X-rays, ECG and EEG were normal. The CT brain scan showed bilateral periventricular hypodensity and mild to moderate ventricular enlargement.

Hard neurological signs appeared towards the end of 1992, with generalized hyperreflexia, stooped and dyspraxic gait and a left extensor plantar response. The patient became very apathetic and withdrawn. Psychometric tests were repeated and revealed a further deterioration (WAIS verbal score 113, WAIS performance score 88).

The final diagnosis was that of BD. The patient was discharged into a nursing home. When reviewed 4 months later she had also developed urinary incontinence.

DISCUSSION

Both cases fulfilled the criteria for the clinical diagnosis of BD, with evidence of dementia confirmed by neuropsychological tests, the presence of one risk factor for cerebrovascular disease (hypertension), subcortical features of parkinsonism, rigidity and gait disturbance. One focal pyramidal sign was present in Case 1 (ST). In both cases there was radiological evidence of bilateral periventricular leukoaraisis (Bennett et al., 1990; Roman et al., 1993). The onset of cognitive impairment prompted the radiological investigations which confirmed the presence of diffuse white matter disease. Before this, the symptomatology was strongly suggestive of a functional psychiatric disorder.

The association between schizophrenia and affective disorders with cerebrovascular disease has been well researched. Rabins et al. (1991) described a small sample of patients with schizophreniform symptoms after a stroke and found that right hemisphere lesions predominated and that the psychotic patients had greater ventricle to brain ratio (VBR), particularly in relation to frontal horns and third ventricle. Cerebral blood flow studies of individuals with schizophrenia have indicated both frontal hypoperfusion and relative hyperperfusion in the left medial temporal lobe depending on symptom profile (Friston et al., 1992; Liddle et al., 1992).

Studies have shown greater third and lateral VBR in patients with depression following stroke (Starkstein, 1988) and the left frontal lobe in particular appears to be implicated (Astrom et al., 1993). Depressed patients have been found to have increased VBRs (Dolan et al., 1985; Bird et al., 1986). Neuroimaging techniques have demonstrated reduction in cerebral blood flow and hypometabolism in the frontal lobes in depressed individuals (Baxter et al., 1985; Buchsbaum et al., 1986).
The association between leukoaraiosis and depression was reported by Coffey et al. (1987) in a prospective study of depressed non-demented patients who had been successfully treated with ECT. Periventricular white matter disease was discovered in all subjects on MRI. However, in 80% of cases the onset of depression was after the age of 60 years. With advancing years, the expectation of finding white matter disease increases. The study of younger psychiatric hypertensive patients with radiological evidence of leukoaraiosis would help clarify the pathoplastic influence of white matter disease on the pathogenesis and progression of some forms of psychiatric illness.

Hypertension in association with white matter disease is not always linked with the development of cognitive impairment, especially when there is a satisfactory degree of cerebral perfusion (Shimada et al., 1990). It may, however, have a bearing on non-cognitive psychopathology and resistance to the successful treatment of functional psychiatric illness. The mechanism of this neuropathological–phenomenological interaction is still unclear and is likely to be complex. Mood disturbance and psychotic phenomena are frequently found in patients suffering from primary cortical degenerative conditions and are widely reported in the scientific literature (Burns et al., 1990).

These two cases are a reminder that significant functional psychopathology may precede ominous neuropsychiatric developments by several months or years in patients suffering from BD. The suspicion of the impending risk of a vascular dementia could have possibly led to the prescription of warfarin, aspirin or other anticoagulant agents as well as hypotensive drugs. The antidepressant and neuroleptic treatment received by both patients does not call for criticism, as functional syndromes in demented patients can be pharmacologically treated, albeit in the short term (Joyce and Levy, 1989).

We suggest that BD should be considered in all hypertensive patients in their fifties with an apparent functional psychiatric illness of recent onset, especially if it proves resistant to treatment. Cognitive impairment may be initially very subtle, but should be vigorously sought. The assessment of this group of patients should include neuroimaging by CT or, preferably, MRI brain scan. Optimal management must also include careful control and treatment of blood pressure and cerebrovascular disease.

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