

Subacute cerebellar ataxia with Anti-SOX1 antibodies and small cell lung cancer

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Introduction: Anti-SOX1 antibody (Ab)-associated paraneoplastic cerebellar degeneration (PCD) is rare immune-mediated neurological disorder. More commonly anti-SOX1 Abs have been linked with other paraneoplastic syndromes such as Lambert-Eaton myasthenic syndrome and sensorimotor polyneuropathy. Here we report a case of Anti-SOX1 Ab-associated PCD presenting as subacute cerebellar ataxia which showed a rapid and dramatic response to chemotherapy.

Case report: A 72-years old man first complained a vomiting, poor oral intake and non-vertigo dizziness. Five days after symptom onset, by then the symptoms had further progressed including dysarthria and the inability to independently walk or stand-up. He had suffered ischemic cerebral stroke 10 years ago but recovered without sequelae. Physical examination revealed dysmetria and severe limb, and truncal ataxia. Brain magnetic resonance imaging (MRI) shown no brain metastases, and no cerebellar or limbic alterations. Brain positron emission tomography computed tomography (PET-CT) showed mildly hypometabolic areas in the cerebella. Cerebrospinal fluid (CSF) revealed pleocytosis of white blood cell 75 cell/mm^3 and absence of hyperproteinorrachia. No atypical cells were evidenced in cytology in the CSF. The anti-SOX1 Abs were detected in the serum. Autoimmune synaptic encephalitis Abs and other paraneoplastic auto-Abs were negative. The chest CT showed multiple lymph nodes enlargement at mediastinum, right upper neck and left sub-clavicular area. Therefore, endobronchial ultrasound guided biopsy was conducted, and pathologic report was small cell lung cancer. We decided to try systemic chemotherapy without steroid pulse therapy. After only 2 cycles of chemotherapy, cerebellar ataxia was almost fully recovered and mediastinal lymph nodes showed decrease markedly in size on follow up imaging.

Discussion: Anti-SOX1 Ab-associated PCD is rare but devastating disease. Our case demonstrates that early detection of anti-SOX1 Ab, diagnosis of underlying neoplasm, and prompt initiation of chemotherapy are essential to achieve a better outcome.

