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Plasmapheresis as a viable treatment option for scleritis

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ABSTRACT

Purpose: We describe a case and our experience with using plasmapheresis as a treatment for scleritis.
Observations: Treating relapsing autoimmune scleritis can be challenging when it inadequately responds to traditional therapy. Our patient could not receive non-steroidal anti-inflammatory therapy for her scleritis due to recent gastrointestinal surgery and previously failed multiple steroid sparing treatments due to intolerance. There was good initial control with high dose oral prednisone, however, the steroid could not be tapered to a safe dosage (<10 mg per day) without relapse. Therefore, we opted to treat our patient with plasmapheresis.
Conclusions and Importance: After undergoing plasmapheresis, our patient experienced total resolution of symptoms with corresponding clinical resolution of scleritis. Plasmapheresis derives great benefit by filtering circulating immune complexes. Although rarely used, plasmapheresis can be effective in treating non-infectious scleritis.

1. Introduction

Scleritis is often relapsing and remitting with varied presentation, from mild to vision threatening scleral inflammation.[1,2] In severe cases, globe integrity can be compromised. Often, there is associated underlying systemic autoimmune disease. Posterior and necrotizing scleritis are particularly difficult to control.[1,2] Some patients with recalcitrant cases require long term high dose steroid therapy and multiple immune modulators. Most forms of non-infectious scleritis are immune mediated and driven by auto-antibodies.[2] Plasmapheresis is a form of extracorporeal filtering procedure that decreases the overall antibody load in the serum. There is a theoretical benefit for antibody driven immune mediated diseases such as scleritis but the current literature for plasmapheresis in the setting of scleritis is sparse.

2. Case report

A 48 year old female with a past medical history of idiopathic intracranial hypertension

presented to the emergency department with a 6 month history of a painful right eye that acutely

worsened 5 days prior to presentation. The patient described the pain as throbbing and tender to

touch. The pain radiated to her right forehead causing severe headaches. On further questioning,

the patient stated she had a history of scleritis for which she was being treated with steroids. She

was started on high dose oral prednisone by an outside physician and tapered to 20 mg per day

currently. Even at the initial high dose, her symptoms were only partially controlled. She was

experiencing systemic and psychological side effects from the steroids. The patient was

previously treated with methotrexate and infliximab which she discontinued due to intolerable

side effects (gastrointestinal upset and rash). Also of note, she was unable to take nonsteroidal

anti-inflammatory drugs (NSAIDS) due to recent gastrointestinal surgery and high risk of

bleeding.

On examination, visual acuity was 20/30 in the right eye and 20/25 in the left eye using a near

card with the patient's own reading glasses. Her intraocular pressure was normal in both eyes.

Anterior exam of the right eye was remarkable for sectoral injection of the sclera with a

violaceous hue superonasally (Fig. 1). There was no anterior chamber inflammation or corneal

abrasion/ulceration. Posterior exam was remarkable for Frisen grade I disc edema of both eyes.

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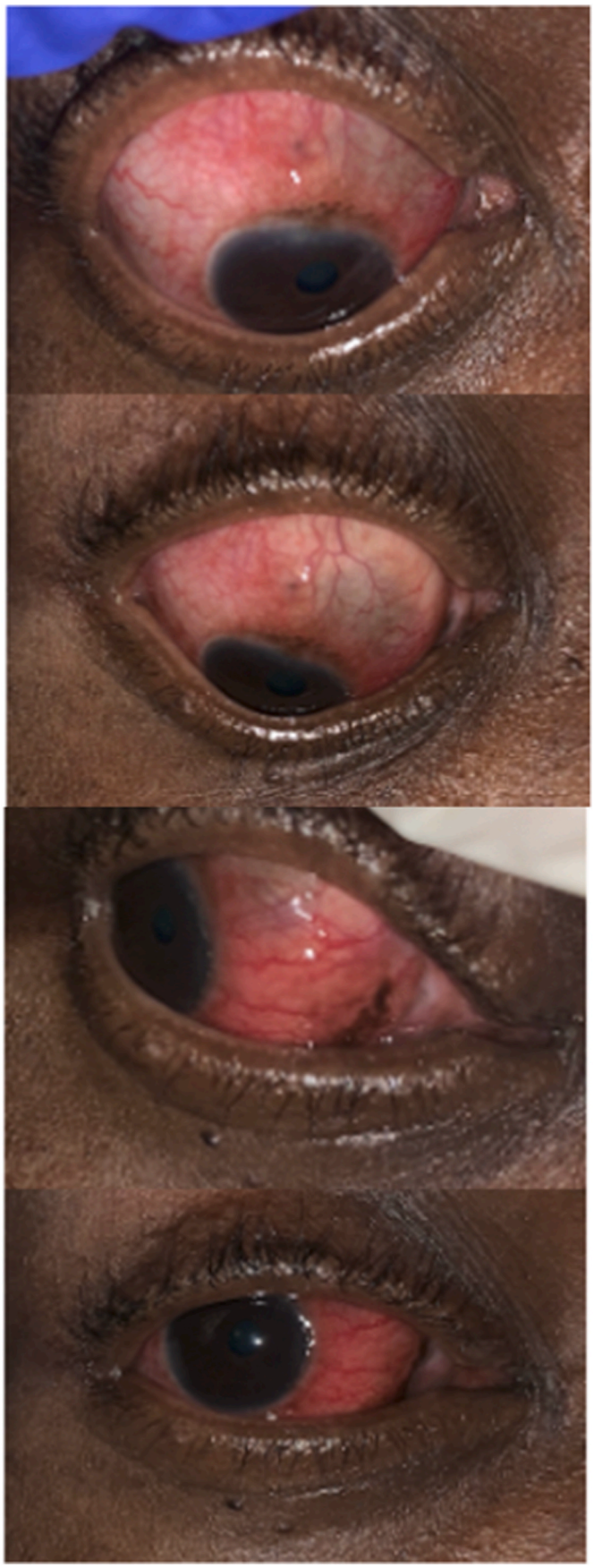

Fig. 1. External photos of the right eye showing scleral injection with violaceous hue superior-nasal.

Otherwise, posterior exam was unremarkable. Of note, she was taking acetazolamide for her

idiopathic intracranial hypertension.

The patient's medical history was remarkable for hypertension and diabetes mellitus. She had

no history of autoimmune disease to her knowledge. Family history was remarkable for

sarcoidosis in her mother.

While the patient was admitted, a rheumatologic work-up revealed positive rheumatoid factor (RF), positive antinuclear antibodies (ANA), elevated erythrocyte sedimentation rate (ESR) and elevated C-reactive protein (CRP). All other inflammatory markers were negative. Chest x-ray was obtained and was negative. Infectious work-up was unremarkable. The patient was evaluated by rheumatology who determined the patient met only 4 of 6 required criteria for rheumatoid arthritis.

Notably, our patient never achieved full resolution of symptoms on steroids. Given that she had a history of steroid induced psychosis and had very poorly controlled blood glucose, IV solumedrol was not a good treatment option in this case. As described above, our patient could not tolerate multiple steroid sparing agents in the past. For these reasons, we decided to treat her with plasmapheresis. Five consecutive daily plasmapheresis sessions were completed successfully with 2.5 L volume exchange and 5% albumin as replacement solution. Our patient did not experience any side effects from the therapy. After just 2 sessions, the patient began to experience clinically significant improvement of ocular pain and headaches. Initially, no additional steroids were given. Repeated clinical exams were done after each session and revealed improvement of scleral tenderness and injection. After the completion of all 5 sessions and prior to discharge, the patient had complete resolution of the scleritis. Mycophenolate mofetil (MMF) was initiated by the rheumatology service prior to the hospital discharge.

The patient presented to our clinic two weeks after discharge. She endorsed total resolution of right ocular pain and headaches. She experienced no flare-ups and experienced no complications. Anterior exam showed significant improvement of scleral injection. (Fig. 2).

3. Discussion

Currently, mainstay therapy for non-infectious scleritis consists of NSAIDs, steroids and various steroid sparing immunomodulatory therapies.[1–3] Many of these treatments have side effects and/or contraindications. Scleritis can be recalcitrant and is not always responsive to traditional therapy. Plasmapheresis is not commonly used for scleritis, but is an effective treatment option given that scleritis is frequently antibody mediated. To our knowledge, this is the first documented case of plasmapheresis used specifically to treat scleritis. The only similar case is a documented case of plasmapheresis used to treat a peripheral corneal ulcer in a patient with benign hypergammaglobulinemic purpura.[4]

At our institution, ocular inflammatory diseases frequently associated with autoantibodies are often treated with plasmapheresis. These include orbital inflammatory syndrome, peripheral ulcerative keratitis and severe scleritis. A protocol similar to those for myasthenia gravis was used. Five to 6 sessions of extracorporeal plasmapheresis is generally performed. This is followed by initiation of a steroid sparing immunomodulatory agent. Our agent of choice is rituximab for preferential targeting of B-cells during this period of hyperactivity following plasmapheresis.

In the literature, other ocular conditions that have utilized plasmapheresis for treatment included idiopathic orbital inflammatory syndrome,[5] neuromyelitis optica (NMO),[6,7] necrobiotic xanthogranuloma,[8] Graves' ophthalmopathy,[9] Behcet's disease,[10] atopic keratoconjunctivitis,[11] bilateral diffuse uveal melanocytic proliferation (BDUMP)[12] and ocular manifestations of toxic epidermal necrolysis (TEN).[13] A recent report also detailed how plasmapheresis can be helpful in treating lupus vasculitis by quickly removing circulating immune complexes.[14]

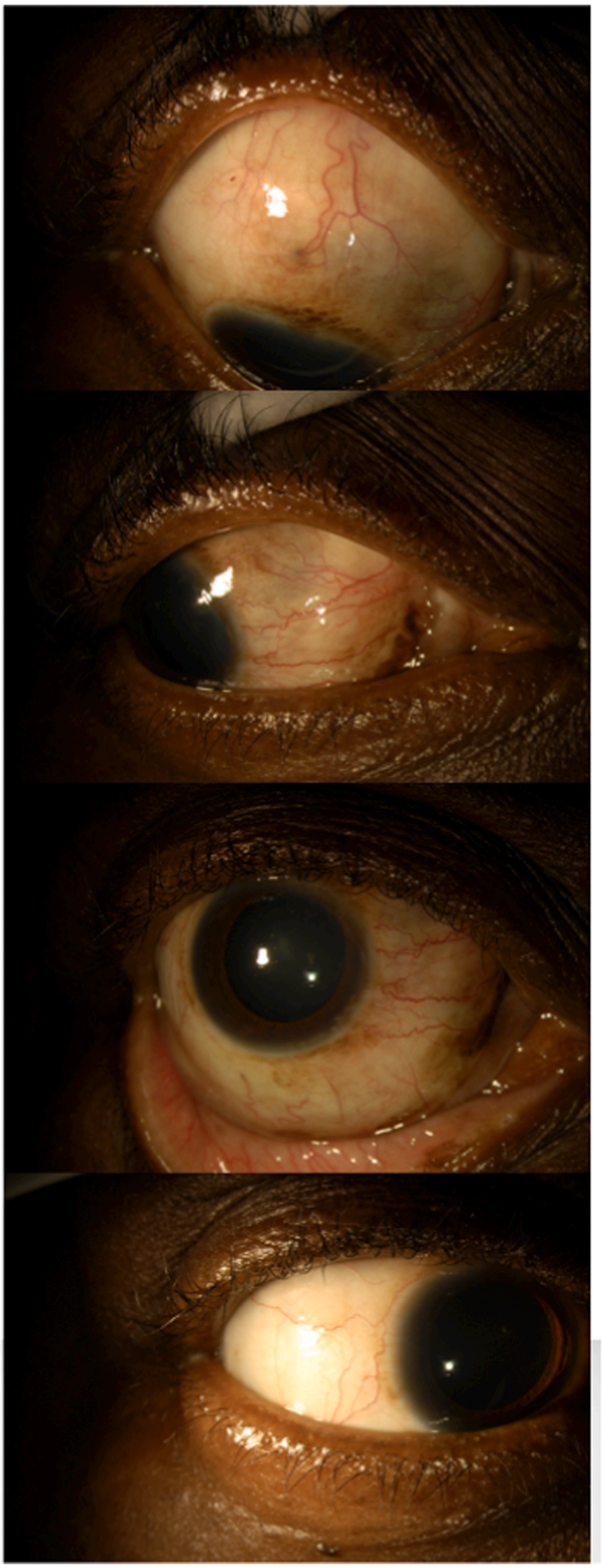

Fig. 2. External photos of the right eye showing resolved scleral injection 2 weeks after plasmapheresis treatment.

4. Conclusions

We present a unique case of a patient with relapsing and remitting scleritis whose symptoms and manifestations resolved quickly after initiating plasmapheresis. Plasmapheresis was effective and well tolerated in our experience. Thus, we believe plasmapheresis should be considered and utilized when treating patients with non-infectious scleritis. Maintenance therapy with immuno-modulatory agents may be needed to achieve long-term remission.

Patient consent

The patient consented to publication of the case orally and in writing. Additionally, this report does not contain any personal information that could lead to the identification of the patient.

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