Corticobasal Syndrome

Corticobasal syndrome (CBS) is a form of atypical parkinsonism (a parkinsonism-plus syndrome), which means that it shares some features with Parkinson’s disease such as stiffness (rigidity), tremor at rest, slowness of movement (bradykinesia) and postural instability (balance difficulties). It may also cause problems with memory and thinking. CBS, however, is distinct from Parkinson’s disease in regards to other clinical features and its response to treatment.

CBS was first described by Rebeiz and colleagues in 1968. There are some variations of the name of CBS such as corticobasal disease or corticobasal ganglionic degeneration. The name implies the parts of the brain damaged. CBS results in gradual loss of nerve cells (neurodegeneration) in the surface of the brain (the cerebral cortical areas) as well as deep structures (the basal ganglia). These brain regions are heavily involved in the control of movement, so CBS causes problems with mobility. In contrast to other types of atypical parkinsonism, the neurodegeneration in CBS is markedly asymmetrical, thus the symptoms usually start on one side of body and remain worse on that half throughout the course of the disease.

Symptoms and Signs

The most characteristic presenting feature of CBS is the gradual loss of use of one hand or leg (called "apraxia"). Patients may also experience abnormal postures of their limbs or neck (dystonia), painful rigidity, muscle jerking (myoclonus), and eventually irreversible muscle contractures, all involving the same side of the body. They may have myoclonus of one hand while at rest or with activity. On occasion, an affected limb can seem to have a "mind of its own," and make seemingly purposeful movements that the patient cannot control. This problem, known as "alien" hand or limb, is sometimes accompanied by the feeling that one's limb is somehow foreign. Some patients also have language dysfunction (e.g. primary progressive aphasia) or slurred speech (dysarthria), difficulty opening or moving their eyes, as well as difficulties with their concentration, and behavior. Although disorders of thinking and memory (cognitive changes) may be noted early in the disease, dementia usually occurs only in more advanced stages. There may be loss of inhibition and changes in behavior such that patients speak rudely or do not show empathy. On examination, there is often loss of sensation in one or both sides of the body, even though patients typically do not complain of numbness. The symptoms of CBS usually worsen over 3-8 years and often result in great disability, including the inability to communicate or ambulate. Walking and balance
difficulties, however, occur later in patients with CBS compared with other forms of atypical parkinsonism such as progressive supranuclear palsy (PSP).

**Diagnosis**

There is no diagnostic test for CBS, but a neurologist usually suspects the diagnosis based on a patient's history, physical examination and clinical course. Early in the disease, it can be challenging to differentiate CBS from other forms of parkinsonism, such as Parkinson's disease or PSP and in some cases, there is an overlap in clinical features between the different parkinsonian disorders. Imaging with CT or MRI may show asymmetrical shrinkage (atrophy) of the cerebral cortex (brain surface) on the side opposite to the more affected limbs. Brain scans, however, cannot yet reliably distinguish CBs from other similar neurodegenerative diseases. In some cases, the diagnosis of CBS cannot be confirmed until an autopsy examination of the brain is performed, which usually shows "ballooned" neurons, protein aggregations (neuronal inclusions) and other characteristic abnormalities resulting from abnormal accumulation of the tau protein (CBS is a "tauopathy").

**Cause**

The cause of CBS is not yet known. Like other neurodegenerative diseases, patients with CBS accumulate misfolded proteins within specific brain cells. Mishandling of tau, a protein that normally acts to stabilize the cellular skeleton of neurons (nerve cells), appears to play a major role but the details remain unclear. CBS is usually not an inherited condition.

**Treatment**

As with the other atypical parkinsonian syndromes, treatment with levodopa and related medications sometimes lessens muscle rigidity and improves mobility, but results are often disappointing. Muscle spasms and jerking can be reduced with muscle relaxants, such as clonazepam, and with botulinum toxin injections into affected parts of the body. Medications for memory loss, depression and anxiety may be useful in patients with these problems.

Other treatments for CBS include physical therapy and stretching exercises designed to relieve rigidity and to prevent contractures and deformities as well as to maintain good strength and condition of muscles. Devices which make walking safer, such as a cane or walker, can be helpful. Speech, physical, and occupational therapy may be beneficial. Because of swallowing problems, some patients require placement of a feeding tube (PEG) directly into the stomach to maintain adequate nutrition and prevent aspiration pneumonia. If general health and nutrition can be maintained, some CBs patients live for several years after the onset of symptoms, although their quality of life in the advanced stages of the disease is usually significantly impaired.
At present, there are no therapies that can reverse or even slow the progression of CBS. Furthermore, since CBS is quite rare, clinical drug trials are sometimes not available for affected patients. Nonetheless, there is reason for hope. Because the biology of CBS may be related to other neurodegenerative diseases, it is possible that therapies designed for other conditions will also prove helpful for patients with CBS.

**Selected References**


Helpful organizations

**CurePSP: Foundation for PSP, CBD and Related Brain Diseases**


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