Case Report

Extensive Onychomycosis in a Patient with Good Syndrome

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Abstract

Thymoma with immunodeficiency was first reported by Dr. Robert Alan Good and was termed Good Syndrome. It is a rare acquired immunological disorder with <200 cases reported in the literature. Although described more than 60 years ago, the syndrome is still largely underrecognized by physicians who deal with the variable manifestations in the absence of diagnostic criteria. Here, we describe a patient with Good Syndrome diagnosed with extensive candidal onychomycosis. The patient had a history of excised thymoma and had myasthenia gravis. Further, laboratory investigations showed very low B-cells, CD4 helper T-cells and reduced CD4/CD8 consistent with the described abnormalities associated with Good Syndrome in the literature. Atypically severe variants of onychomycosis could indicate the presence of an immunodeficiency.

Keywords: Good Syndrome, immunodeficiency, onychomycosis, thymoma

Introduction

Good Syndrome, also known as thymoma with immunodeficiency (TWI), is a rare acquired condition with <200 cases reported in the literature.[1,2] It is an immunological disorder characterized by the association of thymoma, low to absent B cells in peripheral blood, hypogammaglobulinemia, and variable defects in cell-mediated immunity.[1] Morbidity and mortality in patients with Good Syndrome appear to be attributed to secondary complications such as infectious, hematologic, and autoimmune diseases.[3] Here, we report a patient with metastatic thymoma and myasthenia gravis with clinical and laboratory findings matching the diagnosis of Good Syndrome presenting to the dermatology clinic with extensive onychomycosis.

Case Report

A 53-year-old Saudi male who presented in April 2018 to the dermatology clinic with extensive subungual hyperkeratosis with white nail plate (leukonychia) of the left index and right thumb for the last 6 months. Nail clipping for potassium hydroxide and culture was obtained which came positive for *Candida albicans*, thus confirming the diagnosis of onychomycosis. The patient was then treated with amorolfine 5% topical solution twice daily. One week later, both nails had spontaneously detached from their nail bed [Figure 1]. His past medical history included seropositive generalized myasthenia gravis diagnosed in 2005 in which he was started on prednisolone 15 mg daily, azathioprine 100 mg once daily and pyridostigmine 60 mg four times a day. A thymoma was diagnosed and resected in 2006 followed by three cycles of radiotherapy and chemotherapy including adriamycin and cisplatin. He subsequently developed a recurrence in 2009 and received six cycles of adriamycin and cisplatin. Two years later, he had another recurrence, and it was resected. He was also treated with partial pleurectomy and resection of a mediastinal mass in 2014 followed by left thoracoscopic excision of pleural mass in 2015. One year later, paraesophageal and pericardium deposits were found, and he underwent metastasectomy followed by left thoracotomy and metastatic pleural deposit resection in the same year. His history also included pulmonary nocardiosis diagnosed in 2017, for which he was treated with long-term trimethoprim/sulfamethoxazole; herpes labials were diagnosed in 2018, for which he was treated with acyclovir. He has a history of frequent admissions to the

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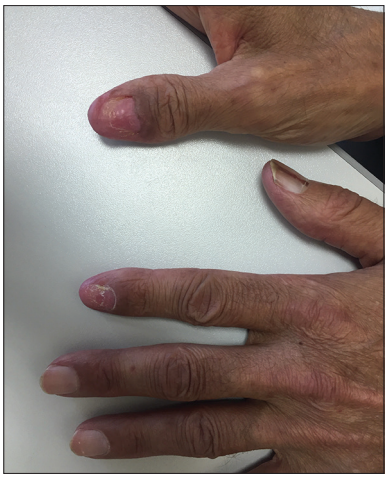

Figure 1: Complete nails detachment with hyperkeratosis of both left index and right thumb following one-week therapy

hospital because of healthcare-associated pneumonia; the last was in February 2018 where he presented to the emergency department with two days history of shortness of breath, fever, and productive cough. He underwent a sputum culture that showed *Aspergillus flavus* and scanty growth of normal respiratory flora and was started on imipenem, azithromycin, and oseltamivir. His recent investigations included a chest computed tomography (February 2018) which showed stable findings and a pulmonary function test (April 2018) that revealed a severe restrictive lung pattern and significant air trapping.

Immunologic analysis showed lymphopenia 6.95% (reference range, 28.00%–39.00%), very low CD4 absolute but normal relative count of 222.00 cells/mcL (reference range, 700.00–1100.00 cells/mcL), very low CD8 absolute but raised relative count of 242.00 cell/mcL (reference range, 500.00–900.00 cells/mcL), critically low B cell count of 3.0 cells/mcL (reference range, 200.0–400.0 cells/mcL), very low NK cell count of 18.0 cells/mcL (reference range, 200.0–400.0 cells/mcL), and reduced CD4/CD8 ratio 0.92 (reference range, 1.00–1.50). His blood smear showed a normochromic anemia with polychromasia, neutrophil was seen with toxic granuloma and normal platelets.

Discussion

TWI was first reported by Dr. Good and Varco in 1955 and was termed Good Syndrome.[4] It is a rare adult-onset, immunological disorder classically defined as thymoma in the presence of low to absent B cells in peripheral blood, hypogammaglobulinemia, and variable defects in cell-mediated immunity with CD4 T lymphopenia and an inverted CD4:CD8 T-cell ratio.[2,3] This syndrome was considered as a subset of common variable immunodeficiency (CVID),[5] however, the reduced or absent number of peripheral B cells that are typically found in patients with Good Syndrome is not a feature of CVID.[6] Good Syndrome is now defined as a diagnostic entity "TWI" independent from CVID.[6,7]

The immunodeficiency in Good Syndrome involves both humoral and cell-mediated immune responses, predisposing the patients to recurrent infections due to bacteria, fungi, viruses and opportunistic infections.[8] As reviewed by Tarr *et al*.,[6] most commonly reported infections in patients with Good Syndrome were recurrent sinopulmonary infections secondary to encapsulated organisms, skin and joint infections, urinary tract infections, and infectious diarrheas. With regard to the autoimmune presentations, myasthenia gravis, pure red cell aplasia, oral lichen planus, aplastic anemia, macrocytic anemia, autoimmune hemolytic anemia, monoclonal gammopathy, myelodysplastic syndrome, diabetes mellitus, and polyarthropathy can occur in association with Good Syndrome.[3] Hematological abnormalities are frequent in routine laboratory examination in patients with Good Syndrome, anemia, and leukopenia were the most prevalent and observed in about half of the patients.[3,9]

Patients with Good Syndrome are at increased risk of death with a greater mortality rate compared to CVID, mainly secondary to infections, autoimmunity, and hematologic complications.[3,10] A high index of suspicion and appropriate investigations are crucially important for early identification and management of Good Syndrome. The management involves resection of the thymoma, intravenous immune globulin, treatment of associated complications such as infections and autoimmune disorders.[3] In addition, live vaccines should be avoided where possible as they may be a serious threat to patients with Good Syndrome.[1]

In conclusion, we identified a patient with Good Syndrome diagnosed with extensive onychomycosis caused by *C. albicans*. The patient had a history of excised thymoma and myasthenia gravis. Further laboratory investigations showed very low B-cells, CD4 helper T-cells and reduced CD4/CD8 consistent with the described abnormalities associated with Good Syndrome in the literature. We propose that unusually extensive or severe onychomycosis may be a guide to consider a primary acquired immunodeficiency such as Good Syndrome. Rising the awareness may lead to earlier recognition, hence, avoid complications and possibly decrease morbidity and mortality in patients with Good Syndrome and related conditions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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