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Case Report

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An Active Duty Patient With Common Variable Immune Deficiency With Early Diagnosis With the Help of the Electronic Record

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Abstract

Electronic medical records have become an integral part to the practice of both military and civilian medicine. One key benefit is the ability to observe the patient as a whole, as opposed to one presenting symptom at a time. We report a case of common variable immunodeficiency in an active duty male who presented with a variety of soft tissue and sinopulmonary tract infections. With the assistance of the electronic medical record, we could view the entirety of the patient's medical history with ease and were able to make the diagnosis. Because treatment with IVIG has been shown to reduce both morbidity and mortality, it is necessary for diagnosis to be made early in our active duty population.

Keywords: CVID; EMR; Immune deficiency; Immunocompromised; Electronic health record; Immunology

Introduction

Common variable immunodeficiency (CVID) is a relatively uncommon diagnosis which is properly made in the setting of an excellent clinical history, laboratory testing, and an insufficient response to vaccinations with no identifiable secondary causes. Frequently practitioners argue about the effectiveness of the advent and incorporation of electronic medical records (EMRs) into common clinical practice. EMRs have been taken with varying degrees of receptivity by physicians and other health care practitioners. Many of them seem frustrated with the inherent redundancies of the EMRs which were specifically highlighted with the earliest iterations. One overlying benefit essential for the diagnosis of our patient was the accessibility of prior health records [1]. Although our patient was seen and treated at numerous facilities to include Fort Benning, Afghanistan, Landstuhl, and Fort Bliss, through the AHLTA electronic record visits for multiple infections and treatment plans were accessed which along with a detailed history and physical exam helped lead to the diagnosis of CVID. The incidence of undiagnosed CVID is far more prevalent than previously believed [2] and early recognition and testing to achieve the correct diagnosis is imperative particularly in the young active duty population. Viewing the entirety of the patient's medical record established a continuity of care previously unavailable in our active duty population, and allowed for the diagnosis of CVID to be made. Because we were able to make the diagnosis, the morbidity and mortality should be significantly decreased in this young, seemingly healthy active duty male.

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Case Report

A 21-year-old Caucasian active duty male combat medic presented to his primary care clinic for treatment of sinusitis, bronchitis and a skin abscess. Searching through AHLTA it was noticed that the patient had a long history of recurrent skin, ear, and lung infections. The patient reported that not only has he had multiple infections during his time in the military but also throughout his childhood he had several bouts of cellulitis, otitis media, bronchitis and methicillin-resistant *Staphylococcus aureus* (MRSA) cellulitis. Three years prior, he joined the military at age 18. While actively involved in basic training, he developed pneumonia confirmed by chest X-ray and was treated with a 7-day course of levofloxacin with symptomatic resolution. Just 1 month later, he developed otitis media of the right ear, for which he was treated with short course amoxicillin. Two months later, he was diagnosed with right thigh cellulitis treated with trimethoprim/sulfamethoxazole (TMP-SMX) treated for 10 days. Interestingly later that month he was deployed to Afghanistan for a 1-year deployment. While he was deployed per modern recommendations for active duty soldiers, he received malaria prophylaxis with daily doxycycline. Of note while in Afghanistan he never suffered an infection. Once he returned from deployment to Landstuhl, he took primaquine daily for 14 days for terminal chemoprophylaxis for malaria. After completion of malaria chemoprophylaxis within the next year and a half, he was diagnosed with four

separate sinopulmonary and skin infections. Two weeks after finishing his primaquine, he was diagnosed with cellulitis of his left arm treated with levofloxacin. The following month, he was diagnosed with pneumonia and treated with levofloxacin. One month later, he was diagnosed with bronchitis treated with azithromycin. One year later, he was diagnosed with bronchitis treated with TMP-SMX.

Medical history

His past medical history aside from his recurrent infections was negative. He took no chronic medications, and had never undergone any surgeries. Regarding his social history, he was an active duty male who lived in the barracks. He smoked a half a pack a day of cigarettes for the past 3 years, and alcohol consumption was limited to approximately five beers a week. He was previously never immunized with the pneumococcal vaccine. Family history was only significant for hypertension, osteoarthritis and pertinently negative for immunodeficiencies, cancers or early deaths of unknown etiology.

Clinical course

On physical exam, his vital signs are normal and he had a BMI of 25. He had bilateral swollen boggy turbinates, sinus tenderness, and pale fluid behind the right tympanic membrane. His lungs were clear to auscultation. There was a small 2 mm pustule on his right thigh over hair follicle with surrounding 3 cm of erythema. Routine labs were performed to include complete blood count, renal panel and liver enzymes which were all within normal limits. HIV test was negative. His IgG, IgA, and IgM levels are all low 53, 11, and 21 respectively. Complete blood count with basic differential showed a normal B-cell level. The aforementioned history of recurrent infections, positive physical exam, and laboratory studies suggested CVID. This was confirmed with a documented lack of immunoglobulin response to the pneumococcal vaccine. Further screening for primary tumors may include in the future a repeat history and physical examination, colonoscopy, and tumor markers, although the efficacy of these tests is yet to be determined. The patient was started on immunoglobulin replacement with human immune globulin, which resolved his recurrent infections.

Discussion

CVID is an idiopathic primary immune deficiency most frequently diagnosed between the ages of 20 and 40, although both children and older adults can be affected [3]. CVID is characterized by low levels of serum immune globulins, IgG, IgA, and/or IgM, with reduced or absent antibody production [4]. This is secondary to defective B cell differentiation and poor production of various subtypes of immunoglobulin. Patients may suffer from acute or chronic infectious processes, and inflammatory, autoimmune, or neoplastic complications [5]. Acute infectious processes are typically caused by encapsulated organisms and involve the sinopulmonary tract, resulting in recurrent sinusitis, otitis media, pharyngitis, pneumonia or bronchitis [6]. The gastrointestinal, urinary, and central nervous systems may also become involved if the condition remains untreated. Chronic lower respiratory tract infections may lead to bronchiectasis and respiratory limitations. Other possible complications include various autoimmune cytopenias, multisystem granuloma formation, and lymphoid malignancies [7]. The minimum age of diagnosis of CVID is 4 years, with exclusion of other immune defects, and requires the following for diagnosis: low serum IgG, low serum IgA and/or IgM, weak or no response to immunization, and no other defined immunodeficiency [8]. A delay in diagnosis of 6 - 7 years is common, possibly due to the disease's heterogeneous nature and with symptom onset in young adult life. Pneumonia prior to treatment for CVID will result in a 10-fold recurrence rate as compared to a normal patient. Treatment with human immunoglobulin can be achieved through various avenues to include intravenous in hospital preparations, intravenous administration in the clinic setting, home subcutaneous therapy, and subcutaneous pump therapy [1]. Dosing is typically 200 - 600 mg/kg body weight given every 2 - 4 weeks [9]. Survival certainly appears to be dose-related as one trial by the UK research council at a dose of 0.1 g/kg/month had a 10-year survival of 37% [1, 10], and another trial showed at 0.4 g/kg/month survival rate of 78% [1, 11]. Furthermore, overall lung function has been shown to be protected in patients with CVID if they are treated with an adequate amount of IVIG [12]. The heterogeneous nature may cause a patient to seek out several different specialists, and without EMRs, the pattern may not be detected. Further investigation involving pattern recognition programs may help guide future physicians and expand their differential diagnosis. For instance, a simple program that isolates a patient with recurrent similar infections based upon parameters such as noted > 5 chronologically separate antibiotic prescriptions generated in the military electronic ordering system, CHCS, per year, recurrence of same site of infection, recurrence of same organism, and noted family history of primary immune deficiency could result in a reflex warning that appears on the providers screen warning the provider that CVID should be a part of the differential diagnosis.

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