



Concurrent primary Sjögren's syndrome and isolated ocular sarcoidosis presenting with bilateral corneal scarring and dry eye

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ABSTRACT

Purpose: To report the case of a patient who presents with multiple progressive ocular diseases who is diagnosed with concurrent primary Sjögren's syndrome and isolated ocular sarcoidosis.

Observation: A 60-year-old woman was referred for dry eye disease, bilateral interstitial keratitis, anterior uveitis, and progressive glaucoma. There was clinical suspicion of an autoimmune etiology due to her ocular history, risk factors, and presentation. Thorough diagnostic testing revealed both primary Sjögren's syndrome and ocular sarcoidosis. After 2.5 years of systemic treatment and follow up, the patient currently remains stable.

Conclusions and importance: Autoimmune disease may underlie those with progressive ocular disease with an unknown etiology. More than one autoimmune disease may be the cause of ocular findings, especially for patients with a complicated presentation. Proper awareness, clinical suspicion, and diagnosis of these diseases can greatly improve a patient's condition and prevent future ocular and systemic complications.

1. Background

Sjögren's syndrome is an autoimmune disease that typically manifests with keratoconjunctivitis sicca and xerostomia, with frequent extraglandular complications.¹ Sarcoidosis is also an inflammatory disease that can affect a variety of organs and tissues.^{2,3} Although ocular findings are present in 20–30% of patients with systemic sarcoidosis,³ only a reported 8% of all sarcoidosis cases present with non-pulmonary findings with even fewer manifesting as isolated ocular sarcoidosis.⁴

While sarcoidosis and primary Sjögren's syndrome are each uncommon in the general population, they are both encountered by ophthalmologists managing patients with clinically significant dry eye,^{5,6} and both can lead to corneal scarring.^{1–3} To our knowledge, this is the only report to date describing a patient with both primary Sjögren's syndrome and isolated ocular sarcoidosis, presenting with corneal scarring and dry eye.

2. Report of a case

An African American woman in her 60s was referred in 2017 after being managed for over a decade by ophthalmologists for dry eye. She

subsequently developed bilateral interstitial keratitis and anterior uveitis. She also had progressive glaucoma and cataracts secondary to chronic topical steroid usage. She had a family history of autoimmune inflammatory bowel disease, and review of systems revealed joint pain, fatigue, and dry mouth with history of dental abscess.

The patient's best corrected visual acuity was hand motion right eye and 20/40 left eye. Slit lamp examination revealed inferiorly located mid-stromal corneal scarring without epithelial defects and bilateral nodules on tarsal conjunctiva (Fig. 1A–D). Further testing for dry eye demonstrated significant bilateral bulbar conjunctival lissamine green staining and corneal punctate epithelial erosions (Fig. 1E and F).

A work-up was ordered to reveal possible underlying systemic disease. Serological testing revealed high titer antinuclear (ANA) and anti-Sjögren's syndrome A (SSA) antibodies, confirming a diagnosis of primary Sjögren's syndrome per the American College of Rheumatology's 2016 guidelines.⁷ She also had anti-thyroid peroxidase antibodies with normal TSH levels. Clinical findings of corneal scarring and nodular conjunctival lesions raised suspicion for sarcoidosis. This was confirmed with a conjunctival biopsy which showed chronic inflammation with multiple non-caseating granulomas. A chest x-ray was normal, without hilar or mediastinal adenopathy or pulmonary infiltrates. The patient

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did not present with any cutaneous or systemic manifestations of sarcoidosis on presentation.

The patient's diagnoses were confirmed by a rheumatologist. In response to the vision-threatening ocular inflammation, the patient was treated with oral mycophenolate mofetil and hydroxychloroquine. The patient subsequently underwent a penetrating keratoplasty with cataract surgery to improve visual acuity of her right eye. Over the 2.5 years of follow-up, her progressive glaucoma mandated a trabeculectomy. The patient's last examination in October 2019 showed stable findings with best corrected visual acuity of 20/40 right eye and 20/30 left eye.

3. Discussion

This case illustrates two important points. First is the importance for ophthalmologists to consider an autoimmune or inflammatory cause of ocular surface disease in any patient with clinically significant dry eye.⁶ Several findings on presentation that raised clinical suspicion for autoimmune disease include: progressive ocular disease with an unknown etiology,^{5,6} the patient is an African American woman,^{2,3} the patient has a familial autoimmune condition,^{8,9} review of systems showed joint pain and dry mouth, and the nodular appearance of her conjunctiva (Fig. 1C and D). Despite having the quintessential presenting features of autoimmune disease and having seen several ophthalmologists for over a decade, our patient was neither suspected nor tested for any

autoimmune condition. Sjögren's syndrome in particular, is an underappreciated and therefore underdiagnosed cause of dry eye disease.⁵

The second point is for clinicians to consider additional autoimmune disorders even in a patient with an existing autoimmune disease.^{5,6} Epidemiologic studies demonstrate that autoimmune diseases are inter-connected: in particular Sjögren's syndrome is associated with increased prevalence of sarcoidosis, thyroid disease, rheumatoid arthritis, and others.^{8,10} In our patient, the finding of nodular conjunctivitis raised concern for ocular sarcoidosis and prompted the diagnostic conjunctival biopsy, even though a diagnosis of primary Sjögren's syndrome was already established. Multiple inflammatory and autoimmune systemic diseases may underlie dry eye.^{5,6}

4. Conclusion

An ophthalmologist should consider autoimmune disease as a differential for ocular pathology, especially for progressive diseases with unknown etiology. Multiple autoimmune diseases may be present in a patient, particularly those with a complex presentation. Proper awareness and clinical suspicion of autoimmune diseases allows for early diagnosis and treatment of these progressive diseases, thereby helping reduce ocular and systemic morbidity and greatly improving a patient's quality of life.

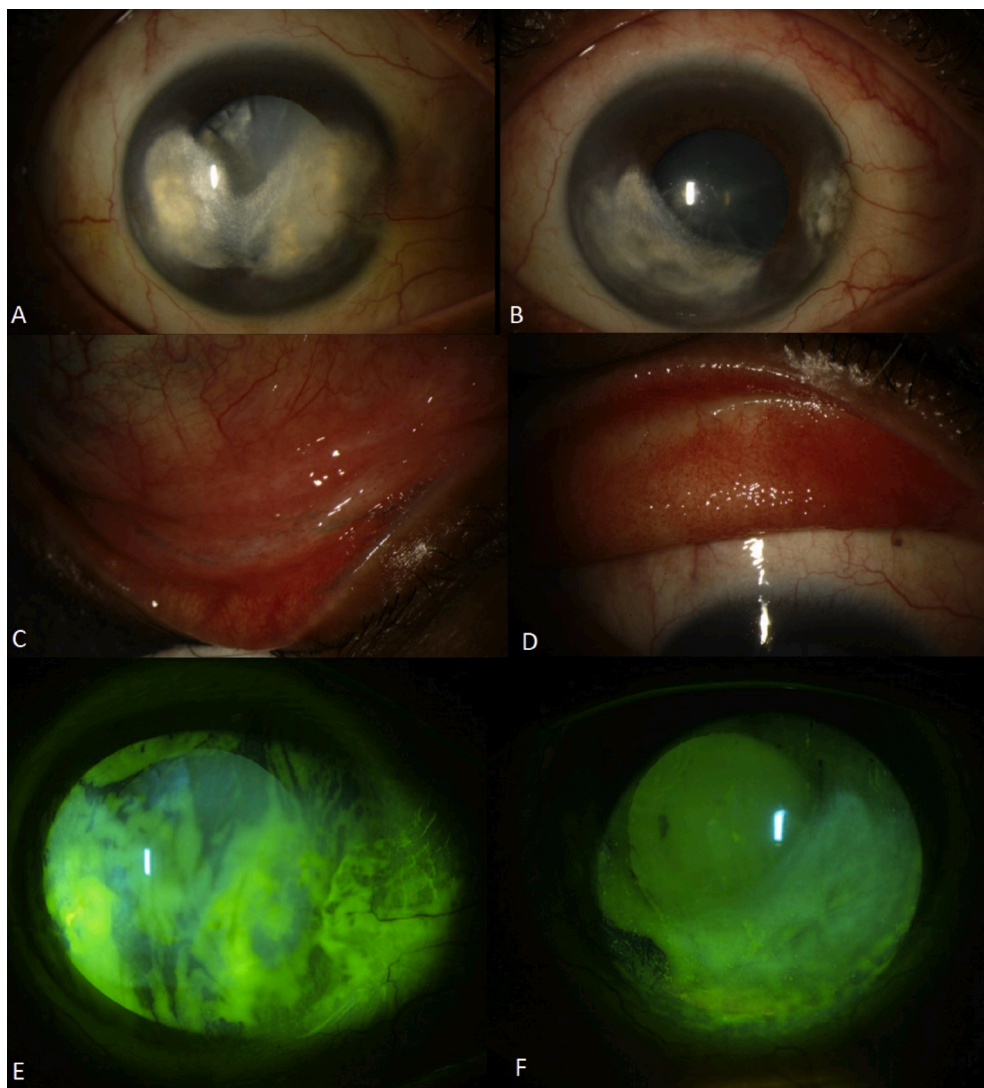


Fig. 1. Clinical findings of a patient with concurrent primary Sjögren's syndrome and sarcoidosis, on presentation.

A–B: Slit lamp photograph similarly in both eyes, centered on the cornea, showing areas of deep stromal scarring and haze spanning from temporal to nasal with overlying lipid deposition and neovascularization.

C–D: Upper and lower tarsal and conjunctiva of the right and left eye respectively demonstrating a nodular appearance.

E–F: Fluorescein staining under cobalt blue light illumination showing poor tear film, coarse punctate epithelial erosions in both eyes. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Patient consent

Consent was not obtained as this report does not contain any personal identifying information.

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Authorship

All authors attest that they meet the current ICJME criteria for authorship.

Declaration of competing interest

No conflicts of interest.

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