

A Case Report of Paraneoplastic Stiff Person SyndromeHyuntae Kim^{1*}, Kyeong Eun Uhm¹, Jongmin Lee^{1†}Konkuk University Medical Center, Department of Rehabilitation Medicine¹**Introduction**

Stiff person syndrome, previously known as stiff man syndrome, is a progressive disorder of the central nervous system causing muscle stiffness and spasms. This uncommon entity gradually hinders one's ambulatory function commonly leading to a disability. Its prevalence is estimated at approximately 1-2 cases per million, with only a small minority of those patients being diagnosed with the paraneoplastic variant type. Herein, we report a very rare case of paraneoplastic stiff person syndrome, secondary to sigmoid colon cancer.

Case Report

A 59-year-old male was presented to the department of rehabilitation with axial stiffness and truncal lurching to the right side when ambulating. These symptoms began 4 months prior to the hospital visit and had increasingly worsened as what was initially stiffness involving upper back had progressively involved lower back and proximal lower extremities. An initial physical examination revealed hyperreflexia in the upper and lower extremities with spasticity at right knee extensor muscles while ankle clonus was bilaterally equivocal. There was no previous history of neurological disorder or other relevant family history, and a electrophysiologic study exhibited no evidence of peripheral neuropathy or radiculopathy, while a lumbosacral MRI showed only disc bulging at L3-4-5-S1 levels. The patient came back two months later when there was severe limitation on spinal extension with knee extensor spasticity also being found on the left side as well. With a likely diagnosis of stiff person syndrome, diazepam was prescribed along with serum anti-GAD Ab, which later came back negative. Upon admission to the department of neurology, the patient underwent additional laboratory test of tumor markers. TFT and other tumor makers turned out to be within normal limits, but elevated CEA at 17.64. For further evaluation, both EGD and colonoscopy were performed and found a 5 cm mass was found at distal colon, which was confirmed to be sigmoid colon cancer on biopsy. Subsequently the patient underwent a surgical resection followed by chemotherapy. With continued treatment with diazepam and addition of clobazam, axial muscle stiffness has gradually improved to the point where patient was able to perform lumbar extension and truncal lurching is minimally evident.

Conclusion

Paraneoplastic variant of stiff person syndrome is an extremely rare and interesting disorder that presents as a progressive truncal muscle stiffness and difficulty in ambulation. Though its rarity, it should be kept in mind when making a differential diagnosis for progressive axial muscle stiffness with impaired ambulation. At the same time, consideration of the paraneoplastic variant with concurrent features of an underlying neoplasm carries a significant diagnostic value in proper evaluation and

management of such patients, as complete resolution may not occur in the paraneoplastic variant unless the cancer is properly managed.



Fig 1. Photographs of a gait sequence with truncal lurching to the right side