Peripheral Ulcerative Keratitis: A Novel Presentation

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Presentation
A 46 year-old man presented to our center with a one week history of foreign body sensation in his left eye and blur. The patient reported no trauma and no inciting event. The patient indicated he was using Besivance eye drops every hour in the left eye for which he was prescribed by his outside provider four days earlier. The patient’s uncorrected visual acuity was 20/20 in the right eye and 20/200 in his left, with no improvement with pinhole. The patient’s pupillary exam was normal and IOP was 7 in both eyes. The patient’s exam was notable for +1 left upper eyelid edema, +2 conjunctival injection, most significantly adjacent to the 10 o’clock position where there was a peripheral 2.5mm circumferential corneal ulcer.

There was 70% thinning of the ulcer in the superior nasal aspect of the lesion. There was no obvious infiltrate, however, there was +1 corneal edema with descemets folds tracking to the ulcer location. The patient had a 1.5mm hypopyon with +2 cell/flare in the anterior chamber. The lens, iris and posterior segment exam were normal.

Photo 1: Slit lamp photo of left eye -day one presentation

Workup, Diagnosis, and Treatment
The ulcer was cultured for bacterial and fungal species and samples for HSV and VZV PCR were also sent. The patient was started on valacyclovir 500mg three times daily, doxycycline 100mg two times daily, vitamin C 500mg daily and fortified vancomycin and gentamicin drops every one hour around the clock. On follow up one day later, the patient’s vision was improved to 20/100, however no improvement in ulcer size or hypopyon. Given the lack of infiltrate and the peripheral nature of the ulcer our differential also included an autoimmune etiology and masquerading bacterial species. Labs were sent for CBC, BMP, UA, RF, ACE, ANCA, quantiferon gold and syphilis screening. Two days later, with the same treatment, the patient’s exam findings were stable. All cultures continued to be negative, PCR for HSV/VZV were negative, however, his labs were concerning for +ANCA and PR3 and moderate blood in his urine. With these lab findings the patient was admitted to our hospital for further workup and treatment as we felt this was atypical peripheral ulcerative keratitis in the setting of +ANCA vasculitis. Additional labs were done upon admission, including a toxicology screen which was positive for cocaine and cannabinoids which were not revealed previously.

The patient was started on IV Solumedrol 500mg daily for 3 days and rheumatology and nephrology consults were made. After just one day of IV steroids the hypopyon resolved and the corneal edema improved. Due to the persistent corneal thinning at the superior nasal aspect of the ulcer, histoacryl glue was applied to the area of concern and a Kontour bandage contact lens was placed. The visual acuity continued to improve over the next several days to 20/40. The patient underwent sinus evaluation and kidney biopsy to rule out sinus and kidney involvement. CT showed erosive sinus disease which was felt to be a result of his cocaine abuse and his kidney biopsy was negative for glomerulonephritis. This patient’s +ANCA and PR3 were felt to be a result of the patient’s cocaine dependency which he failed to admit up to the time of his positive drug screen.

Levamisole is an antihelmintic drug and a common cocaine contaminant. Levamisole is believed to potentiate cocaine’s euphoric effects and it is present in an estimated 71% of cocaine samples in the US [1]. Levamisole-contaminated cocaine has been linked to an ANCA-associated vasculitits [1, 2]. The pathogenesis and prevalence of levamisole-induced vasculitis in patients who use cocaine is unclear, although researchers speculate that a byproduct of levamisole may play a role [2, 3].

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The most common initial presentation of levamisole/cocaine-associated vasculitis is purpura (1). Our patient did not have these specific skin findings, however, cocaine itself is also associated with autoimmunity formation, and it is conceivable that exposure to both drugs may contribute to the formation of +ANCA vasculitis [1].

The main treatment of cocaine induced vasculitis is abstinence [3]. Systemic corticosteroids are also used in the treatment of this condition and a multidisciplinary approach is necessary to properly care for these patients [1, 2 & 3]. After 3 days of IV solumedrol, our patient was transitioned to oral prednisone at 1mg/kg/day and he was discharged for outpatient follow up.

At one week follow up the glue and contact lens were still in place with stable 20/40 vision. At the three week follow up the glue had dislodged, so the BCL was removed. The conjunctiva was no longer injected, and the ulcer was now scarred with improvement in thinning to appx 40% depth and there was corneal neovascularization into the affected area.

Compliance with follow-up visits was a challenge in this patient. We have encouraged the patient to continue to follow up with our office and with rheumatology and his primary care physician [4].

References