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A Complex Case of Rhupus Syndrome with Vogt-Koyanagi-Harada Syndrome in a Patient with Type 2 Diabetes Mellitus: A Therapeutic Challenge and Multidisciplinary Approach

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ABSTRACT

Vogt-Koyanagi-Harada Syndrome (VKH) syndrome is a autoimmune disorder affecting melanin-pigmented tissues, with significant ocular manifestation, ear, skin, and central nervous system. We report a rare patient with a VKH Associated With Rhupus syndrome.

Case Report

49-year-old lady with a history of type 2 diabetes mellitus presented with abdominal pain, breathlessness, and visual impairments. Diagnostic evaluation confirmed VKH syndrome, characterized by bilateral diffuse uveitis and other systemic involvements.. Further investigations revealed an overlap syndrome of SLE and RA, commonly known as Rhupus syndrome, complicating her clinical course. The patient was treated with intravenous methylprednisolone, mycophenolate mofetil, and oral prednisone, leading to significant improvement in ocular symptoms and visual acuity

Conclusion

This case underscores the complexity of diagnosing and managing patients with multiple overlapping autoimmune disorders.

Keyword: *Vogt-Koyanagi-Harada (VKH) syndrome, Rhupus syndrome, Systemic lupus. erythematosus (SLE), Rheumatoid arthritis (RA), Autoimmune disorders, Bilateral diffuse uveitis, Visual impairments*

1. INTRODUCTION

VKH is a multisystem autoimmune disorder characterized by bilateral granulomatous panuveitis, often accompanied by neurological, auditory, and integumentary manifestations(4). The standard treatment for VKH involves high-dose systemic corticosteroids, typically initiated with intravenous methylprednisolone followed by oral prednisone. Overlap syndromes in autoimmune diseases are characterized by the presence of clinical features and immunological markers of two or more distinct autoimmune disorders in a single patient(1). In this case, the patient exhibits feature of both SLE and RA, a condition known as Rhupus. The diagnosis of rhupus typically requires fulfilling classification criteria for both SLE and RA, with the presence of specific autoantibodies such as anti-dsDNA (characteristic of SLE) and anti-CCP (characteristic of RA)(2).A multidisciplinary approach involving rheumatologists, ophthalmologists, pulmonologists, and endocrinologists is crucial for optimal management of this complex case.

CASE PRESENTATION

A 49-year-old lady with a history of type 2 diabetes mellitus for 6 years, Presented with gradual onset and progressive impaired vision in both eyes, more notably in the right eye, over the past two months. Additionally, she experienced migrating joint swelling and morning stiffness in multiple small joints, particularly the PIL, MCP, wrist, elbow, hip, knee, and MTS joints, as well as the distal interphalangeal joint,

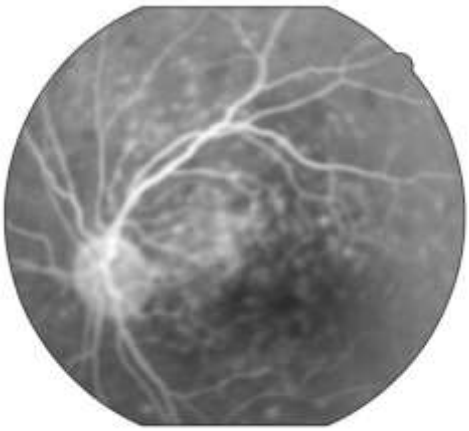
Ocular examination revealed normal facial symmetry, orthophoric eyes and Pigments were observed on the corneal endothelium, and laser iridotomy was noted at 5 o'clock in the right eye and 7 o'clock in the left eye. The pupil measured 4

mm in diameter and was non-reactive to light. No abnormalities were observed in the lens. Best corrected visual acuity was 6/18 in the right eye and 6/36 in the left eye. Fundus examination showed clear media in both eyes with a normal optic disc, arteriolar attenuation, and an epi-retinal membrane with multiple exudates in the macular region. The right eye had an inferior retinal detachment involving the macula and multiple leaks, while the left eye displayed superior and inferior exudative retinal detachments.

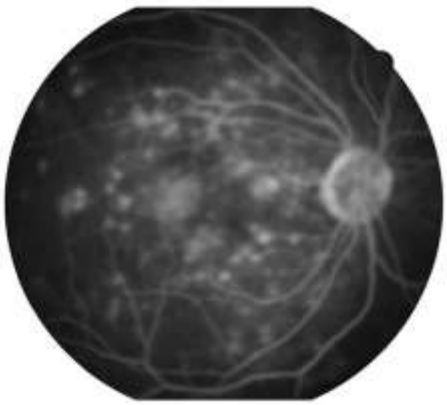
The patient had a history of type 2 diabetes mellitus for 6 years managed with oral hypoglycemic agents. She was evaluated by Rheumatologist and Ophthalmologist. Her RF titer, ANA(IFA), Anti-dsDNA, Anti-CCP titer were positive, suggesting Rhupus syndrome. Other antibodies like Nucleosome Ab, Sm Ab, RNP 68kd/A/C Ab, Sm/RNP Ab, Ssa/Ro 60kd Ab, Scl-70 Ab also were Positive. Her metabolic and biochemical parameters were unremarkable.

The patient received intravenous methylprednisolone at a dosage of 1 gram daily for 3 days, combined with mycophenolate mofetil at 1 gram twice daily. Oral prednisone was initiated at a dose of 1 mg per kilogram of body weight starting on the fourth day. Concurrently, topical antiglaucoma medications and NSAIDs (Nepafenac eyedrops) were continued for both eyes. At a follow-up examination sixteen months later, her eyes were completely quiet with complete resolution of exudative retinal detachment, and her bilateral best corrected visual acuity improved from 6/18 in right eye and 6/36 in left eye to 20/20 (Figure 2). Presently she is asymptomatic on mycophenolate and low dose corticosteroid with significant resolution of vision. Treatment commenced with a three-day course of pulse injectable methylprednisolone followed by oral prednisone. Hydroxychloroquine and oral methotrexate were initiated.

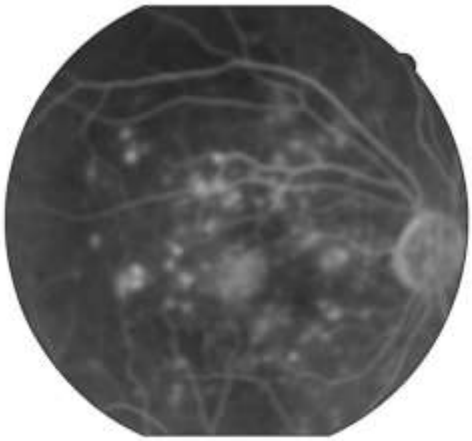
Additionally, she began treatment with sildenafil, perfenidone, and torsemide-spironolactone for her interstitial lung disease and pulmonary arterial hypertension



FFA– Left eye – mid venous phase.



FFA – RE – Mid venous phase.



FFA – RE – Late venous phase.

FFA images showed multiple leaks in the retina favoring the diagnosis of VKH.

2. CASE DISCUSSION

Vogt-Koyanagi-Harada syndrome is an uncommon systemic autoimmune disorder marked by bilateral granulomatous inflammation of the posterior or panuveitis of the eye. This condition can also be accompanied by neurological, auditory and skin-related symptoms.[4].

Rhupus syndrome is a rare condition consists of SLE and RA, that has its own clinical manifestations and laboratory markers, with female predominance (1).RA manifestations presented first, then those of SLE, in most cases. Conversely, reports of concurrent symptoms and vice versa are infrequent. In our case, our female patient had a diagnosis of DM for 6 years and then began having articular manifestations given by chronic, symmetric, ANA Positive, Anti-dsDNA, Anti-CCP titer, Nucleosome Ab, Sm Ab, RNP 68kd/A/C Ab, Sm/RNP Ab, Ssa/Ro 60kd Ab, Scl-70 Ab also were positive, suggesting Rhupus syndrome (RA+SLE). for this reason, it differs from the generality of cases of Rhupus previously reported.

Rhupus patients have been found to have a lower incidence of malar rash, haemolytic anaemia, and renal and neurological involvement compared with the SLE group, as in our case. In addition, rhupus patients rarely have severe renal disorders such as nephrotic syndrome and renal insufficiency. The disease activity in SLE, initial corticosteroid dosages, or pulse have been found to be lower in the rhupus patients (2),. Previous studies have also shown that rhupus patients have a incidence of visceral organ involvement compared with SLE patients without RA. SLE shows 3 types of articular involvement: intermittent non-erosive polyarthritis usually found in the hands, wrists, and knees; non-erosive deforming arthritis referred to as Jaccoud's joint; and arthritis with joint deformities and specific erosion, i.e., Rhupus syndrome. Most patients with SLE have transient, migratory, and reversible arthritis without erosion (2). Some rheumatologists suggest that the presence of rheumatoid nodules in SLE patients could be a risk factor for Rhupus syndrome (2).

The definitive diagnosis of Rhupus is done according to the presence of specific immune markers such as anti-Smith or anti-CCP. In addition to the positivity of ANAs, and rheumatoid factor((2) (3). The presence of anti-CCP has been used to distinguish RA and Rhupus syndrome from SLE (2)In our case, the positivity of anti-Smith and anti-CCP antibodies supports the criterion that Rhupus syndrome is an overlap syndrome and not a variable of SLE.

Hands X-ray may show juxta-articular bone demineralization and erosions which are part of the diagnostic criteria for RA.

Hydroxychloroquine, corticosteroids, and methotrexate are used to control the inflammatory arthritis and alleviate constitutional symptoms.((2), (3), as we did. Other treatments such as mycophenolate mofetil and biological therapy have been used in the presence of important organ involvement. (2) (3), To the best of our knowledge, VKH combination with Rhupus has not been reported in literature. (2)

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