

Immune Checkpoint Inhibitor Causes Autoimmune Effect Resulting in Stiff-Person Syndrome: A Case Report

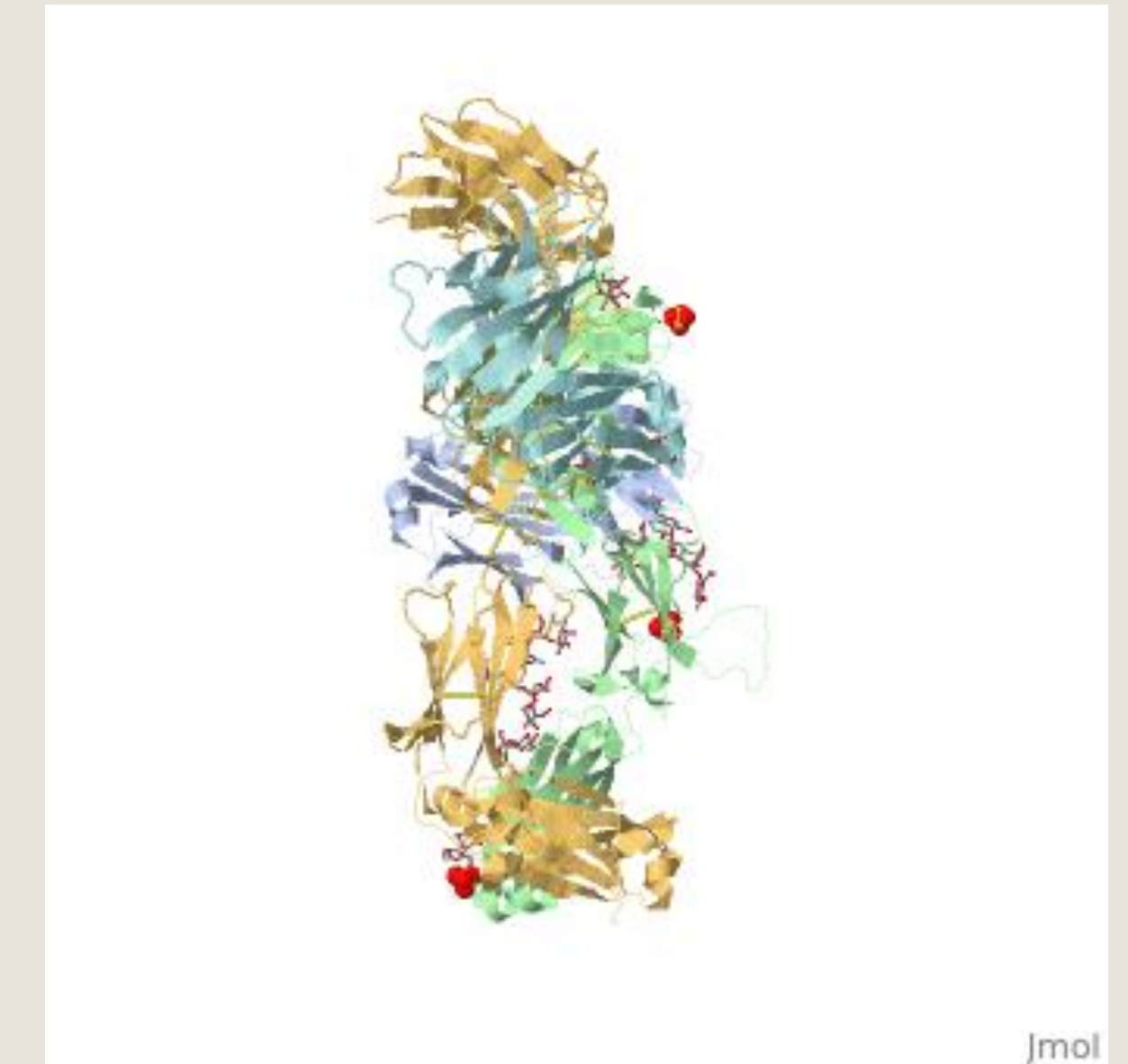
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Case Description: This patient is a 71 year old female, previously independent and living alone, with a history of stage IV lung cancer. She was started on a different chemotherapy regimen including Premetrexed, Carboplatin, and Pembrolizumab due to progression of her disease. After two cycles, she experienced mid to low back pain which progressed to lower extremity “weakness”, muscle spasms, and falls over two weeks. She was admitted to the hospital for further evaluation. Physical examination and EMG was consistent with Stiff-Person Syndrome, likely due to Pembrolizumab.

Discussion: Stiff-Person Syndrome is characterized by muscle spasms and rigidity that causes a functional decline. Usually the axial skeleton (abdominal muscles and thoracolumbar paraspinal muscles) is involved initially, and it progresses to the extremities later causing a functional decline in ADLs and ambulation. This disease process can be either autoimmune or paraneoplastic in nature. Diagnosis can include work up with physical examination, EMG findings, and lab work up with Anti-GAD antibodies in the serum or CSF. Current treatment recommendations include high dose steroids, GABAergic medications to reduce muscle spasm and spasticity, plasmapheresis and IVIG that can result in improved function and ambulation.

Use of Pembrolizumab can be associated with an autoimmune effect causing Stiff Person Syndrome



Conclusion: Even though it is rare, there have been case reports that show immune checkpoint inhibitors, like Pembrolizumab, can cause immune-related neurological symptoms such as SPS. This case reveals the importance of taking a thorough medical history and reviewing recent medication changes and/or additions in assessing for new progressive functional decline. SPS should be on the differential in the setting of recently initiated immune checkpoint inhibitors and noted axial stiffness that worsens to the extremities. Appropriate identification and treatment can result in improved function and ambulation.

Outcome: The patient was started on high dose steroids as well as Diazepam and demonstrated significant improvement in her rigidity and pain symptoms. However, her functional status did not significantly improve despite the above therapies and an acute rehab admission due to the development of significant cognitive deficits. She ultimately transitioned to skilled nursing for further rehabilitation and care.