Cutaneous T-Cell Lymphoma and Paraneoplastic Syndromes

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Abstract

We describe the case of a patient with cerebellar ataxia resulting from Lambert Eaton Myasthenic Syndrome in the setting of a recent diagnosis of cutaneous T-cell lymphoma. Cutaneous T-cell lymphoma is not well-associated with any paraneoplastic syndromes. Though both the malignancy and paraneoplastic syndrome are individually rare phenomena, dermatologists need to evaluate systemic symptoms suggestive of paraneoplastic syndromes when presented with patients with cutaneous malignancies.

Keywords: Cutaneous T-cell lymphoma • PCD • Purkinje cells • LEMS • Antibodies

Introduction

Cutaneous T-cell lymphoma is a relatively rare malignancy [1]. Paraneoplastic syndromes are also relatively uncommon sequelae often associated with particularly malignancies and can affect a variety of body systems. Lambert Eaton Myasthenic Syndrome (LEMS) and Paraneoplastic Cerebellar Degeneration (PCD) are autoimmune paraneoplastic syndromes that are caused by cross-reacting onconeural antibodies, which arise in response to malignant tumors, producing neurological symptoms [2].

Paraneoplastic cerebellar degeneration is one of the more commonly seen paraneoplastic neurological syndromes. It is caused by immune-mediated injury to cerebellar Purkinje cells and is found to be associated with a high prevalence of anti-yo antibodies [2]. It is most seen with breast and pelvic malignancies, though it is very rare and is thought to affect less than 1% of patients with cancer. Patients with paraneoplastic cerebellar degeneration can present initially with mild symptoms such as unsteady gait, double vision, and difficulty with fine hand movements. These symptoms usually progress to limb and truncal ataxia. MRI of the brain is usually normal. For a definitive PCD diagnosis, the patient must have severe cerebellar symptoms for less than 12 weeks with a normal brain MRI, and the patient must also have a moderate disability with at least a score of 3 on the Modified Rankin Scale. Cerebellar symptoms with onconeural antibodies are also definitive for PCD. There are currently no evidence-based treatment guidelines. PCD has been associated with Hodgkin's Lymphoma, but there has been no association with Cutaneous T-Cell Lymphoma [3].

LEMS is a neuromuscular disorder involving autoantibodies against presynaptic voltage-gated calcium channels. It often arises in a paraneoplastic setting and is particularly associated with small cell lung cancer. LEMS is not highly associated with lymphomas and no association has been made with cutaneous T-cell lymphoma [4]. In 50% of the cases with lymphoma and LEMS or myasthenia, the neurological disorder develops around the time of the diagnosis of lymphoma. In SCLC, the presence of anti-SOX1 antibodies can help distinguish paraneoplastic LEMS from non-paraneoplastic LEMS [5].

Cerebellar ataxia is a broad term encompassing a variety of disorders wherein ataxia is the primary symptom [6]. It can arise in the setting of paraneoplastic syndromes, as well as in the setting of infection, shock, vascular disease, and metabolic deficiencies among other causes. It can also be associated with anti-P/Q-Calcium Channel antibodies. The presence of antibodies against P/Q-VGCC and N-VGCC has been shown to not be particularly useful in determining neurological phenotype, autoimmune diagnosis, or cancer type [7].

Case Report

A 67-years-old African American female with a recent diagnosis of CTCL presented with blurry vision, lightheadedness, and gait instability. She had not yet begun treatment of her CTCL nor had she undergone any staging workup. She was noted to initially be hypertensive, though symptoms persisted after blood pressure returned to normal limits. She was found to have gait and heel-to-shin ataxia. The remainder of her neurologic exam was normal.

She underwent a full malignancy workup, including an MRI with and without contrast of the brain and spine to rule out meningeal carcinomatosis. There was no evidence of metastatic spread. A lumbar puncture revealed no abnormalities and a paraneoplastic panel was negative. On serum testing, she was found to have anti-P/Q-Calcium Channel antibodies.

Before antibody results returning, the patient was started on five days of IVIG 2 g/kg per a high clinical suspicion of paraneoplastic cerebellar degeneration. Her ataxia resolved, and she was discharged without further episodes.

Discussion

Given the overall low incidence of CTCL and paraneoplastic syndromes, it is unsurprising that CTCL has not been readily associated with paraneoplastic syndromes.

This patient was initially treated under the presumptive diagnosis of paraneoplastic cerebellar degeneration given the preponderance of cerebellar and ataxic symptoms. Ultimately, her lab testing supported a diagnosis of cerebellar ataxia in the setting of Lambert Eaton Myasthenic Syndrome based on the presence of anti- P/Q-Calcium Channel antibodies in her serum and the absence of anti-yo antibodies in her cerebrospinal fluid. With five days of IVIG, she was able to fully recover without further incidences.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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