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Review Article

Binswanger's Disease

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Abstract

On autopsy of brain, in some cases ischemic periventricular leukoencephalopathy is found not only this, CT SCAN and MRI has also shown deep white matter lesions in the aged persons usually present between 54 to 66 years of age ,this conditions is called as Binswanger's disease is a progressive neurological disorder and is also called as sub cortical vascular dementia .There is no cure for this disease, which was resulted from thickening or narrowing of arteries (atherosclerosis) that supply blood to white matter of brain and that leads to the death of brain, brain tissues particularly basal ganglia and thalamus which are the deep structures of brain .it may be caused by rare hereditary disease called CADASIL. Symptoms usually seen in the patient's are, mood changes, loss of the ability to focus on tasks, a deterioration in thought processes (e.g., loss of memory and cognition), and mood changes. It does posses some risk factors such as hypertension and amyloid angiopathy, impaired auto regulation of cerebral blood flow in the elderly, and periventricular hypo-perfusion due to cardiac failure, arrhythmias, and hypotension. But for this disease, there is no specific treatment. Treatment is symptomatic; the most characteristic feature of Binswanger's disease is psychomotor slowness. Persons with this disease often die within five years of the onset of the disease. It can coexist with Alzheimer's disease and some other diseases. Scientists at the National Institute of Neurological Disorders and Stroke are reevaluating the definitions for many forms of dementia, including Binswanger disease.

Key words: Peri ventricular leukoencephalopathy, Abulic, CADASIL, Atherosclerosis.

Introduction

Binswanger disease is a progressive neurological disorder which is considered as a type of vascular dementia and Previously known as multi-infarct or post-stroke dementia, associated with sub cortical white matter disease Caused by arteriosclerosis and thromboembolism Affecting the blood vessels that supply blood to the white-matter and deep structures of the brain (basal

ganglia and thalamus) Occurred in the people by the age of 55-65 or above this leads to death of the patient.

History

Binswanger in 1894 described patients with a subtype of cerebral arteriosclerosis[1]. Their clinical courses were characterized by slowly progressive intellectual deterioration starting in the sixth decade, punctuated by seizures and stroke like events. Extensive white matter atrophy, predominantly in a periventricular and temporal-occipital distribution, spared the basal ganglia and the cortical mantle. He named this disease 'encenphailitis subcorticalis chronica progressive.' But he didn't conduct any microscopic investigation, which was incomplete report and then after. Alzheimer in studying Binswanger's work with pathological evidence that concluded and supported Binswanger's ideas and hypotheses. Alzheimer renamed this disease as a Binswanger's disease.

In the late 19th century vascular dementia was heavily studied, however, by 1910 scientists were lumping Binswanger's disease with all other subcortical and cortical dementia and labeling everything senile dementia despite all previous research and efforts to distinguish this disease from the rest[2,3].In 1962 J. Olszewski published an extensive review of all literature about Binswanger's disease so far. He discovered that some of the information in the original reports was incorrect and that at least some of the patients studied in these cases probably had neurosyphilis or other types of dementia. Even with these errors, Olszewski translated the articles by Binswanger, Alzheimer, and Nissl and presented two new cases[4]. He emphasized the presence of associated lacunes and cortical infarcts, and suggested the title "Sub cortical Arteriosclerotic Encephalopathy. "Concluded that Binswanger disease did exist as a subset of cerebral arteriosclerosis and then the term multi-infarct dementia was coined and all vascular dementia was grouped into one category. Because of this, the specific names of these types of this dementia, including Binswanger's disease were lost.

Classification of dementia[5]

Alzheimer's disease	Includes about 50%-70% of cases	Difficulty in remembering names and recent events is often an early clinical symptom: apathy and depression are also often early symptoms. Later symptoms include impaired judgment, disorientation, confusion, behavior changes and difficulty speaking and etc
Vascular dementia	Includes20%of all cases	It is a decline in thinking skills caused by conditions that block or reduce blood flow to the brain, depriving brain cells of vital oxygen and nutrients. This often results from a stroke or mini strokes.
Lewy body dementia	Includes15%-20%of all cases	Have memory loss and thinking problems common in Alzheimer's, but are more likely than people with Alzheimer's
Parkinson's disease dementia	Includes about5% of cases	Problems with movement are a common symptom early in the disease. If dementia develops, symptoms are often similar to dementia with lewy bodies.
Front temporal dementia	Includes about 5% of all cases	Typical symptoms include changes in personality and behavior and difficulty with language. Nerve cells in the front and side regions of the brain especially affected.

And some other cases were also found but that occurrence is less when compare.

Alzheimer's diagnostic centers created specific criteria known as the Hachinski's Ischemic Scale which became the standard for diagnosing MID or vascular dementia. Because of the complicated history of Binswanger's disease and the fact that it was overlooked as a disease at all for so many years, leads us to believe that many patients have been misdiagnosed as Alzheimer's for years[6].This leads us to believe that Binswanger's is more prominent in the population than once thought

Etiology

Actually this disease mainly occur due to affecting the blood vessels that supply blood to the brain and this was due to

- The width of the media of arterioles in BD was found to be greater than in comparable cases with intracerebral haemorrhage or hypertensive encephalopathy[7].
- Amyloid deposition in the long medullary arteries could produce distal tissue ischemia and vessel wall hypoxia with consequent transudation of plasma.
- Ischemic nature, of the characteristic white matter lesions.
- Vasculopathy including abnormal blood brain barrier.
- Hyper coagulation: significant elevation in levels of thrombin-antithrombin complex, have been reported in patients with Binswanger's disease.

  In some cases this disease can be caused be to CADASIL.

What is Cadasil?

Cerebral autosomal dominant arteriopathy with sub cortical infarcts and leukoencephalopathy, usually called CADASIL, is an inherited condition that causes stroke and other impairments.[8]This condition affects blood flow in small blood vessels, particularly cerebral vessels within the brain. Mutations in the on chromosome 19q12 involving the*NOTCH3*[9] results in a small vessel and arterial stenosis secondary to fibrotic thickening of the basement membrane of the vessels.

The *NOTCH3* gene provides instructions for producing the Notch3 receptor protein, which is important for the normal function and survival of vascular smooth muscle cells

Symptoms of the patients who are having a Binswanger's disease

- Mutism-Inability or unwillingness to speak
- Abulia-Absence of or decreased ability to exercise willpower
- Apathetic-Without interest
- Dysarthria-Impairment or clumsiness of words
- Dysphagia-Inability to swallow or difficulty in swallowing
- Hemipalgia-Pain in one side of hand[10].

This are the symptoms that's based on physical behavior in order to confirm the presence of disease patient need to diagnose

Diagnosis

- **CT scan**

It has shown diminished white matter density and enlarged ventricles, frequently with mild cortical atrophy and lacunar infarctions. White matter hypodensity has been mainly periventricular, extending into the Centrum semiovale, with the region just anterior to the frontal horns of the lateral ventricles frequently abnormal[11].Posterior white matter has occasionally been the initial site of involvement radiographically.

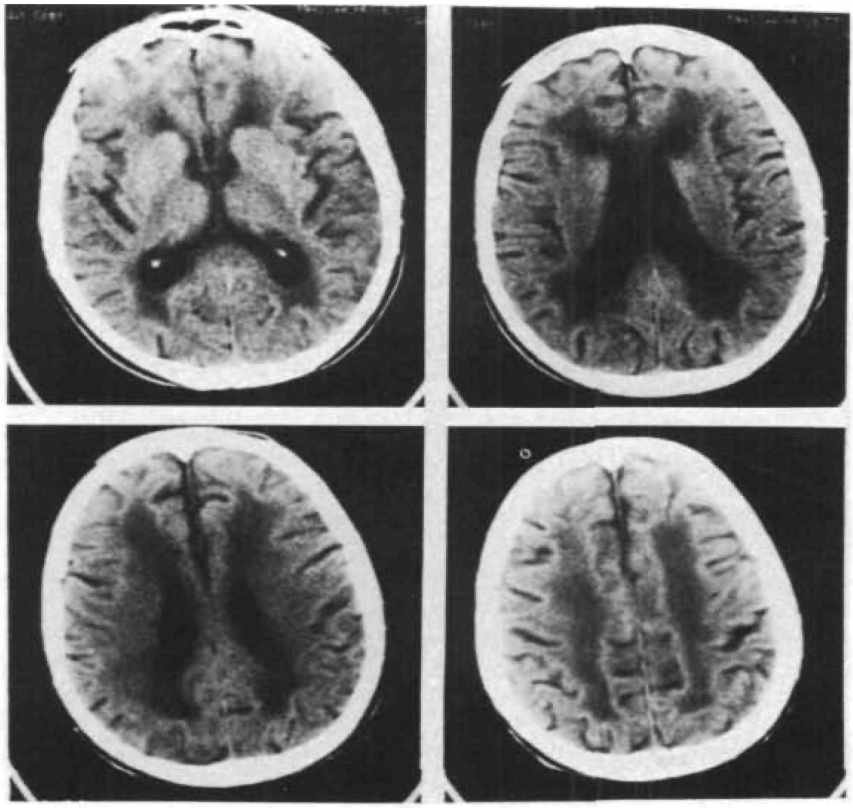

Figure 1: CT scan shows peri ventricular and Centrum semiovale low density with enlarged ventricles and widened sulci

- **Electroencephalogram. EEG,** Predominantly focal or asymmetric slowing, especially over the temporal or frontal areas, was also frequent[12].
- **Mini Mental Test (MMT)** But there is a little confusion for confirmation of this disease in order to confirm physician should follow following criteria

Criteria for the clinical diagnosis of possible Binswanger's disease

1. Dementia must be established.
2. One finding from two of the following three groups must be present
   A. Presence of vascular risk factor
   B. Evidence of focal cerebrovascular disease
C. Evidence of sub cortical cerebral dysfunction
3. The radiological criteria require bilateral leukoaraiosis on CT[13].

Treatment

The ischemic brain damage in Binswanger disease is not reversible, which can be cured easily, so treatment is focused on reducing risk factors for stroke, thereby retarding progression of the disease[14].

Treatment usually involves the use of Anti-hypertensive drugs to control blood pressure, Antiplatelet drugs (e.g. Aspirin) or warfarin to reduce thromboembolism, Statins to reduce atherosclerosis and Antidepressant drugs are helpful in the management of depression associated with Binswanger disease. Other treatment is symptomatic and supportive recent drug trials with the drug *Memantine* have shown improved cognition and stabilization of global functioning and behavior. It has been shown that current Alzheimer's medication, *Donepezil* (trade name Aricept), may help Binswanger's Disease patients as well.

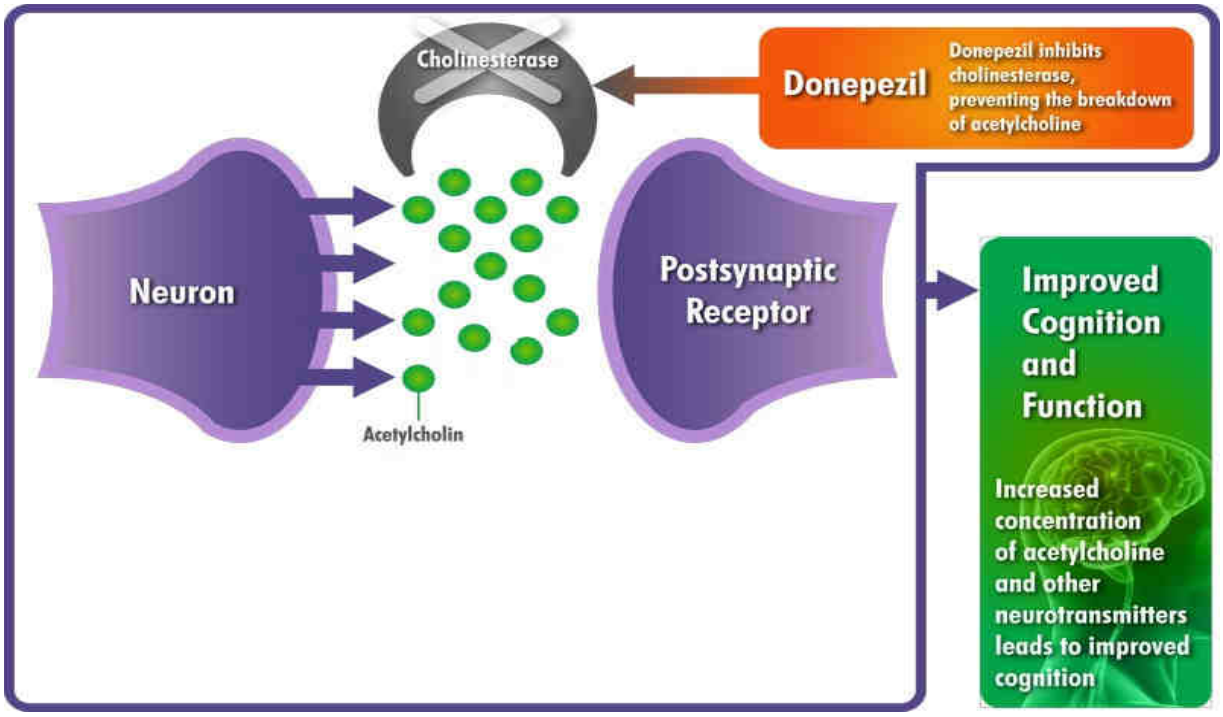

Figure 2: Mechanism of action of donepezil, it inhibits cholinesterase which leads to inhibition of breakdown of acetylcholine and this leads to improvement of cognition in patients

Management of Binswanger's diseased patient

Patient with Binswanger's disease can be maintained and at first he/she need to be encouraged to lead a healthy lifestyle. cessation of smoking, dietary modifications and regular exercise are key in counseling. low fat diet provides an adequate nutrition and promotes glycemic control should be used to control diabetic conditions and slow the process of atherosclerosis[17]. Exercises have been shown to aid in weight control, blood control stabilization and its conditions Management of cognitive and psychological symptom is also important in order to do this patient should be Challenge brain-Improve ability to retain and retrieve memories. Set aside some time in the evening to recall the day's events, which can build memory capacity. Learning new skills, such as a foreign language or how to paint, can also help build brain capacity if done consistently. Maintain social activity-Holding up a conversation may require more effort, but staying in touch with friends and family, face to face, can help maintain cognition. Regularly schedule activities that lead to involvement of interaction with other people and by some other ways patient conditions canbemaintained[15].

Conclusion

Leukoencephalopathy of Binswanger's disease appears will be due to chronic ischemia in the terminal zones of the medullary arteries, which will be determined by anatomic factors, variations in perfusion pressure of blood, and probably by hemorrheologic factors[16]. More controversial is the role of alterations of the blood-brain barrier linked to hypertensive episodes with chronic vasogenic edema. Recently, ventricular dilatation from Binswanger's-type lesions leading to normal pressure hydrocephalus has been noted, [15] as well as the possible causal contribution of periventricular venous collagenosis.

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