CASE REPORT

Subcortical Arteriosclerotic Encephalopathy (Binswanger's Disease)

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Abstract

We report the case of a patient who presented to us with dementia, progressive gait instability and features of pseudobulbar palsy. Her MRI showed bilateral leukoaraiosis. She was diagnosed as a case of subcortical arteriosclerotic encephalopathy (Binswanger's disease) based on the clinical and radiological findings as per the diagnostic criteria proposed by Bennet.

Key Words

Subcortical Arteriosclerotic Encephalopathy, Binswanger's Disease, Dementia

Introduction

Binswanger's disease (BD) is a poorly understood form of vascular dementia and for the years was considered a relatively rare disorder diganosed at necropsy (1).We report the case of a patient who presented to us with dementia, progressive gait instability and features of pseudobulbar palsy. Her MRI showed bilateral leukoaraiosis. She was diagnosed as a case of subcortical arteriosclerotic encephalopathy (Binswanger's disease) based on the clinical and radiological findings as per the diagnostic criteria proposed by Bennet *et al* (1).

Case Report

A 52 years female presented to us with history of gait disturbances for last 1 ½ years and difficulty in speaking for the same period of time. On examination she was found to have "magnetic" gait and features of pseudobulbar palsy like dysarthria, dysphonia and inappropriate laughing and crying. Her power was 4/5 in all the four limbs, reflexes were brisk and plantar showed bilateral extensor response. Rigidity was present in all the four limbs. Dementia was present and her score on MMSE was 19/30. On admission,her BP was 190/130 mm Hg and fundoscopy revealed grade II hypertensive retinopathy. With these clinical findings, in the background of chronic hypertension, diagnosis of vascular dementia was suspected.

CT head showed multiple focal hypodense lesions in the periventricular region, bilateral basal ganglia and pons. MRI brain was done which revealed multiple focal lesions, measuring 2-8 mm, hypointense on T1-weighted (*Fig 1*) and hyperintense on T2-weighted images (*Fig 2 & 3*), in periventricular region, centrum semiovale, bilateral basal ganglia and pons. Multiple punctate haemorrhagic foci were seen interspersed within these lesions. There was sparing of the subcortical U-fibres and the corpus callosum. Ventricular system, cisternal spaces and sulcul spaces were prominent and mild cerebral atrophy was also seen. MR Angiogram was normal.

CSF examination was normal and her tests for syphilis were negative. There was no family history of similar complaints.

With these white matter changes detected on CT and MRI and clinical findings of dementia, gait ataxia and pseudobulbar palsy, in the background of chronic hypertension, a diagnosis of subcortical arteriosclerotic encephalopathy (SAE) (Binswanger's disease) was kept in accordance with the diagnostic criteria proposed by Bennetet *et al* (1990) (1).

Discussion

White matter changes, as observed in our present case (hypodensities on CT and hyperintensities on T2-weighted MRI in periventricular region), are termed "leukoaraiosis" (2). These changes may be seen in various pathological conditions like Binswanger's disease (BD), Alzheimer's disease, multiple sclerosis, CADASIL, progressive multifocal leukoencephalopathy as well as in normal elderly individuals. Clinical picture helps to differentiate most of these causes. Clinical features of BD include mental deterioration, neurological deficits, frequent

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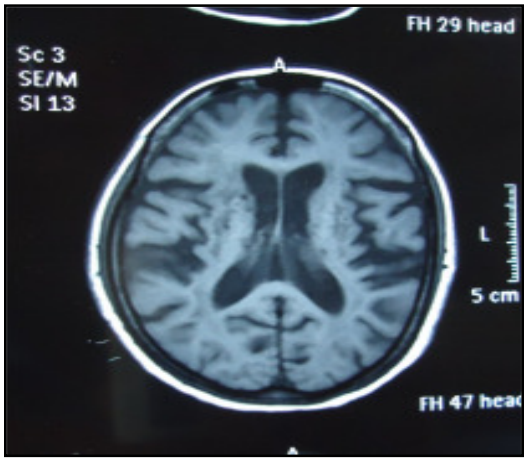

Fig 1. Axial T1-Weighted Image Showing Multiple Focal Hypointense Lesions in the Region of Basal Ganglia Interspersed with Haemorrhagic Foci

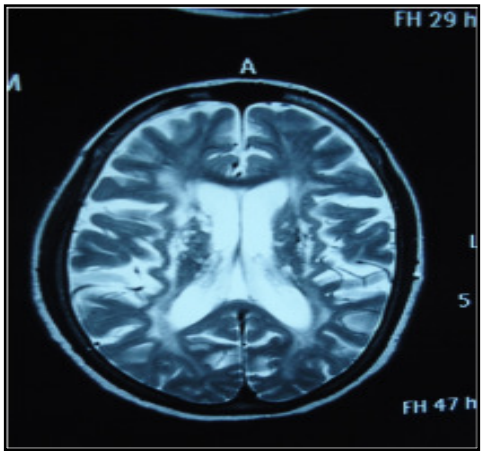

Fig 2. Axial T2-Weighted Image Reveals Multiple Focal Hyperintense Lesions in the Region of Basal Ganglia Interspersed with Haemorrhagic Foci. Subcortical U-Fibres, Corpus and Corpus Callosum are Spared

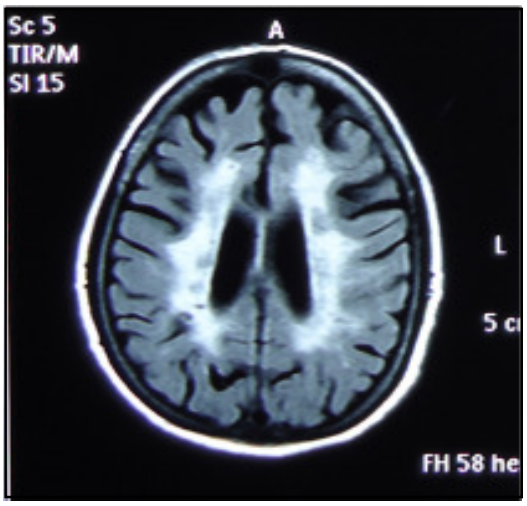

Fig 3. Axial T2-Weighted FLAIR Image Showing Multiple Focal Hyperintense Lesions in Periventricular Region, Centrum Semiovaleand Bilateral Basal Ganglia

occurrence of TIAs, pseudobulbar disturbances (e.g. dysarthria, dysphagia, inappropriate laughing and crying, small stepped gait), and cerebellar and extrapyramidal signs. The course is punctated by frequent falls, seizures, syncopes and urinary incontinence. Associate vascular abnormalities, hypertension, in particular, are found in majority of patients (3,4).

Diagnostic criteria proposed by Bennetet *et al* in 1990, help in making antemortem diagnosis of possible BD with high specificity (1). These include (1) dementia; (2) one finding from two of the following three groups: (a) presence of vascular risk factor or evidence of systemic vascular disease (e.g. hypertension, diabetes) (b) evidence of focal cerebrovascular disease (e.g. a history of stroke) (c) evidence of subcortical cerebral dysfunction (e.g. a Parkinsonian, magnetic or senile gait); and (3) bilateral leukoaraiosis on CT or MRI.1 The patient, in our present case, had dementia, was hypertensive, had gait ataxia (magnetic gait) and exhibit features of pseudobulbar palsy. Her CT and MRI showed bilateral, multiple subcortical white matter changes (leukoaraiosis). Hence, the diagnostic criteria proposed by Bennetet al were fulfilled in the present case.

Therefore, leukoaraiosis, along with the clinical picture and associated signs (e.g. arterial hypertension), may allow, in particular cases, a clinical diagnosis of possible SAE (BD). A definite diagnosis of SAE can be achieved only on pathological grounds (4).

This case highlights the consideration for rare possibility of SAE (BD) in the patients presenting with dementia and neurological deficits and having evidence of bilateral leukoaraiosis on CT and/or MRI.

References

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