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Stiff Person Syndrome- the forgotten cause of Depression

Dr. Anshul Arora*

Corresponding Author: Dr. Anshul Arora, Board-certified Neurologist based in Germany

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I am honored and excited to present an exciting case during my rotation in the Department of geriatric Psychiatry.

Before getting into the case, I would like to introduce myself. I am Dr. Anshul Arora, a Board-certified Neurologist based in Germany.

During my last rotation in Psychiatry, I encountered several interesting patients, one of them on whom this case report is based on.

Case Report

A Patient aged 50 was admitted because of progressive cognitive Impairment and depression. Over the years there was a decline of movement. The mood war reported depressed. The Patient would just be lying in bed without doing anything during the day. The appetite was decreased as well as a result he lost a sufficient amount of weight.

Apart from that there was cognitive decline with episodes of aggression during the evening. From the past medical history chronic consume of Alcohol there was noted but was never proved. On Examination the Patient was afebrile. He was alert to Person but not oriented to Place, Situation and time.

Furthermore, the mood looked depressed. The Affect was reduced. The drive decreased as well. In the minimental Test the patient achieved 10 Points out of possible 30 points, which correlates with a severe care upon neurological Examination the cranial nerves were intact. Hyperreflexia was noted. Signs after Babinksi, Hoffmann, Chaddock as well Trömer were negative. The Gait displayed a reduced step length. Apart from that spasm and random myoclonic jerks were noted which worsened after tactile stimuli.

To rule out a cerebral Pathology a computer tomography of the head was done which yielded a slight general atrophy. Indications for an acute cerebral ischemia or intracranial bleeding could not be detected.

The laboratory analysis didn't show any pathological changes. To exclude an infection of the central nervous system and to the detect the markers for Dementia as well to exclude an encephalitis an analysis of the cerebrospinal fluid was done which ruled out an infection.

The further Evaluation yielded a positive Titer antibody against GAD. To rule out a paraneoplastic syndrome a further evaluation of the thorax and abdomen with the aid of computer tomography was conducted which showed no indication of malignancy. In view of the clinical and paraclinical findings the suspicion of Stiff Person Syndrome was raised.

We initiated a treatment consisting Methylprednisolone intravenous. Because of the cognitive impairment, the patient kept removing the intravenous line so that it had to be changed to Prednisolone per os. Addition to the patient received Lorazepam and Baclofen.

After duration of 2 Weeks there were clinical improvements. The myoclonic jerks were reduced. The affect was much better. The hyposmia was reversible. The Patient was transferred to neurological ward for further diagnostics.

Definition of Stiff Person Syndrome

Stiff person Syndrome also known as Stiff Man Syndrome is a rare neurological disorder of unclear cause characterized by progressive rigidity and stiffness. The Stiffness primarily affects the truncal muscles and is superimposed by spasms, resulting in postural deformities.

Pathogenesis:

In Reference to literauture, Stiff Person Man Syndrome is said to be caused by an underlying autoimmune disease. The autoimmune response causes a cascade of reaction resulting in impairment of GABA mediated interneuronal activity.

There have several antibodies associated with Siff Person Man Syndrome, mostly glutamic acidid decarboxylase (GAD), Amphysin, Glycin R, Dipeptidyl Peptidase Like Protein (DPPX,Gamma aminobutyric acid type A receptor. Understanding Stiff Person Man Syndrome is all about understanding the pathophysiology.

After establishing the correlation between the antibodies and GABA mediated interneuronal activity, its important to understand what happens between them. [2],[3],[4],[5],[6]

GAD Antibody, the mostly noted antibody associated with Stiff Person Man Syndrome is the rate limiting enzyme in the synthesis of γ-aminobutyric acid (GABA). We could go into details of all the antibodies, but the end result remains the same, that every enzyme listed as antibody is used in the synthesis of GABA. GABA in turn inhibits the stimulation of neuros. In simple words, an impairment of GABA caused by the antibodies causes constant firing of neurons.

Thus, in SPS patients motor neuron unit neurons fire involuntarily in a way that resembles a normal contraction. Motor unit potentials fire while the patient is at rest, particularly in the stiff muscles. The excessive firing of motor neurons may be caused by malfunctions in spinal and supra-segmental inhibitory networks that utilize GABA. Hence, involuntary actions show up as voluntary on EMG scans; even when the patient tries to relax, there are agonist and antagonist contractions.

Diagnosis

Personally, I feel the most important diagnostic feature are the clinical findings which can be as listed below.

The diagnosis should be carried by the following;

1. Clinical findings
2. Analysis of cerebrospinal Fluid
3. EMG
4. Exclusion of paraneoplastic cause

Clinical Findings

Hyperlordosis

Hyposmie

Reduced movement

Stiffness

Rigidity

Myoclonic jerks stimulated by tactile stimuli

Hyperreflexia

Positive Babinski

Cerebrospinal Fluid

Analysis of cerebrospinal Fluid is important to obtain the antibodies as well to exclude other conditions. Apart from an increased Titier against the above mentioned antibodies, oligoclonal bands (30 %) and an increase of Protein can detected.

EMG

EMG can provide a useful clue to the diagnosis by demonstrating characteristic continuous motor unit activity in agonist and antagonist

Exlusion of paraneolplastic cause

There have been cases associated with lung cancer, ovarian cancer as well thymom. In order to rule them out its adviseable to conduct a ct scan of the thorax and abdomen.

Treatment

By understanding the pathophysiology of the disease, we understand that there are two approached towards the treatment

1. Enhance the activity of GABA

2. To suppress the autoimmune response

Enhance the activity of GABA?

By aiding the GABA mediated pathway, we decrease the interneuronal firing which in return decreases the stiffness. Medications which help with are as

Benzodiazepines (i. e Diazepam or Lorazepam)

The benzodiazepine binding site is in a specific pocket at the pairing (intersection) of the α and γ subunits. Within the α subunit of isoforms 1, 2, 3, and 5 resides a histidine residue (H101, H101, H126, and H105, respectively) that possesses a high affinity for BZDs. x Isoforms 4 and 6 of the α subunit contain an arginine residue and do not have an affinity for BZDs.x BZDs bind to the pocket created by the α and γ subunits and induce a conformational change in the GABA-A receptor, allowing GABA to bind. BZDs bind to the pocket created by α and γ subunits and induce a conformational change in the GABA-A receptor. In simple words, Benzodiazpines enhance the function of GABA which in turns causes the inhibitory effect.

Baclofen
Baclofen is a gamma-aminobutyric acid (GABA) agonist used as a skeletal muscle relaxant.

Immunsupression

Steroids

Rituximab

Plasmapheresis

Treatment

Discussion & Conclusion

Having a patient of psychiatric disorder can be difficult. Not only because of the difficulty of the patient´s condition but because of the difficulties coming along with it. Usually once a patient has been diagnosed with a psychiatric disorder it's difficult to overlook it. In other field there are Diagnosis of Exclusions which are concluded after a thorough work up where as patients with psychiatric clinical manifestations are concluded instantly without a in depth work up. Even when a patient is seeking justice, he/ she is innocent until proven otherwise but in psychiatric patients its mostly psychiatric disorder until proven otherwise.

Similar situation occurred with our patient. Because of his commodities he was subjected first as a patient with dementia with depression as a result of his presumed alcohol misuse, where as he actually had an autoimmune encephalitis. If an early detection would have helped his cognitive decline is a subject to imagination.

Over the years I have encountered several patients who actually had an underlying neurological disorder but were overlooked by their psychiatric manifestations.

The take home message is that a diagnosis of psychiatric disorder at any age should be a diagnosis of exclusion and not otherwise. Colleagues from different field need to be comfortable with patients with psychiatric manifestations rather than directing them to psychiatric ward. At the end we are solely there for the patient and should the patient should not be stigmatized for disorder he develops.

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