CASE REPORT

Autoimmunity and Immunodeficiency Coexisting in the Same Patient

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Abstract

Myasthenia gravis is a well-known autoimmune disorder. Very few cases in literature showed association of myasthenia gravis, thymoma, and oral candidiasis. Here, we present a case of myasthenia gravis with thymoma, significant weight loss, extensive oral and esophageal candidiasis mimicking an immunodeficiency state which often creates a therapeutic dilemma in instituting immunosuppression.

Keywords: Acetylcholine receptor antibodies, Esophageal candidiasis, Good syndrome, Mucocutaneous candidiasis, Myasthenia gravis, Thymoma, Weight loss.

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Background

Myasthenia gravis is a well-known and extensively studied autoimmune disorder, where antibodies are mediated against nicotinic receptors at neuromuscular junction. Antibodies against acetylcholine receptor (AchR) are detected in 80% of patients with myasthenia gravis.[1] About 10% have associated thymoma.[1] Approximately 40–50% thymomas are associated with myasthenia gravis.[1] Here, we present a case of myasthenia gravis and thymoma presenting with significant weight loss, oral mucocutaneous, and esophageal candidiasis mimicking an immunodeficiency state.

Case Report

A 37-year-old female came with complaints of right eyelid drooping with diurnal variation of 2 years duration. She also complained of vomiting immediately after taking food, burning sensation in the mouth and heart burn in the previous 1 year and lost 15-kg body weight over this period, she also complained of head drop on and off over the past 3 months, difficulty in chewing and swallowing, difficulty in getting up from chair and squatting position, difficulty in combing and brushing, difficulty in lifting head from supine position, and dependent on her daughter for day-to-day activities due to fatiguability over the past 3 months. She was investigated outside for immunodeficiency and malignancy which had no yield. On examination, she was pale with extensive oropharyngeal candidiasis and candidal stomatitis, she also had right eyelid ptosis (Fig. 1), bedside fatiguablity test was positive and quantitative myasthenia gravis (QMG) score was 15. Blood investigations mild iron deficiency anemia, renal, liver, and thyroid function tests were normal, and AchR was more than 8 nmol/L. Serum immunoglobulin G (IgG), immunoglobulin M (IgM), and immunoglobulin A (IgA) levels were normal; cluster of differentiation (CD4) and CD8 ratio was also normal. The repetitive nerve stimulation (RNS) testing showed decremental response of more than 10% in both orbicularis oculi. Computed tomography (CT) chest showed lobular mass in the anterior mediastinum without evidence of local infiltration suggestive of thymoma (Fig. 2). Computed tomography abdomen was normal. In view of recurrent vomiting, medical gastroenterology opinion was sought and upper gastrointestinal endoscopy was done which showed pangastritis with mid and lower esophageal candidiasis. Barium swallow showed shaggy mid and lower esophagus suggestive of candidiasis. In view of

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Fig. 1: Right eyelid ptosis

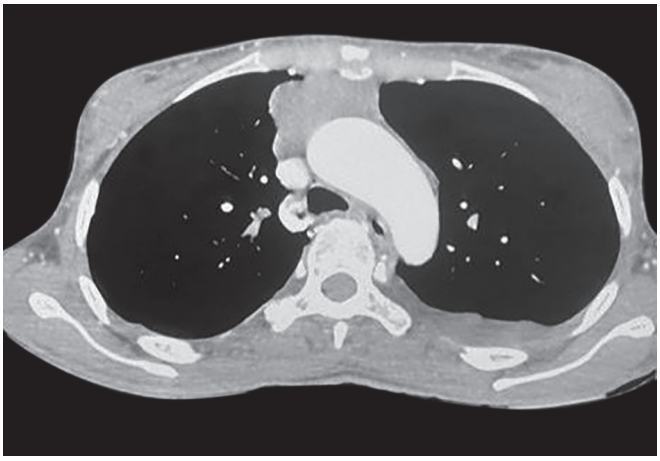

Fig. 2: Computed tomography scan showing lobular thymic enlargement

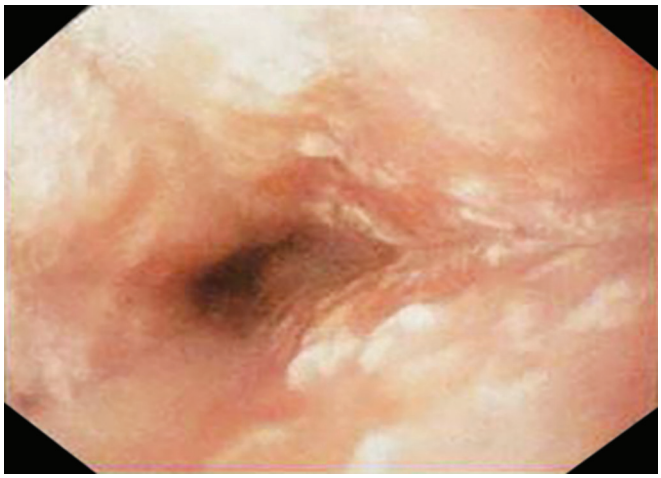

Fig. 3: Esophageal candidiasis

oropharyngeal and esophageal candidiasis (Fig. 3), workup for HIV, HbSAg, HCV, and Good syndrome was done and was found to be negative.

Discussion

Very few cases have been published in literature showing association of myasthenia with thymoma and candidiasis. Boras et al. reported a case of myasthenia gravis with thymoma and oral candidiasis.[1] Such combination can occur in isolation or in association with good syndrome. Good syndrome is a rare acquired immunodeficiency syndrome first described by Robert Good in 1954, which has got worst prognosis with mortality in the range 44–50%, when compared with other acquired immunodeficiency syndromes.[2] Moreover, 0.2–6% of thymomas are associated with Good syndrome.[3] This rare condition can present with variety of infections such as Pneumocystis carinii pneumonia, cytomegalovirus (CMV) retinitis, disseminated aspergillosis, tuberculosis, and candidiasis, herpes zozter, Kaposi sarcoma, chronic diarrheal illness secondary to salmonella and shigella like any other immunodeficiency states.[4] Variety of autoimmune diseases are found to be associated with thymoma. Thymomas tend to release immature T cells into circulation. Normal thymic architecture is essential for normal development of T lymphocytes which is impaired in Thymoma.[5] However, in our case, workup for Good syndrome was negative. Porter and Scully reported that ulceration of tongue, buccal mucosa, and palate in patients with thymic tumor.[6] Rothberg et al. proposed screening for thymoma as a standard procedure in late onset severe and persistent oral candidiasis.[7] Often this association leads to decreased food intake and rarely spread of oral to esophageal candidiasis and further dissemination leading to higher mortality.[8] Often this immunodeficiency-like state creates a therapeutic dilemma in prescribing immunosuppressants. Our case was started on steroids, pyridostigmine and mycophenolate mofetil, proton pump inhibitors, and antifungals. Thymectomy was planned at a later date.

Conclusion

The coexistence of myasthenia gravis, thymoma, and candidiasis in the same patient with features of autoimmunity and immunodeficiency proved to be a diagnostic and therapeutic challenge. Recognition of this rare association is essential to decide long-term management of these patients.

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