Waldenstrom Hypergammaglobulinemia Secondary To Primary Sjögren Syndrome In An Adolescent: A Case Report

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Abstract

Introduction: Waldenstrom hypergammaglobulinemia is a rare condition, particularly when secondary to primary Sjögren syndrome in adolescents. This combination of disorders presents with atypical symptoms, making diagnosis challenging, especially in younger patients. This case report highlights a rare presentation of Waldenstrom hypergammaglobulinemia secondary to primary Sjögren syndrome in an adolescent female.

Case report: An 18-year-old Indian female presented with recurrent petechiae and purpura, exacerbated by tight clothing and orthostatic positioning, along with burning sensations at rash sites, dry eyes, and intermittent facial swelling. Laboratory tests revealed mild anemia, elevated liver enzymes, and a positive antinuclear antibody (ANA) profile with SSA/Ro antibodies. She was diagnosed with Waldenstrom hypergammaglobulinemia secondary to primary Sjögren syndrome and treated with hydroxychloroquine and prednisolone, resulting in symptom improvement.

Discussion: The association between Waldenstrom hypergammaglobulinemia and primary Sjögren syndrome in this age group is rare, highlighting the need for clinicians to consider autoimmune etiologies in similar presentations.

Conclusion: Early diagnosis and a multidisciplinary treatment approach are essential in managing this rare presentation of Waldenstrom hypergammaglobulinemia secondary to primary Sjögren syndrome in adolescents, ensuring better outcomes and preventing complications.

Keywords: Waldenstrom hypergammaglobulinemia, primary Sjögren syndrome, purpura, adolescent, autoimmune disorders.

Introduction

Hypergammaglobulinemic purpura of Waldenström is an uncommon condition, primarily affecting women, and typically presenting on the lower extremities. It is sometimes associated with underlying immune dysregulation, with Sjögren syndrome being the most common associated condition. However, there have been rare cases where the syndrome resolves spontaneously or is linked to lymphoma or myeloma.1 The symptoms of this disease are due to the infiltration of the bone marrow and extramedullary sites by malignant B cells and elevated IgM levels. When associated with primary Sjögren syndrome, it presents unique diagnostic and management challenges. Primary Sjögren syndrome is relatively uncommon in adolescents, and its presentation in this age group can involve atypical symptoms, making diagnosis more difficult. Cutaneous features are among the most characteristic extra-glandular manifestations of primary Sjögren syndrome. A recent study described a wide spectrum of cutaneous lesions in patients with primary Sjögren syndrome, with vasculitis detected in 10% of cases.2 A diagnostic workup for suspected hemolytic anemia is essential and should include cold agglutinin titers, direct Coombs test, haptoglobin, lactate dehydrogenase, and reticulocyte count.3

Case report

An 18-year-old Indian female presented with a persistent history of petechiae and purpura, which were exacerbated by tight clothing and orthostatic positions. The patient also experienced burning sensations at the sites of the rash, dry eyes, occasional facial swelling, and intermittent abdominal pain over the past 3-4 years. Her medical, family and psychosocial history were largely insignificant, except for a history of mumps two years ago. There was no significant family history of autoimmune disorders.

Upon diagnostic assessment, laboratory investigations revealed mild anemia with an elevated red blood cell (RBC) count. Liver function tests indicated increased levels of serum glutamic-pyruvic transaminase (SGPT) and serum glutamic-oxaloacetic transaminase (SGOT), suggesting possible hemolysis, a condition associated with Waldenstrom's macroglobulinemia. An antinuclear antibody (ANA) profile demonstrated a speckled pattern with an intensity of 4+, and specific bands for SSA/Ro60kD, SSA/ Ro52kD, and SSB antibodies were detected. These clinical and laboratory findings led to the diagnosis of Waldenstrom hypergammaglobulinemia secondary to primary Sjögren syndrome.

The patient was initiated on a treatment regimen that included hydroxychloroquine, prednisolone, and topical ocular lubricants. Hydroxychloroquine, an antimalarial medication with immunomodulatory properties, was prescribed to reduce disease activity. Prednisolone was administered to manage the underlying autoimmune component of primary Sjögren syndrome, alleviate inflammation, and control symptoms such as facial swelling. The patient was advised to use carboxymethylcellulose sodium ophthalmic drops to relieve dry eye symptoms.

Following the initiation of treatment, the patient showed significant improvement in her cutaneous vasculitis and a reduction in erythrocyte sedimentation rate (ESR), serum gamma globulins, and IgM and IgG rheumatoid factors. The patient was subsequently referred to a rheumatologist for further evaluation and management. Continued follow-up assessments demonstrated sustained improvement in her symptoms with ongoing treatment.

Discussion

The association between Waldenstrom hypergammaglobulinemia and primary Sjögren syndrome is particularly rare in adolescent patients. Some studies have linked cutaneous purpura with an increased risk of lymphoma and mortality,4,5 although it is generally mild, self-limiting, and considered an unusual form of chronic leukocytoclastic vasculitis. The presentation of petechiae and purpura, worsened by tight clothing and orthostatic positions, along with dry eyes and facial symptoms, creates a complex diagnostic challenge. This case underscores the importance of considering autoimmune causes in young patients with unusual skin symptoms and highlights the necessity for a multidisciplinary approach in management and treatment. CD20, a protein expressed exclusively on B-cells, is a viable therapeutic target for B-cell malignancies, including Waldenstrom macroglobulinemia (WM).6

While some reports have suggested that familial cases of WM may present at a younger age,7 this observation has not been confirmed in subsequent studies.8 The presence of systemic symptoms combined with significant immunological abnormalities can raise concerns, leading to extensive and sometimes unnecessary investigations. A similar report described three young women with hypergammaglobulinemic purpura of Waldenström (HGPW), who experienced recurrent, asymmetrical purpuric lesions on their lower extremities, triggered by prolonged standing or tight clothing, accompanied by constitutional features.9 Increasing awareness of hypergammaglobulinemia in Waldenstrom may improve diagnosis, especially in patients with non-thrombocytopenic purpura or cutaneous vasculitic syndromes characterized by palpable purpura. While many patients with primary hypergammaglobulinemic purpura may not require specific treatment, those with frequent or severe

symptoms have responded to therapies including colchicine, prednisolone, hydroxychloroquine, indomethacin, chlorambucil, rituximab, and plasmapheresis.10-12
In this case, the differential diagnosis included other autoimmune disorders such as systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA), as well as infectious causes of purpura. However, the presence of specific autoantibodies characteristic of primary Sjögren syndrome supported the final diagnosis. Long-term management will require regular follow-up to monitor treatment response and manage potential complications.

Conclusion

This case highlights an unusual instance of Waldenstrom hypergammaglobulinemia linked to primary Sjögren syndrome in a young woman. The patient's symptoms—such as rashes, dry eyes, and facial swelling—were not typical for someone her age, making the diagnosis challenging. However, by recognizing the possibility of an underlying autoimmune condition and using a team-based approach, her symptoms were successfully managed. This case serves as a reminder for doctors to consider autoimmune disorders even in young patients with atypical dermatological manifestations.

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