Granulomatosis with polymyositis (GPA), previously known as Wegener granulomatosis, is a rare and potentially lethal autoimmune disorder. Ocular involvement can occur in up to half of patients with GPA. This article details some of the ocular signs and symptoms, tests, and treatments for GPA that ODs should be aware of as the primary eye care providers for their patients.

As its name implies, GPA is characterized by a necrotizing granulomatous vasculitis of small and medium-sized blood vessels throughout the body. This inflammation often occurs in the upper and lower respiratory tracts, leading to frequent episodes of sinusitis, epistaxis (bloody noses), difficulty breathing, and hemoptysis (coughing up blood). Renal involvement, in the form of necrotizing glomerulonephritis, is also common. In its early stages it may be asymptomatic; however, as renal inflammation progresses it can cause proteinuria and hematuria (protein and blood in urine) and can result in renal failure.

**Ocular Signs and Symptoms**

Orbital inflammation is one of the most common ocular manifestations of GPA, and it may present as proptosis, diplopia, and/or pain. Epiphora secondary to nasal lacrimal duct obstruction has also been reported. Conversely, dacryoadenitis (lacrimal gland inflammation) in GPA can result in severe keratoconjunctivitis sicca.

Scleritis can often be the initial clinical manifestation of GPA. Patients

**Not Your Typical Scleritis**

What ODs should know about granulomatosis with polymyositis.

**By Erich A. Hinel, OD, MS, FAAO, DipL ABO**

- Although rare, ANCA-associated vasculitic diseases such as granulomatosis with polymyositis (GPA) carry a high risk of mortality if left untreated.
- Ocular and orbital involvement can often be early clinical manifestations of GPA.
- An ANCA panel inclusive of PR3 and MPO antibodies should be part of the workup of every patient with scleritis.
- Optometrists can play a crucial role in the early recognition and management of this potentially lethal autoimmune vasculitis.
A 55-year old Black woman presented with a nodular scleritis in her right eye (Figure 1A). A scleritis laboratory workup revealed an elevated p-ANCA immunofluorescence pattern and myeloperoxidase. A detailed medical history was remarkable for symptoms suggestive of chronic sinus disease and renal dysfunction. A renal panel confirmed elevated blood urea nitrogen and creatinine levels as well as reduced glomerular filtration, possibly indicating a renal vasculitis.

Referral to rheumatology and nephrology for suspected ANCA vasculitis was initiated. The patient’s nodular scleritis improved with prednisone and methotrexate therapy and topical ophthalmic corticosteroids. Three months after her initial visit, the patient showed complete resolution of her scleritis with a remaining area of scleral translucency (Figure 1C). This feature is best visualized outside the slit lamp with external lighting and can be present in patients with chronic or resolved scleritis.

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A 70-year-old White woman presented with a 6-month history of a unilateral red eye that had been treated unsuccessfully by multiple eye care providers. Her examination revealed a crescent-shaped peripheral ulcer along the limbus (Figure 2A) associated with deep scleral inflammation. This finding is consistent with scleritis and PUK, raising suspicion for a systemic vasculitis.

Laboratory testing revealed elevated c-ANCA and PR3 antibodies. The patient was referred to rheumatology for a probable diagnosis of GPA. Initial response with prednisone and methotrexate resulted in decreased inflammation and corneal epithelialization; however, the patient regressed during prednisone taper. Increasing corneal neovascularization and keratitis with increasing scleral inflammation (Figure 2B) and focal stromal necrosis (Figure 2C) were observed.

Working with rheumatology, the patient was started on rituximab, and she responded favorably. Despite this more aggressive therapy and resolution of her scleritis and PUK, the patient eventually developed a small perforation at the site of the focal stromal melt, requiring a conjunctival patch graft (Figure 2D).

rates of patients with GPA. In particular, rituximab, a monoclonal antibody directed against specific B-cells, is FDA-approved for the treatment of GPA in patients as young as 2 years. Administered as an infusion, rituximab has been shown to be highly successful in achieving steroid-sparing remission in patients with scleritis and PUK associated with GPA.6,7


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