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Musculoskeletal Manifestations in Patients with Common Variable Immunodeficiency

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Abstract

Musculoskeletal manifestations are not rare in patients with common variable immunodeficiency (CVID). This study aims to describe different osteoarticular manifestations seen within CVID patients. Eleven patients were collected over 20 years (2000-2022). There were 9 women and 3 men. The median age at diagnosis was 24.2 years (4 years –40 years). Osteoarticular manifestations were reported in 8 cases (72.7%).

Keywords: common variable immunodeficiency, Musculoskeletal manifestations

1. Introduction

Musculoskeletal manifestations are not rare in patients with common variable immunodeficiency (CVID) (Itescu S., 1996; Mucke J, Cornet A, Witte T & Schneider M, 2022; Azizi G, Kiaee F, Hedayat E, Yazdani R, Dolatshahi E, Alinia T, Sharifi L, Mohammadi H, Kavosi H, Jadidi-Niaragh F, Ziaee V, Abolhassani H & Aghamohammadi A, 2018). They include broad-spectrum manifestations from different origin and which are dominated by infections (Mucke J, Cornet A, Witte T & Schneider M, 2022). This study aims to describe different osteoarticular manifestations seen within CVID patients.

2. Methods

This retrospective cohort study was initiated in the Internal Medicine department in Farhat Hached Hospital, Sousse, Tunisia. All adult patients followed for CVID were included. The CVID diagnosis is based on the European Society for Immunodeficiencies (ESID) diagnostic criteria.

3. Results

Eleven patients were collected over 20 years (2000-2022). There were 9 women and 3 men. The median age at diagnosis was 24.2 years (4 years –40 years). Osteoarticular manifestations were reported in 8 cases (72.7%), it's about: acute femur osteomyelitis in 1 case, infectious myositis of gluteal muscle in 1 case, microcrystalline arthritis in 1 case, kienbock's disease in 1 case. Osteoporosis was reported in 2 cases and was complicated by shoulder and wrist fractures. Slow growth with skeletal malformation was observed in 3 cases, the malformations were dorsal kyphosis, syndactyly, macrognathia, and pectus excavatum. Arthralgia secondary to Sjogren syndrome was noted in 1 case. Non-specific degenerative affections were also reported: one case of tendinopathy in the rotator cuff, and 1 case of lumbar disc protrusion.

4. Discussion

Autoimmunity is more frequent among patients with CVID (Azizi G, Tavakol M, Rafiemanesh H, et al, 2017; Azizi G, Ahmadi M, Abolhassani H, et al, 2016) and the CVID is typically observed in childhood and early adulthood, but the autoimmune diseases in patients with CVID increases by age (Tak Manesh A, Azizi G,

Heydari A, et al, 2017; Blancas-Galicia L, Ramirez-Vargas NG & Espinosa-Rosales F, 2010). However, Rhueumatologists should measure Ig levels in patients with autoimmune diseases before the initiation of immunosuppressive therapy and later on.

A wide variety of rheumatologic disorders occurs in patients with CVID, similarly, with a higher prevalence in women than men (Blancas-Galicia L, Ramirez-Vargas NG & Espinosa-Rosales F, 2010; Amer R, Bamonte G & Forrester JV, 2007). Rheumatologic disorders have early onset in comparison with other autoimmunities. Therefore, rheumatologic manifestations seem to be one of the first presentations of the CVID (Mucke J, Cornet A, Witte T & Schneider M, 2022; Azizi G, Kiaee F, Hedayat E, Yazdani R, Dolatshahi E, Alinia T, Sharifi L, Mohammadi H, Kavosi H, Jadidi-Niaragh F, Ziaee V, Abolhassani H & Aghamohammadi A, 2018). In order to reduce the delay in diagnostic and establish timely Ig replacement therapy in these patients, it is critical to clinical immunologist and rheumatologist to be aware of the main manifestations of CVID.

Morover, additional CVID-manifestations are present in a substantial number of patients with CVID and rheumatic diseases. These include bronchiectasis, liver dysfunction, primary biliary cirrhosis, and granulomatous disease as well as gastrointestinal disorders. These patients may have severe manifestations and this can mimic organ disease of the respective rheumatologic disorder (Mucke J, Cornet A, Witte T & Schneider M, 2022).

The musculoskeletal manifestations in CVID patients are polymorphous and nonspecific. The association of osteoarticular manifestations and hypogammaglobulinemia or other immune dysregulations should evoke CVID, after elimination of lymphoproliferative syndrome and different infectious and iatrogenic causes of hypogammaglobulinemia.

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