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Case report ***Open Access***

Stiff person syndrome misdiagnoised as oxaliplatin induced neurotoxicity

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ABSTRACT

Stiff person syndrome is characterized by muscle rigidity that waxes and wanes along with concurrent spasms. The closest related disease to Stiff person syndrome is Tetanus because both conditions affect peripheral inhibition via central mechanisms and both conditions inhibit central gamma-amino butyric acid (GABA) systems. We herein report the case of stiff-person syndrome in a 65-year-old man. He experienced pain with bleeding from the rectum. The tests included screening tests to detect the presence of amphiphysin antibodies and electromyography. His clinical examination was suggestive of generalized muscle involvement with laryngeal involvement. He was evaluated with a CT scan abdomen, which was suggestive of Liver Limited cancer and rectal examination suggested ulcero-proliferative growth of around 4 cm from the anal verge. Colonoscopy was suggestive of circumferential growth with multiple session polyps, which was to document the metastatic nature of the disease. FNAC was done from the government which was conclusive of metastasis. He was initiated on Capecitabine[7] and Oxaliplatin protocol. He underwent NCV studies which came out to be mild to the moderate sensory deficit. During the hospital stay, he was treated with high dose Methyl Prednisolone 1gm for 5 days and sequentially with immunoglobulin 2gm per day for 5 days, concomitantly Benzodiazepam was given 10mg three times a day for 3 days but later withdrawn as the patient started experiencing dizziness. He achieved no clinical benefit in neurological status. Eventually developed aspiration pneumonia and succumbed to death after one month of diagnosis of SPS.

Keywords: Stiff-person syndrome, Autoimmune Disorder, Methyl Prednisolone, Bevacizumab.

INTRODUCTION

Stiff person syndrome [1] is a rare acquired neurological disorder characterized by progressive muscle stiffness and repeated episodes of painful muscle spasms. The muscular rigidity often grows worse and then improvises and usually occurs with muscle spasms. Spasms occur randomly or a variety of events acts as triggers include a sudden noise or light physical contact. The severity and progression of SPS vary from one person to another. The exact cause of SPS [2] is unknown; it is an autoimmune disorder and sometimes can occur along with other autoimmune disorders. The exact prevalence [3] and incidence of SPS are unknown, although one estimates place the incidence [4, 5] at about 1 in 1,00,000 people in the general population. SPS usually becomes prevalent sometime between 30 - 60 years of age.

CASE REPORT

A 65-year-old male presented with complaints of pain and PR bleeding in January 2019. He was evaluated with a CT scan abdomen, which became one of the factors in the suggestion of Liver

Limited cancer and rectal examination suggested ulcero-proliferative growth of around 4 cm from the anal verge [6]. Colonoscopy was suggestive of circumferential growth with multiple session polyps, which was to document the metastatic nature of the disease. FNAC was done from the government which was conclusive of metastasis. He was initiated on Capecitabine [7] and Oxaliplatin protocol. At the time of the presentation, his performance score was ECOG 1. However, the patient discontinued chemotherapy given severe loose motion after the first dose of capecitabine and oxaliplatin regimen. He presented again in March 2019 after a gap of 2 months with worsening of symptoms of weakness and bleeding from the rectum. Revaluation with CT scan abdomen again showed suggestive of Liver Limited disease. His mutational status of KRAS, BRAF was not done because of financial constraints. Bevacizumab was not added given financial constraints. He was initiated with a dose reduction of the CAPOX regimen. However, the patient again defaulted for 2 months because of personal issues. He did not develop severe enteropathy this time. He presented again in May 2019 and was re-initiated with CAPOX regimen on 2 weekly bases, which he completed to 4 cycles till August 2019. From July 2019 patient started to develop hypoalbuminemia and generalized weakness. In August 2019 patient developed generalized weakness, on clinical examination, he was found to have severe tightening around the body. Based on clinical examination there was no neurological weakness attributable to the sensory neural deficit or due to cerebellar involvement. MRI brain was negative for any brain metastasis or leptomeningeal involvement. CSF analysis showed a mild rise in CSF protein. CSF was also sent for anti GAD [6] antibody analysis which came out positive. His clinical examination was suggestive of generalized muscle involvement with laryngeal involvement. He was planned for Tracheostomy but as a familial consensus tracheostomy was withheld. He underwent NCV studies which came out to be mild to the moderate sensory deficit [8].

TREATMENT

During the hospital stay, he was treated with high dose Methyl Prednisolone 1gm for 5 days and sequentially with immunoglobulin 2gm per day for 5 days, concomitantly Benzodiazepam was given 10mg three times a day for 3 days but later withdrawn as the patient started experiencing dizziness. He achieved no clinical benefit in neurological status. Eventually developed aspiration pneumonia and succumbed to death after one month of diagnosis of SPS.

DISCUSSION

A diagnosis of SPS is based upon identification of characteristic symptoms, detailed patient history, and thorough clinical evaluation. The tests include screening tests to detect the presence of amphiphysin antibodies and electromyography. An EMG can prove continuous muscle motor unit firing in stiff muscles which is characteristic of SPS.

In this case, the patient's stiffness of muscle was misdiagnosed as distal dysesthesia which is observed in oxaliplatin-induced neurotoxicity. His condition is also unique as it didn't show any novel identification factors for the diagnosis of stiff person syndrome. This case report stresses the fact that the physicians should be open to the hypothesis for uncommon diseases even when a minuscule of doubt arises thereby giving early management of the disease.

The main aim of SPS is to give symptomatic relief and improving the quality of life of the patient. Due to its rarity, there are limitations in the quality of treatment options [8]. Over the years, treatment modalities for SPS included with Benzodiazepines and Baclofen as the first line of drugs followed by IVIG, plasmapheresis, immune modulators, and Rituximab. IVIG and plasmapheresis are either used alone or in combination in refractory cases. Corticosteroids are used as monotherapy or as a combination with other drugs for SPS. However, their efficacy is not determined by any clinical trial yet. In this case, the patient was treated with a high dose of Methyl Prednisolone, immunoglobulin and Benzodiazepam.

In a para-neoplastic variant of SPS the stiffness localizes to the arms and legs, makes up to only 5% of SPS cases. Generally, classical SPS patients respond well to treatment, but in about 10% of cases, sudden deaths occur due to autonomic dysfunction. Repeated spasms or sudden withdrawal of medicine may lead to autonomic dysfunction and may lead to sudden death.

ABBREVIATIONS

SPS- Stiff Person Syndrome, CT- Computerized Tomography, Bleeding PR- Rectal Bleeding, FNAC- Fine Needle Aspiration Cytology, ECOG- Eastern Cooperative Oncology Group Performance Test, CAPOX- Capecitabine and Oxaliplatin Regimen, KRAS, BRAF- Tumor Driven Genes, MRI- Magnetic Resonance Imaging, CSF- Cerebrospinal Fluid, GAD antibodies- Diabetes Associated Antibodies, NCV- Nerve Conduction Study, EMG- Electromyography, IVIG- Intravenous Immunoglobulin.

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