Medical and Research Publications
International Open Access

Review Article **Journal of MAR Neurology and Psychology (Volume 6 Issue 4)**

Report of an Interesting and Progressive Case of the Disease

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Received Date: February 28, 2023

Published Date: April 01, 2023

Progressive Nuclear Palsy (P.S.P)

When the Cuivd19 epidemic became global, including in the country of Albania, we were faced with a large number of patients like all other countries in the world. My colleagues visited the patient, a 63-year-old man, who came in due to shaking of the chin and hands, along with fever.

He was observed as a corona patient. But as his problems got worse, his family admitted him to a private hospital. In that hospital, an MRI of the brain was performed and the doctors diagnosed brain hydrocephalus. There was a hospital with limited medical facilities, the doctors recommended that after the end of Corona in the country, he should visit a neurosurgeon and check the possibility of installing a Brain shunt. Corona lasted more than a year.

Then we were able to re-evaluate him medically. On axial views of the brain, MRI showed, although the middle ventricles are slightly dilated, but was not hydrocephalus. For this reason, neurosurgeons rejected the existence of hydrocephalus. They said:

All radiological views reject this issue. Also, the clinical symptoms and complaints of the disease and the new neurological examinations were not compatible with the diagnosis of hydrocephalus.

In hydrocephalus patients that I have seen so far, usually the symptoms: imbalance in walking were very obvious. The severity of the imbalance was so severe that they could not walk. They were patients who complained of progressive headache caused by a dramatic increase in C.S.F, nausea, vomiting. While the most important symptom and complaint of this patient was the tremor of his right hand and chin.

My colleagues and I re-evaluated the patient and during the neurological clinical examinations, we accepted that the patient's problems were not hydrocephalus, but Parkinson's.

With this diagnosis, we immediately started drug treatment. Tab Amantadine 100 mg twice, Tab Medopar 250 mg three times, a quarter of a tablet each time. With this initial treatment, after a while, although his symptoms decreased.

Three months later, the friends who lived with him raised two new issues:

- Memory disorders and dementia
- Lack of balance in keeping oneself even in normal traffic. They reported cases where the patient fell due to lack of balance control.

We did M.M.S.E test for the patient to evaluate his memory disorder. The test number was reported as 22. It means the beginning of memory disorders, which of course, considering his age, this number was not too low for us.

What was clear in his clinical examinations, was his inability to perform tasks and quick movements. We even encountered a lot of pauses in answering questions, like someone who has forgotten something.

Friends of the patient kept complaining about his falls. According clinical observations did not justify his falls.

Clinical examinations and muscle stiffness were confirmed along with early dyskinesia and tremor of the right hand and chin. Ophthalmology examinations were normal. His hearing was perfect and he walked like a Parkinson's patient.

After two years, we noticed that his condition has seriously changed. He was treated with anti-Parkinson drugs. But his speech had changed and he was unable to speak. He wanted to say a word or a name, he looked at our faces and wanted to say a word, but he couldn't and finally gave up.

We also saw that, apart from memory problems, concentration, calculation, judgment was absolutely disturbed. He was different from a Parkinsonian patient in his normal walking. It was very strange for me that in a short period of time, apart from the accompanying dementia disorders, he has also found some psychological functions. I have known him and his family for more than four decades. And this development of the disease was very strange and unexpected for me. We noticed that for no apparent reason, the left side of his upper limb was very weak and his muscle strength was reduced. Although his muscle reflexes were normal.

Along with these symptoms, his inability to relate to those around him and even his closest friends, led us to a new diagnosis, which was more than a typical Parkinson's. In my specialty, I observed many parkinsonian patients, but his condition was beyond the classical parkinsonian patients.

My colleagues and I focused on a new term, Progressive Supranuclear Palsy (P.S.P), which in some books is also called Richardson-Steele-Olszewski syndrome. In this syndrome, the patient faces focal dystonia along with dementia disorders, which usually face life threats such as pneumonia and swallowing disorders. In this patient, in contrast to Parkinson's patients, who respond even partially to drug treatments, while in Progressive Supranuclear Palsy (P.S.P), not only do they not respond adequately to anti-Parkinson drugs, but the patient's condition increases rapidly. We had enough reasons to confirm the above-mentioned progressive paralysis disease, including:

- Lack of acceptable response to common anti-Parkinson's treatments, despite increasing the dosage of drugs and their variety
- Significant changes in sensory and motor disorders in a progressive manner
- Swallowing disorder and even difficulty in eating solid foods routinely
- Cognitive disorders
- Sleep disorders, he could not sleep for several days and nights, even for a short time, accompanied by symptoms of depression and unwillingness to open his eyes and establish a relationship with his family and friends. Despite the use of treatments such as melatonin and citalopram, his sleep problems were not resolved, so we had to start him with Tab Clonotril 2mg t, which reduced his problems
- Breathing Abnormality and feeling of respiratory distress. (He was a smoker thirty years ago and but had quit smoking in the past three decades, and normal chest radiographs did not show significant chest disease.)

The first problem that made me realize that the patient's problems are not only Parkinson's, but may also be other problems, was his imbalance in walking. I also found a bewilderment on his face, along with a long silence to answer questions. This silence was so long that every doctor was surprised at the first encounter. Unnecessary laughter, loud voice when talking, emphasis on his words, i.e. repeating them, were strange for me too. I also encountered other symptoms in this patient, such as:

- Slow eye movements,
- Difficulty in voluntarily moving the gaze vertically (that is, down and/or up),
- Difficulty in controlling the eyelids.
- Tendency to move the head to look in different directions.
- Involuntary closing of the eyes.
- Long blinking,
- Difficulty opening eyes was so evident at times that, I assumed, he was asleep, and the main problem was inability to maintain eye contact during conversation.

I was also faced with other signs and symptoms with this patient, including:

- Indifference to people far away from him and even a regular relationship with family and old friends.

- Change in judgment, insight and problem solving
- Difficulty in finding common and not necessarily new words
- Loss of interest in normally pleasurable activities.
- Increased irritability and forgetfulness,
- Laughing or crying suddenly or showing angry outbursts for no apparent reason.
- Personality change
- Slowness of thinking
- Slow, slurred or monotonous speech.
- Mask-like facial expressions, especially when some of his friends had seen him after many years, they told me about this issue.

In this patient, who had a mixture of Parkinson's symptoms and unlike other Parkinson's patients who have been under my care for years, I saw that he had differences with Parkinson's. Because he had many symptoms including stiffness, movement problems, clumsiness, bradykinesia and muscle stiffness. However, the rapid progress of this problem, in contrast to the slowness of the process in Parkinson's patients, was something to think about. I found some differences in the clinical symptoms of this patient with Parkinson's patients:

- He stands exceptionally straight or sometimes tilts his head back (or tends to fall back). This is called "axial rigidity", but people with Parkinson's disease usually lean forward.
- Speech and swallowing problems in this patient were more severe and attracted a lot of attention, so that if someone did not know him, he would remember the lesions left from a cerebral infarction.

It is interesting to note that in this patient, unlike Parkinson's patients, after two years, hand tremors were no longer very evident, while in Parkinson's patients, hand tremors are one of their primary symptoms.

- I understood very well that in this patient, unlike Parkinson's, Leva Dopa or Seferol drugs did not work, and it bothered me that even the primary symptoms of the so-called Parkinson's were not controlled in this patient.

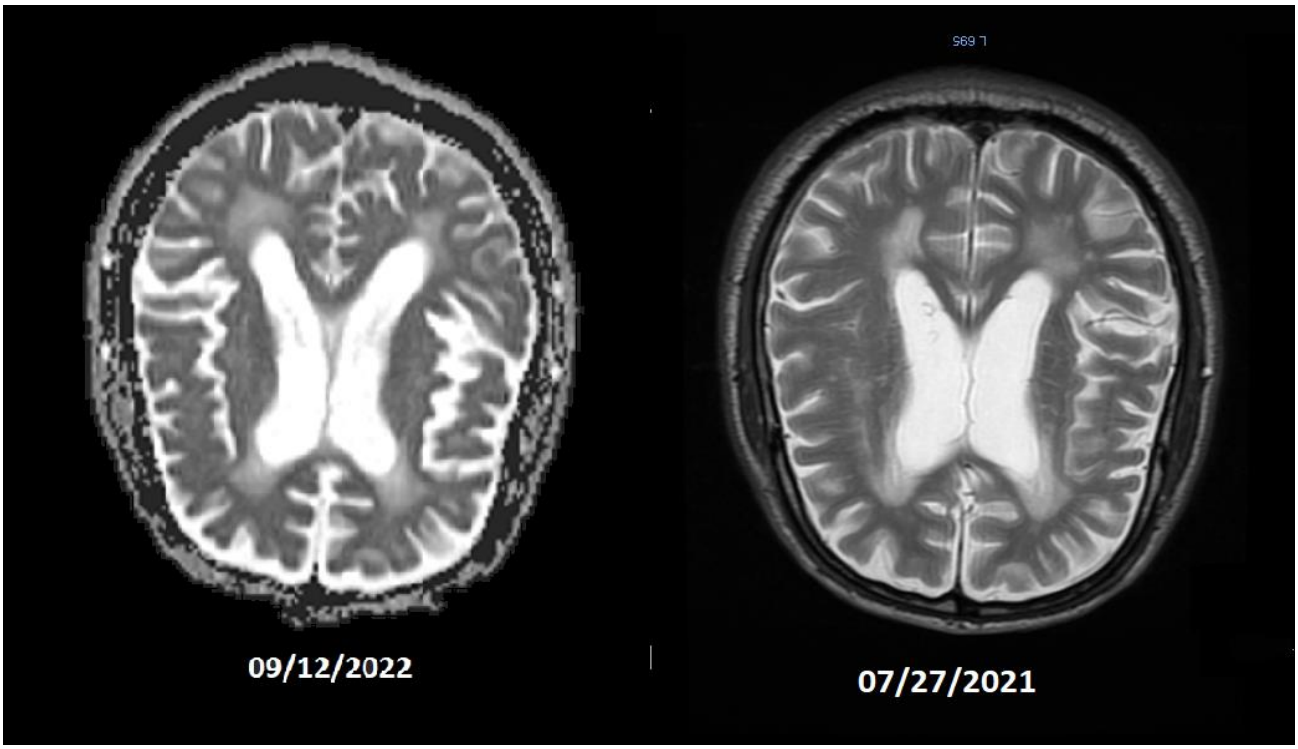
Unfortunately, I did not have the possibility to perform blood tests for Tau ramen protein accumulation at my place of residence, and the patient's financial condition did not allow us to follow it abroad, and the only diagnoses by me and my colleagues, were based on clinical symptoms, clinical examinations, and the patient's process. This the report of patient's Brain MRI:

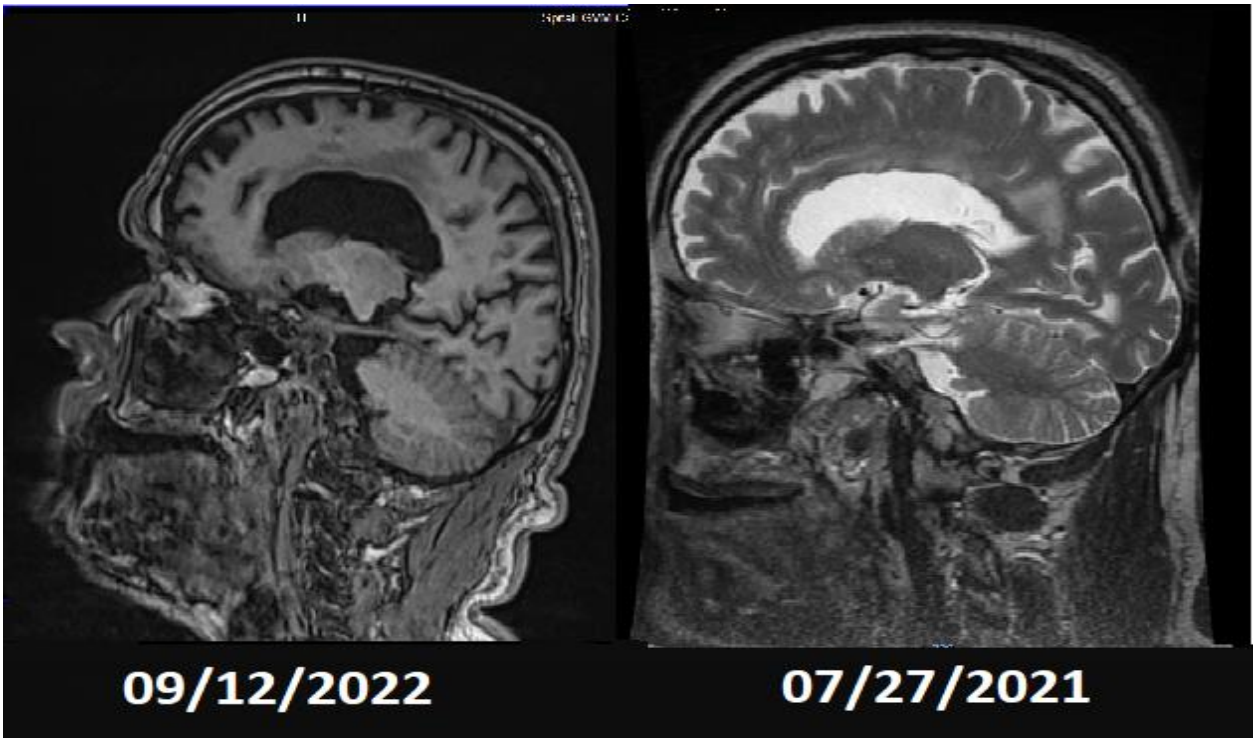
…Cerebellar tonsils superiorly foramen magnum.

- Cerebellar hemispheres, Vermis and Truncus cerebra in normal, without lesions.
- Free basal cisterns.
- Aqueduct with normal caliber, without trans ductal void flow.
- With moderately active hydrocephalus.
- Hyper signals in periventricular T2-dependent sequences in favor of active hydrocephalus
- Cortex with sub atrophy mainly insula-temporal.
- No hemosiderin signal in T2 * GRE. No intra-extra axial collections. Pituitary and orbits in normal. Paranasal sinuses and free mastoid cells. Nasopharynx is not the norm.
- Intracerebral arteries with normal trajectory and caliber. No pathological post-contrast contrasts. Dural sinuses with normal filling.

Conclusion

Moderate active hydrocephalus, probable malabsorption





And the new Brain M.R.I Report:

Cerebral and cerebellar cortical sub atrophy. Free basal tanks. Free bilateral cerebellar Ponto kendet. Internal auditory channels and semi-horizontal channels with normal aspects. Ventricle with normal size in the median line. Giotic lesions of the bilateral periventricular substance (they confluent with one of. No retraction them focus on diffusion and no hemosiderin deposition in hemosensitive sequences. No posting of media structures. No pathology of orbits. Pituitary glandular with signal and dimensions in the norm. There is no any about Hydrocephalus!

A serious incident:

At the end of November 2022, a person with a patient came to me in a worrying manner and reported that: he did not wake up to eat breakfast and lunch. He said:

No matter what I tried to wake him up, it didn't work.

I quickly rushed to the patient's resting place and when I did the initial examination, I realized that he had gone into a coma. And Glasgow's score was below 8. He had no verbal reactions and only opened his eyes for a short moment and closed them again due to deep stimulation. His blood pressure was below 100 over 70 and his pulse was 65 and his breathing was calm. I immediately called the ambulance and sent him to the emergency department of Nene Teresa Hospital. Due to his general

condition, he was admitted and monitored in the neurology department. In the investigations, it was found that the patient had metabolic problems caused by hyponatremia, which caused his coma, which was solved with a nasogastric tube along with serum therapy. After five days, he opened his eyes and looked at the people around him. He asked why am I here?

... After being discharged from the hospital, I encountered numerous sores in the area of his right elbow and right heel, which were evidently the effects of five days of coma. I asked the people who take care of this patient Do not give him any sleeping pills considering that he is not active at all and rests on the bed all the time. One of the factors that can lead the patient to coma is the use of sleeping pills. New food recipes for I have given him and have his people do physiotherapy for him right there. I have asked them to stay with the patient and try to make an emotional connection with him and encourage him to talk and...

Some questions:

Question 1: Is this his situation over?

Answer: Not

Question 2: Is it possible for him to confront with this same board or even violent boards?

Answer: Yes, is it possible

Question 3: What is the half-life of these patients with this diagnosis?

Answer: Maximum one to five years

Question 4: What can we do for the patient with P.S.P?

Answer: No radical treatment has been defined, unfortunately Anti PD treatments do not have much effect on patients.

Question 5: What causes PSP?

Answer5: The exact cause of PSP is unknown, but research suggests that the disease involves the gradual deterioration of brain cells in a few specific areas of the brain, mainly in the brain stem. Death

of brain cells in one of these areas, the substantia nigra, is partly the cause of the movement symptoms shared by P.S.P and Parkinson's.

P.S.P is characterized by the accumulation of abnormal deposits of Tau protein in the nerve cells of the brain. These deposits cause cell dysfunction and death, which stops the flow of information to other nerve cells. The accumulation of Tau Places P.S.P in a group of disorders called Tauopathy, which includes Alzheimer's disease, cortico - basal degeneration, and some forms of frontotemporal degeneration.

P.S.P is usually sporadic, meaning it occurs rarely and without a known cause. In very few cases, the disease is caused by a mutation in the MAPT gene. This mutation gives the nerve cell incorrect instructions for making Tau. Genetic factors have not been involved in most people.

Several theories suggest that P.S.P may be caused by:

- Abnormal accumulation of Tau protein in a cell causes an attached cell, which then spreads through the nervous system.
- An unconventional infectious agent that takes years or decades to start producing visible effects (as seen in disorders such as Creutzfeldt-Jakob disease).
- Random genetic mutations—the kind that happen all the time in people—occur in specific cells or specific genes, in a specific combination that harms those cells
- Exposure to some unknown chemical in food, air or water, which slowly damages some vulnerable areas of the brain, similar to a neurological disorder found on the Pacific island of Guam and several nearby islands.
- Cell damage caused by free radicals, which are reactive molecules that are continuously produced by all cells during normal metabolism. Although the body has internal mechanisms to clear free radicals from the system, scientists suspect that - under certain conditions - free radicals can react with other molecules and damage them.

Question 6: Is there a cure?

Answer: There is currently no effective treatment for P.S.P, and symptoms usually do not respond to medications.

- Parkinson's disease drugs, such as ropinirole, rarely provide additional benefits. In some people, other antiparkinsonian drugs, such as levodopa, can treat the slowness, stiffness, and balance problems associated with P.S.P, but the effect is usually minimal and short-lived.
- Botulinum toxin, which can be injected into the muscles around the eyes, can treat excessive closing of the eyes.
- Some antidepressants may have benefits beyond treating depression, such as pain relief and reduced drooling.
- Non-pharmacological treatment for P.S.P can take many forms.
- Heavy walking aids can help people avoid falling backwards.
- People with P.S.P are sometimes prescribed bifocals or special glasses called prisms to help them look down.
- Exercise under the supervision of a health care professional can keep the joints straight, but formal physical therapy has no proven benefit in P.S.P.
- A gastrostomy (a surgical procedure that involves placing a tube through the skin of the abdomen into the stomach for feeding purposes) may be necessary if there is difficulty swallowing or a definite risk of severe choking.
- D.B.S. deep brain stimulation—which uses surgically implanted electrodes and a pacemaker-like medical device to deliver electrical stimulation to specific areas in the brain to block the signals that cause the motor symptoms of several neurological disorders—and other surgical procedures commonly used in people It is used in patients with Parkinson's disease. Effectiveness has not been proven in P.S.P. While they have been reported to be very useful in Parkinson's patients, especially in those who do not respond to drug treatment.

Question7: What is the prognosis?

Answer: The disease gradually worsens, and people become severely disabled within three to five years of onset. Affected people are prone to serious complications such as pneumonia, suffocation, head trauma and fractures. The most common cause of death is pneumonia. With good attention to medical and nutritional needs, it is possible for people with PSP to live a decade or more after the first symptoms of the disease appear.

Question8: What research is being done?

Answer: The National Institute of Neurological Disorders and Stroke (NINDS), a component of the National Institutes of Health, is the primary provider of research on the brain and nervous system. NIH is the world's leading funder of biomedical research.

- P.S.P is one of the diseases being studied as part of the NINDS Parkinson's Disease Biomarkers Program. This major NINDS initiative aims to discover ways to Identifying people at risk of Parkinson's disease and related disorders and tracking the progress of these diseases. NINDS also supports clinical research studies to develop brain imaging that may allow for earlier and more accurate diagnosis of P.S.P. Genetic studies of P.S.P may identify underlying genetic causes. Previous studies have linked regions of chromosomes containing multiple genes, including the Tau protein (MAPT) gene, to P.S.P.
- Researchers hope to identify specific disease-causing mutations and are also studying how genetics and environment interact to contribute to disease susceptibility.
- Scientists hope to understand the Tau-related mechanisms that lead to P.S.P and its symptoms. Tau can exist in many forms or combinations, and research has shown that some of these combinations are harmful, leading to the formation of toxic aggregates and disruption of normal molecular signaling pathways within.

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