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Anterior Scleritis in the Setting of Multiple Autoimmune Disorders: A Rare Case Report and Literature Review

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Case Report

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Abstract

Background:

Scleritis is an uncommon inflammatory ocular disorder with heterogeneous etiologies. Although scleritis has occasionally been reported in association with Sjögren's syndrome and antiphospholipid syndrome, its occurrence in patients with multiple concomitant autoimmune diseases is exceedingly uncommon. This is also the first case suggesting a possible association between polymyositis and anterior scleritis. This case may serve as a useful reference for clinicians in managing similar conditions with multiple concomitant autoimmune diseases.

Case presentation:

A 65-year-old woman with long-standing polymyositis, Sjögren’s syndrome, and antiphospholipid syndrome developed severe left ocular pain following treatment for suspected polymyositis-related pulmonary hypertension with prednisolone and mycophenolate mofetil. Ophthalmic examination revealed mild ptosis, eyelid swelling, diffuse bulbar conjunctival injection with prominent corkscrew vessels, and superficial keratopathy in the left eye. A diagnosis of anterior scleritis was made. Initial treatment with topical fluorometholone and supportive therapy resulted in resolution of ocular pain and improvement of inflammation within one week. However, recurrence with superimposed conjunctivitis occurred one week later. Oral diclofenac, topical levofloxacin, frequent preservative-free artificial tears, and carbomer gel were subsequently administered. The ocular inflammation gradually resolved over three weeks.

Conclusion:

This case highlights a rare occurrence of anterior scleritis in the setting of multiple coexisting autoimmune diseases and suggests a potential association between polymyositis and scleritis that warrants further investigation. Scleritis may develop despite ongoing systemic immunosuppressive therapy, emphasizing the importance of prompt ophthalmic evaluation in patients with autoimmune diseases who develop ocular pain or inflammation.

Background

Scleritis is an uncommon inflammatory ocular disorder with heterogeneous etiologies, including infectious, autoimmune, and masquerade conditions.(1) Approximately 40% of patients with scleritis have an underlying systemic autoimmune disease, most commonly rheumatoid arthritis (RA) and systemic vasculitis. (1) We report a rare case of anterior scleritis occurring in a patient with multiple autoimmune diseases—polymyositis, Sjögren’s syndrome (SjS), and antiphospholipid syndrome (APS)—each of which is infrequently associated with scleritis. This study was conducted in accordance with the

Declaration of Helsinki and was approved by the Taipei Medical University Joint Institutional Review Board (Approval No. N202605104). Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

Case Presentation

A 65-year-old woman with a long-standing history of polymyositis, SjS and APS was receiving prednisolone 5 mg once daily and mycophenolate mofetil 180 mg once daily. Previous serological testing revealed positive anti-β2 glycoprotein I IgM, anticardiolipin IgM, antiphospholipid IgM, anti-Sjögren's syndrome-related antigen A (anti-SSA), and antinuclear antibodies, with negative anti-Sjögren's syndrome-related antigen B (anti-SSB), anti-myeloperoxidase, anti-proteinase 3, and anti–double-stranded DNA antibodies.

She was admitted to the internal medicine ward due to shortness of breath for three days, suspected to be polymyositis-related pulmonary hypertension. Cyclophosphamide 150 mg was administered once, along with dexamethasone 4 mg twice daily for two weeks. After clinical improvement, dexamethasone was tapered to prednisolone 5 mg once daily.

Subsequently, the patient developed severe pain in the left eye. Best-corrected visual acuity was 0.3 (decimal) in both eyes. Intraocular pressure was 24 mmHg in both eyes. As shown in Fig. 1A, mild ptosis and puffy eyelid was noted in her left eye. Slit-lamp examination of the left eye revealed diffuse bulbar conjunctival injection with corkscrew vessels, inferior superficial keratopathy, and upper eyelid edema (Fig. 1B). The anterior chamber was clear, without cells or flare. Nuclear sclerosis was noted. Dilated fundus examination demonstrated a clear vitreous without cells or haze, and no retinal or choroidal abnormalities were identified. Ocular ultrasonography revealed no T-sign.

A diagnosis of anterior scleritis of the left eye was made. Topical fluorometholone and artificial tears were prescribed four times daily, and brimonidine tartrate was administered twice daily to control elevated intraocular pressure. Oral non-steroidal anti-inflammatory drugs (NSAIDs) were recommended but declined by the patient. After a week, ocular pain resolved, intraocular pressure decreased to 18 mmHg in the right eye and 19 mmHg in the left eye, and conjunctival injection and eyelid swelling subsided. However, one week later, a relapse of scleritis with superimposed conjunctivitis occurred, characterized by ocular pain, eyelid swelling, conjunctival hyperemia, superficial keratopathy, and increased discharge. Oral diclofenac 25 mg three times daily, topical levofloxacin four times daily, preservative-free artificial tears hourly, and carbomer gel once daily were added. The inflammation gradually resolved over three weeks as shown in Fig. 1C and Fig. 1D.

Discussion

We report a rare case of anterior scleritis occurring in a patient with the coexistence of polymyositis, SjS and APS. Scleritis is most frequently associated with RA and antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis, while other autoimmune diseases—including SjS, systemic lupus

erythematosus, Behçet disease, relapsing polychondritis, inflammatory bowel disease, and sarcoidosis—are less commonly implicated.(1–4) APS is an uncommon cause of anterior scleritis, although isolated case reports have described necrotizing scleritis in APS.(5) To our knowledge, scleritis has not previously been reported in association with polymyositis. The coexistence of multiple autoimmune conditions may amplify immune dysregulation, thereby increasing susceptibility to ocular inflammatory manifestations. This case underscores the complexity of autoimmune-mediated ocular inflammation and highlights the importance of comprehensive systemic evaluation in patients presenting with scleritis.

In SjS, lymphocytic infiltration of exocrine glands and systemic immune activation leads to lacrimal gland dysfunction and ocular surface disease, as well as extraglandular manifestations.(6, 7) The reported incidence of scleritis in Sjögren's syndrome ranges from 0.6% to 1.5%.(6, 8) Although rare, scleritis in SjS is clinically significant because it has been associated with an increased risk of vision-threatening corneal complications, particularly in patients with concomitant RA.(8) Therefore, close monitoring for corneal involvement is essential in SjS patients with scleritis. Systemic glucocorticoids and immunosuppressants have been recommended in treating scleritis in SjS. (9, 10) Rituximab has been reported to be effective in refractory cases that failed immunosuppressant treatment.(11)

APS is characterized by autoantibodies directed against phospholipid-binding proteins, leading to a propensity for thrombotic events.(12) Ocular manifestations predominantly involve the posterior segment, particularly retinal vascular occlusive disease.(13) Anterior segment involvement is uncommon but includes dry eye, keratitis, uveitis, episcleritis and scleritis.(13) Anticardiolipin antibodies have also been associated with scleritis, suggesting that microvascular injury and immune-mediated inflammation may contribute to scleral involvement.(14) Previous reports have described successful management using a combination of topical steroids, artificial tears, cycloplegic agents, systemic corticosteroids, immunosuppressants, NSAIDs, and anticoagulants.(5) Our patient demonstrated a favorable response to a similar therapeutic approach.

Polymyositis is a chronic inflammatory myopathy characterized by proximal muscle weakness.(15) Ocular manifestations are rare but may include ptosis, dry eye, cataract, retinal hemorrhage, cotton-wool spots, macular edema, vascular occlusion, and proliferative vasculopathy.(16) Anterior segment involvement in polymyositis is rare and no previous research had reported an association between polymyositis and scleritis. Whether polymyositis directly contributes to the development of scleritis remains unclear and warrants further investigation.

Management of scleritis requires a multidisciplinary approach and a combination of local and systemic immunomodulatory therapies, including NSAIDs, corticosteroids, immunosuppressive agents, and biologic therapies.(1) In this case, scleritis developed despite ongoing systemic immunosuppressive treatment, highlighting that ocular inflammation may occur even under apparently adequate systemic disease control. The favorable clinical course following therapeutic adjustment emphasizes the

importance of prompt recognition and tailored treatment to prevent recurrence and potential vision-threatening complications.

Conclusion

This case highlights a rare occurrence of anterior scleritis in the setting of multiple coexisting autoimmune diseases and suggests a potential association between polymyositis and scleritis that warrants further investigation. This case may serve as a useful reference for clinicians in managing similar conditions with multiple concomitant autoimmune diseases.

Abbreviations

RA
rheumatoid arthritis
SjS
Sjögren’s syndrome
APS
antiphospholipid syndrome
anti-SSA
anti-Sjögren's syndrome-related antigen A
anti-SSB
anti-Sjögren's syndrome-related antigen B
NSAIDs
non-steroidal anti-inflammatory drugs
ANCA
antineutrophil cytoplasmic antibody

Declarations

Ethics approval and consent to participate: This study was approved by the Taipei Medical University Joint Institutional Review Board (Approval No. N202605104). Written informed consent for publication was obtained from the patient.

Consent for publication: Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

Availability of data and materials: Not applicable, as no datasets were generated or analyzed during the current study.

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Authors' contributions: CCS contributed to the acquisition and interpretation of the clinical data, and drafting of the manuscript. ICL contributed to the acquisition and interpretation of the clinical data and reviewed the manuscript. TYT supervised the study, contributed to the conception and design of the study, and critically revised the manuscript for important intellectual content. All authors read and approved the final manuscript and agreed to be accountable for all aspects of the work.

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Figures

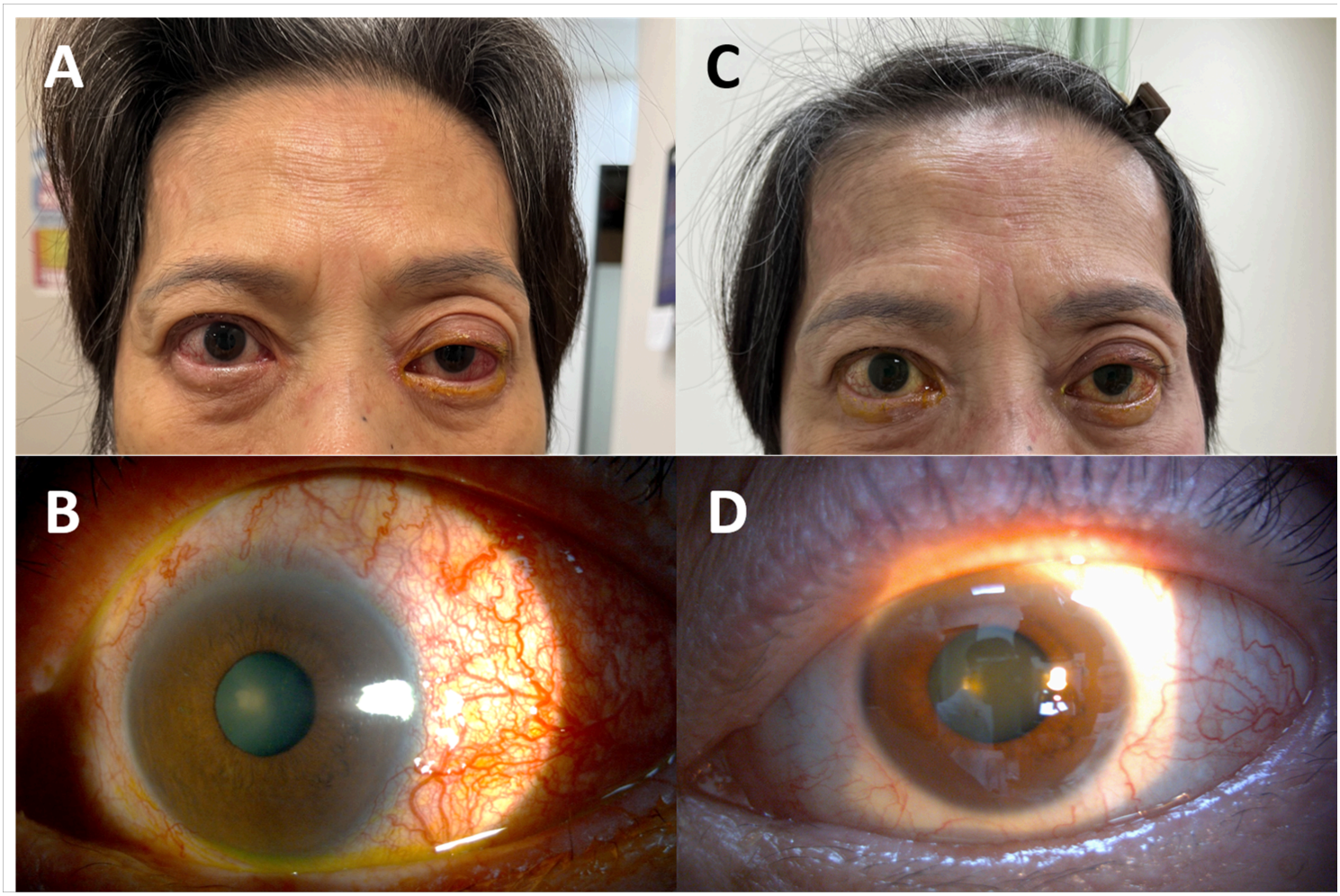


Figure 1

External photographs of this case. At presentation, the patient showed (A) mild ptosis with eyelid swelling in the left eye and (B) diffuse anterior scleritis with conjunctival hyperemia and prominent

corkscrew vessels. Three weeks after systemic and topical treatment, (C) ptosis improved and (D) scleral inflammation significantly decreased.