

Case Report

Improving Quality of Life & Possibly Increasing Life Expectancy with Multiple Complementary Therapies in a Case of Corticobasal Degeneration [CBD]

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Abstract

Corticobasal degeneration (CBD) is a rare neurodegenerative disorder of mid- to late-adult life. Corticobasal degeneration (CBD) is a sporadic tauopathy that presents with a varied combination of motor, cognitive and behavioral features, making its diagnosis difficult. CBD has high morbidity and poor prognosis, with no effective therapy at present. The clinical symptoms of CBD, predominantly extrapyramidal signs (bradykinesia and rigidity) and apraxia, affect often only one body side in the onset phase. Corticobasal degeneration (CBD) is an uncommon condition that can cause gradually worsening problems with movement, speech, memory and swallowing. Clinical-pathological studies have illustrated that CBD can present with diverse clinical phenotypes, including a non-fluent, agrammatic primary progressive aphasia syndrome, a behavioral, dysexecutive and visuospatial syndrome, as well as a progressive supranuclear palsy-like syndrome. Neuropathological studies reveal neuronal loss, gliosis, and achromasia chiefly in frontal and parietal cortex, as well as in basal ganglia and substantia nigra.

Herein we want to present a case of 48 years man, a diagnosed case of early CBD by a neurologist. He is a hairdresser by occupation & owns a hair, Salon. He specifically complained of inability to hold scissors or razors in his Right hand to shave or to do a haircut for his customer. His hair salon being only income source for him & his family, his inability to do his job had made him & his family suffer a lot.

We treated him with our Unique Treatment Method which comprises of Physical therapy combined with complementary therapies-Acupuncture & Panchakarma. After completing a 10-day Therapy session which was given on daycare basis, patient could hold scissors/razors in his hand & could do the fine movements required for the task of doing a haircut & shave.

We wanted to highlight that if diagnosed in early stage of disease & treated with the Unique Treatment Method which we have developed over three decades of practice & experience, even in a rare, progressive, neurodegenerative case of CBD, we could see significant reversal of symptoms which ultimately improved his quality of life.

Keywords: *Corticobasal degeneration (CBD), Complimentary Therapies, Acupuncture, Physical Therapy, Panchakarma.*

Background

Rebeiz et al. first described in 1967 three patients with slowly progressive ‘clumsiness’ and a variety of asymmetrical extrapyramidal signs, including rigidity, dystonia and myoclonus, with relatively intact cognitive functions. They introduced the term “corticonigral degeneration with neuronal achromasia” to describe this entity. This disorder was largely forgotten until 1989, when Marsden et al. introduced the term ‘Corticobasal degeneration’ Corticobasal refers to two parts of the brain that the disease affects. Degeneration refers to deterioration or loss of function. CBD affects-Cerebral cortex & Basal ganglia.

Corticobasal degeneration (CBD) is a rare condition in which areas of the brain shrink and nerve cells degenerate and die over time. CBD is caused by increasing numbers of brain cells becoming damaged or dying over time. The disease affects the area of the brain that processes information and brain structures that control movement. This degeneration results in growing difficulty in movement on one or both sides of the body. The condition may cause one to have poor coordination, stiffness, difficulty thinking and trouble with speech or language. People with CBD may describe having a hard time controlling their arm or leg. Some people with CBD have language problems first and may develop movement problems later. Thinking and behavior changes may happen either at the beginning or later in the disease. Robust epidemiological data on CBD is lacking because of the rarity of the disease, the great clinical pathological heterogeneity of the disorder and the lack of established criteria until recently. It is estimated that it is ten times rarer than progressive supranuclear palsy (PSP). The mean age at disease onset is about 64 years, whereas the youngest pathologically confirmed case was 43 years old. There could be a slight female predominance. Mean survival is estimated at 6.5 years.

Case Report

A 48-year-old man, having no addiction with no past history of any major illness, hairdresser by occupation, presented to us with complaints of clumsiness in his Right hand, abnormal shaky movements of right thumb, inability to do fine movements like buttoning the shirt & combing the hair. He specifically complained that he could

not hold scissors/ razor blade in his hand to do his daily job of shaving & hair cutting in his salon. He also complained of difficulty in walking, inability to wear chappals/footwear. He has been suffering from these symptoms for the last 1.5 years & his complaints have been increasing day by day.

He consulted a neurophysician a couple of times before coming to us. The neurologist clinically diagnosed it as a case of Corticobasal Degeneration (CBD). The treating neurologist prescribed him Levodopa+ Carbidopa combination, Trihexyphenidyl 2 mg, clonazepam 0.5 mg & baclofen 10 mg. But there was not much satisfactory response to these drugs.

When he presented in our OPD, on examination- he had abnormal gait with short steps, had tendency to drag his right leg, weak grip on his Rt. Hand with dystonia, bradykinesia & spasticity on his Rt. Side. His cognition was normal, speech & language were normal, vision & eye movements were normal, memory & behavior was normal. His vital parameters were within normal limits, His Cardiovascular, Respiratory & abdominal system did not reveal any abnormality.

His MRI brain was normal. His blood reports were-Hb-12.6, blood sugar level (Random-124), serum creatinine-1.2, HIV-Not Reactive, HBsAg-Negative.

Going with the diagnosis of Early CBD done by previously treating neurophysician, we continued with the same medicines as these are the medications that help manage the symptoms. As there is no cure for CBD yet & no disease modifying medicine is available & approved yet, we continued the symptomatic treatment. In addition, we treated him with a session of Unique Treatment Method with multidisciplinary approach combining & giving multiple complementary therapies simultaneously under one roof. It is a 10-day session comprising of Acupuncture, Panchakarma & Physical therapy.

Acupuncture

Acupuncture -Traditional Chinese medicine explains acupuncture as a technique for balancing the flow of energy or life force — known as chi or qi (chee) — believed to flow through pathways (meridians) in our body. Acupuncture involves the insertion of very thin needles through our skin at strategic points on our body. By inserting needles into specific points along these meridians, our energy flow gets re-balanced, achieves balance of Yin & Yang which ultimately helps in curing disease & restoring health.

Acupuncture Points given were- GV 20, ST 4, ST 36, SP 9, SP 6, GB 34, UB 60, K 3, ST 41, ST 44, GB 21, LI 15, SI 9, SI 10, TW 5, TW 4, LI 4, GV 14, UB 11, GV 6, GV 4, UB 40, UB 51

Panchakarma

Panchakarma is a treatment program for the body, mind and consciousness that cleanses and rejuvenates. It is based on Ayurvedic principles that every human is unique phenomenon manifested through the five basic elements of Ether, Air, Fire, Water and Earth. Combination of these elements are three doshas (tridosha): Vata, Pitta, and Kapha, and their balance is unique to each individual. When this doshic balance is disturbed, it creates disorder resulting in disease. Panchakarma is done individually for each person with their specific constitution and specific disorder in mind, thus it requires close observation and supervision. Treatment starts with pre-purification Measures of Snehana and Swedana, and then cleansing methods – Shodhanas are applied.

In **Ayurved Panchakarma** he was given-

- Snehan** - Snehan refers to the massage of medicated oil over whole body for a specific period.
- Sweadan**- Swedana, means to "perspire". It is used in Ayurvedic treatment. It is also called steam therapy.
- Shirodhara** -Shirodhara is a classical and a well-established ayurvedic procedure of slowly and steadily dripping medicated oil or other liquids on the forehead. This procedure induces a relaxed state of awareness that results in a dynamic psycho-somatic balance.
- Nasya**- wherein Ayurvedic medicine in the oil forms is infused into the nostrils.
- Basti**-Basti is the introduction of herbal decoctions and medicated oils into the colon through the rectum
- In Physical therapy** he was given-Strengthening exercises, Relaxed Passive Movements, Bridging, Trunk rolling, Wall squats & Therapeutic exercises.

Physical therapy and stretching exercises aim to improve movement difficulties & balancing difficulties. Physical therapy improves functionality of extremities and helps to reduce rigidity in affected limbs.

After first session of Therapy, we could see significant reversal of his symptoms, his gait improved, tendency to drag right leg decreased moderately, he could wear chappals/ footwear which was not possible earlier, clumsiness & abnormal movements in his Right hand decreased significantly, He could do fine movements with his hand, could hold objects in his hand. As he could hold scissors/ razor blades, he started doing his regular job of haircut & shaving in his hair salon which he had left for last few months & which was his only income source for his family.

He is still under our treatment for therapy sessions & we are closely monitoring his signs & symptoms to understand the nature & progression of disease while patient is under our unique treatment with complementary therapies.

Discussion

The cause of CBD is unknown. Scientists know that in some people with CBD, there is a large build-up of a protein called tau. Tau occurs normally in the brain, but we do not yet understand what causes it to build up in large amounts. Others may have a large build-up of amyloid plaques similar to those seen in people with Alzheimer's disease. As more and more proteins build up in the nerve cells, the cells lose their ability to function and eventually die. This causes affected parts of the brain to shrink.

CBD symptoms:

- difficulty controlling limbs on one side of the body.
- a clumsy or "useless" hand
- muscle stiffness
- feeling like the limb doesn't belong to you (an "alien" limb/ also known as ALS- alien Limb Syndrome)
- shaking (tremors), jerky movements and spasms (dystonia)
- problems with balance and co-ordination
- dementia
- visual disturbance
- slow, staggered, effortful speech
- difficulty in swallowing
- Trouble completing tasks like using utensils or buttoning a shirt.

- Difficulty coordinating fine movements (apraxia).

One limb is usually affected at first, before spreading to the rest of the body. The rate at which the symptoms progress varies widely from person to person.

The first sign of CBD is usually trouble with movement. The symptoms may begin in one hand, arm or leg. The limb may feel stiff and might shake. A person with CBD may experience a slight change in the feeling of the limb and may have trouble making the limb move, or the movement may be slower than usual.

Later in the disease, language problems may begin. People may have increased trouble finding the right word, naming objects and people or just getting the words out. Their reading skills might also get worse, and writing might become especially difficult if the movement symptoms affect the hands. Moving around might also get harder as time goes on. Symptoms that start on one limb or on one side of the body might move slowly to other limbs or to the other side of the body. In the late stages of CBD, people may have some personality changes. They might become obsessive or impulsive, or they might say inappropriate things. Some people may also develop memory problems, such as misplacing objects or repeating questions.

Diagnosis

A diagnosis of corticobasal degeneration (CBD) is based on clinical features, signs and symptoms of the patient. However, signs and symptoms could be due to other degenerative diseases such as progressive supranuclear palsy, Alzheimer's disease, Pick's disease and Creutzfeldt-Jakob disease (CJD). You may need an imaging test, such as a CT scan or MRI, to rule out these other conditions. Sometimes, these tests are performed every few months to look for abnormal changes in the brain. Researchers are looking at whether positron emission tomography (PET) and single-photon emission computerized tomography (SPECT) scans can uncover changes in the brain that are consistent with corticobasal degeneration. However, more research needs to be done in this area.

Differential Diagnosis

Predominant parkinsonian features might not be easy to distinguish from idiopathic PD and atypical parkinsonian syndromes (e.g., PSP and multiple system atrophy, MSA). Akinetic-rigid syndrome, early imbalance and poor response to dopaminergic treatment are typical symptoms for CBD, PSP, and MSA, in contrast with PD. PSP is clinically and pathologically related to CBD, but with differences in symmetrical versus asymmetrical parkinsonism, and in the pattern of rigidity, which tends to be more nuchal/axial in PSP and more limb-accentuated in CBD. Also, while the supranuclear gaze palsy can occur in both conditions, in CBD it tends to affect the horizontal more than the vertical gaze and the saccade latency (delay in initiating saccades) more than saccade velocity (the speed of saccades). In patients with cognitive presentation, the diagnosis depends on the specific case: pronounced visuospatial deficits related to dementia with Lewy bodies (DLB), language involvement to primary progressive aphasia (PPA), and prominent behavioral features to fronto-temporal dementia (FTD).

Treatment

There is currently no treatment that has been shown to stop CBD getting gradually worse. No disease-modifying treatment has been approved for CBD yet. However, many symptoms of CBD can be symptomatically treated. Parkinsonism in CBD can be treated with levodopa but only with poor and transient response. Dystonia in CBD can be successfully treated with botulinum toxin, which can temporarily improve the functionality of the affected limb.

Trihexyphenidyl is a muscarinic acetylcholine receptor antagonist, which can also be used to treat dystonia. Clonazepam is the first-choice treatment for myoclonus. Baclofen can be used to treat spasticity.

Outlook / Prognosis

CBD presents high morbidity and poor prognosis because of the lack of effective therapy at present. The symptoms of corticobasal degeneration (CBD) progress to serious complications, such as pneumonia, blood clots in the lungs, or sepsis, a life-threatening response to an infection. Difficulty in swallowing can cause choking, or inhaling food or liquid into the airways. This can lead to pneumonia, which can be life-threatening. Corticobasal degeneration complications ultimately lead to death. As a result of these complications, the average life expectancy for someone with CBD is around 6 to 8 years from the onset of symptoms. However, this is only an average and CBD is very variable.

As we could see our patient in an early stage of the disease, we treated him with the therapy which we are specialized in & which we have designed & developed over three decades of practice & experience in our center & could see a convincingly positive outcome in an early CBD case.

Conclusion

Considering the character of the CBD disorder and keeping in mind our knowledge about the prognosis of this disease, we do not know how the disease would progress in coming time in this patient, but we just wanted to highlight that we could intervene in earlier stages of the CBD with our Unique Treatment Method with multidisciplinary approach combining complementary therapies & physical therapy & with this we could see significant reversal in patient's symptoms. We would like to emphasize that with this approach we could improve the quality of life of the patient & could probably increase life expectancy in a CBD patient. Convincingly we would also like to add that this kind of Therapy will not only achieve physical wellbeing of patient but also socio-economic wellbeing of his family in a condition wherein patient himself is the sole earning member of his family.

Every medical science/ Therapy aims at curing the disease and restoring physical and mental well-being of the patient. When practiced together these multiple therapies combined with modern medical science could harmoniously complement each other & we could achieve far better outcome regarding patient's health.

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