Review Article

Subcortical arteriosclerotic encephalopathy (Binswanger's disease)

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Summary

Subcortical arteriosclerotic encephalopathy is a chronic vascular dementia with hydrocephalus characterized clinically by : (*i*) subacute focal neurological deficit; (*ii*) acute strokes; (*iii*) dementia; (*iv*) motor signs and pseudobulbar palsy; (*v*) hydrocephalus; (*vi*) persistent hypertension and systemic vascular disease; and (*vii*) a lengthy course. The pathogenesis is most probably ischaemic change related to subacute hypertensive encephalopathy. The pathological changes include severe central nervous system disease characterized by loss of white matter with gliosis, and arterial and arteriolar sclerosis of small penetrating cerebral blood vessels.

The differential diagnosis includes vascular pseudobulbar palsy, multi-infarct dementia and senile dementia (Alzheimer's disease). Treatment includes blood pressure control as well as management of other factors known to affect vascular disease (diabetes mellitus).

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In 1894, Otto Binswanger[1] reported 8 cases of pronounced atrophy of cerebral white matter associated with hydrocephalus. He summarized the clinical picture as follows: the disease usually begins between 50 and 65 years of age. There is gradual mental deterioration, manifested in particular by disorder of associations between motor and sensory regions, with aphasia, hemianopia, hemiparesis and hemi-anaesthesia. These symptoms are quite stable, and are accompanied by progressive mental decline leading eventually to complete loss of mental faculties. During the course of the disease 'apoplectiform attacks' with dizziness, hemiplegia and aphasia occur. In most cases progression is very slow, extending over up to 10 years.

In 1902, Alzheimer[2] described numerous foci of severe gliosis in the cerebral white matter, internal capsule, lenticular nuclei, thalamus and pons. He attributed these to arteriosclerosis of the long, deep vessels, causing atrophy of the white matter. In 1965 Olszewski[3] called the condition subcortical arteriosclerotic encephalopathy (SAE), emphasizing the extensive pathological changes in the small arteries and arterioles of the basal ganglia and cerebral white matter.

Until recently diagnosis has been based on autopsy data, the clinical features being common to other disorders such as multi-infarct dementia and Alzheimer's disease. Biemond[4] was the first to propose a scheme for diagnosis during life. A comprehensive account of the clinical features was subsequently given by Caplan and Schoene.[5] Their patients' clinical features included hypertension, acute strokes, subacute neurological deficit, long plateau periods, dementia, pseudobulbar palsy and hydrocephalus. Computed tomography (CT) has made a further contribution to the premorbid diagnosis; a low grade of attenuation of white matter was the main feature correlating with a clinical picture of dementia, focal neurological deficit and hypertension in the cases reported by Loizou *et al.*[6]

Main clinical features of SAE

Pre-existing hypertension is present in all cases. All pathologically confirmed cases had moderate-to-severe disease of cerebral arteries and other features of longstanding hypertension (such as left ventricular hypertrophy). Focal neurological deficit develops gradually, over weeks or months in most cases. This is the principal feature separating this condition from a lacunar state characterized by multiple focal infarcts.

Acute stroke, often followed by an acute neurological deficit in less than 72 hours, occurs in nearly all cases. Motor involvement includes asymmetrical weakness with pyramidal signs and pseudobulbar palsy (invariably present). Dementia occurs in nearly all patients, but memory loss is less common than in senile dementia. Hydrocephalus is characterized by ventricular dilatation without cortical atrophy (in all cases) secondary to loss of white matter.

The clinical course is usually 3 - 10 years (average 6,5 years). Clinical 'plateaux' or periods of improvement may occasionally occur.

The CSF pressure is usually normal and carotid angiography is usually negative. CT reveals symmetrical hydrocephalus and often low-attenuation areas in the white matter of the frontal and parietal lobes, usually symmetrical with subcortical lacunae or cortical infarction and mild-to-moderate cerebral atrophy and symmetrical ventricular dilatation.

Differential diagnosis

SAE must be differentiated both clinically and radiologically from similar conditions, of which vascular pseudobulbar palsy and multi-infarct dementia are the commonest. The former is characterized by multiple focal infarcts due to lacunae, whereas the latter is due to accumulation of cerebral infarcts from extracranial thrombo-embolic disease or intracranial small-vessel disease. In SAE the focal deficit usually develops over weeks or months, whereas a gradual onset is uncommon in occlusive disease of the large cerebral vessels. Also, in SAE the subacute neurological deficit does not show up on arteriography. The presence of subacute neurological deficit with low attenuation of white matter on CT is not found in multi-infarct dementia.

SAE should be further differentiated from 'normal-pressure' hydrocephalus, with which it shares the features of ventricular dilatation and mild cortical atrophy, by the invariable presence of more laterally extending low attenuation of frontal and parietal white matter, and the frequent occurrence of infarcts.

Other vascular conditions may give rise to diagnostic difficulties on CT scanning. In hypertensive encephalopathy, low-density areas may occur in both grey and white matter owing to micro-infarcts and cerebral oedema; the absence of cortical atrophy and infarcts of a size visible on CT will differentiate this from SAE.

Pathogenesis and pathological findings

Three mechanisms have been suggested to explain the development of regional loss of white matter with gliosis, which is the major pathological feature of the condition:[5] (*a*) SAE represents a special type of vascular disease similar to that found in hypertensive patients with lacunae; (*b*) poor perfusion of diffuse regions of white matter is secondary to thickening of the long, penetrating cerebral vessels; and (*c*) the changes in the white matter are due to a subacute or chronic form of hypertensive encephalopathy, with local fluid transudation and focal cerebral oedema leading to loss of tissue with gliosis.

It is now believed that the essential change in SAE is thickening of the long, penetrating cerebral arteries, arterioles and capillaries, usually related to systemic hypertension and in some patients possibly related to atherosclerosis or other vascular factors. The vascular disease produces loss of white matter by diffuse ischaemia or leakage of fluid with subsequent gliosis. Recognition of the syndrome may explain some subacute neurological deficits in hypertensive patients with normal arteriograms and may lead to more detailed investigation.

Therapy

The only recommended treatment is control of blood pressure and management of any other factors which may be associated with vascular disease, such as diabetes mellitus. Shunting procedures are not likely to be of benefit in patients with SAE.

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