Sweet syndrome with ophthalmologic complications

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Introduction

Sweet syndrome, also known as acute febrile neutrophilic dermatosis, is an autoimmune disorder mainly impacting the skin and mucous membranes. Up to one third of cases have ocular involvement. Conjunctivitis is the most common ocular finding.[1]

There are three types: classic or idiopathic, malignancy-associated and drug-induced. The exact pathophysiology is unclear.

Sweet syndrome is more common in females. It tends to affect females aged 30-50 and males aged 50-90.

Case Presentation

A 68 year old female presents with several days of itching, desquamating skin and mucosal lesions. Significant past medical history includes end stage renal disease on peritoneal dialysis, hypertension, history of brain aneurysm s/p clipping and recent diagnosis of ANCA vasculitis. She had been started on hydroxychloroquine by rheumatology for her autoimmune disease two weeks earlier.

She underwent an inpatient work up with dermatology, rheumatology and infectious disease and was initially treated with broad spectrum antibiotics. Skin biopsy suggested autoimmune etiology and she was started on corticosteroid taper with improvement of her symptoms.

Ophthalmology followed closely during her inpatient stay and post-discharge for bilateral ulcerative conjunctivitis primarily affecting her palpebral conjunctiva and eyelid margins. This was treated with amniotic membrane placement, topical steroids, cyclosporine A and prophylactic topical antibiotics with good response. Chronic sequelae included cicatricial entropion, dry eye disease and trichiasis. She was referred to Oculoplastics for entropion repair and trichiasis treatment. Ocular surface disease was treated with frequent lubrication and low dose topical steroid taper.

Case Presentation (continued)

Figure A: ulcerated palpebral conjunctiva and tarsus exposing meibomian glands on initial outpatient presentation

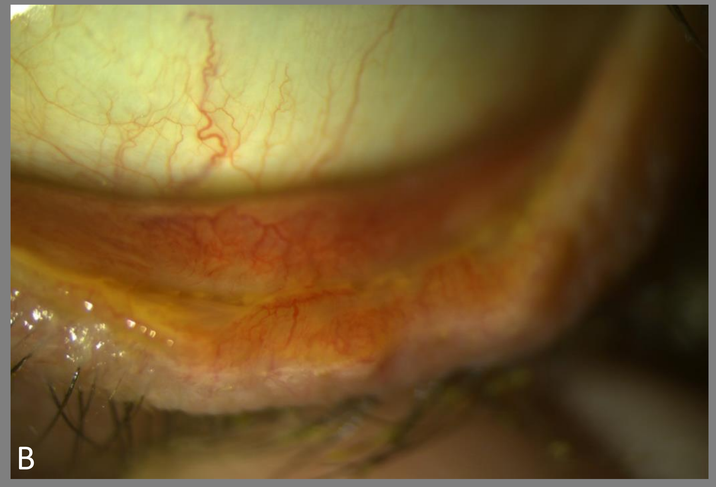

Figure B: 2 months after initial presentation with improvement of ulceration on topical steroid therapy

Figure C: external photo showing impact of posterior lamellar scarring resulting in cicatricial entropion and trichiasis

Discussion

Sweet syndrome is an idiopathic auto-immune disease. It presents with fever, malaise, arthralgias and violaceous or erythematous skin lesions tender to touch and rarely pruritic.[2]

Ocular involvement is highly variable and may affect the orbit and periorbital adnexa as well as the anterior and posterior segment.[1]

Differential diagnosis includes Stevens Johnson syndrome/toxic epidermal necrolysis, erythema nodosum, Behcet disease and erythema multiforme. Diagnosis can be made with skin biopsy showing dense neutrophilic infiltrates without leukocytoclastic vasculitis. Bloodwork shows neutrophilia with elevated acute phase reactants.[4]

Conclusion

While conjunctivitis is the most common ocular complication of Sweet syndrome, it can have a wide range of ophthalmologic manifestations from periorbital skin lesions to severe conjunctivitis and retinal vasculitis.[3]

Sweet syndrome with ocular involvement responds well to systemic corticosteroid therapy with local steroid therapy improving outcomes in severe disease.

Recurrence of ocular disease is uncommon and visual prognosis is generally favorable.

References

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