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Images in Rheumatology

Necrotizing scleritis as the initial presentation of granulomatosis with polyangiitis

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A 70-year-old woman presented to the ophthalmology clinic for evaluation of her eye pain and redness and decreased vision in her right eye. She was referred for urgent rheumatological evaluation for suspected necrotizing scleritis. Her past medical history included left eye herpes zoster infection, atrial fibrillation, and chronic bilateral maxillary sinusitis refractory to treatment but improved partially after antrostomy.

On physical examination, she had diffuse anterior scleritis with a well-localized necrotic area of sclera and surrounding inflammation (Figure 1) and bilateral tender maxillary sinuses with no signs of arthritis or skin rash. Further laboratory workup showed an elevated creatinine level of 2.3 (0.5-1.2) mg/dL, with a normal baseline level reported one year ago. Initial urinalysis was negative, but when repeated 1 week later, proteinuria (1+) and 7 red blood cells/high-power field were observed. Serology was positive for anti-proteinase-3 antibody (>8 IU; normal, <1 IU), but it was negative for antinuclear antibody, rheumatoid factor, and hepatitis C antibody. Chest X-ray showed multiple pulmonary nodules (Figure 2), and maxillofacial computed tomography revealed moderate mucosal thickening in the right maxillary sinus and severe mucosal thickening in the left maxillary sinus (Figure 3). Necrotizing scleritis, coupled with recurrent sinusitis, pulmonary nodules, and glomerulonephritis with positive anti-Proteinase 3 supported the diagnosis of granulomatosis with polyangiitis (GPA). Treatment was started with high dose of prednisone (80 mg given orally daily) and rituximab, which reduced her right eye redness and pain but also persistently decreased visual acuity.

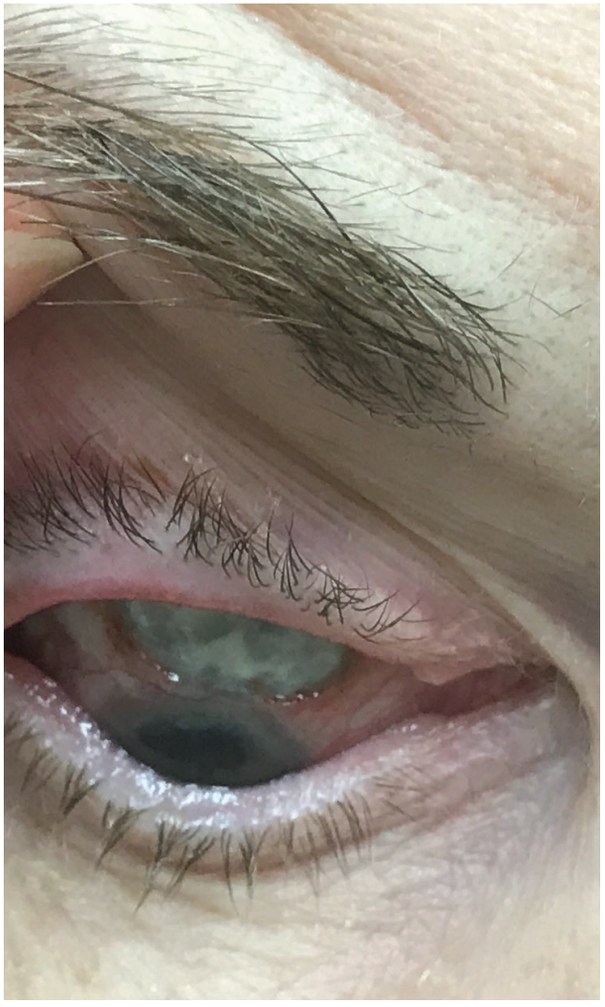

Figure 1. Right-eye necrotizing scleritis.

Scleritis is defined as a painful, potentially blinding scleral inflammation. More than 50% of cases are associated with systemic disease. Necrotizing scleritis is the most severe form of scleritis. It is associated with autoimmune conditions, most notably rheumatoid arthritis, but it is also reported with systemic lupus erythematosus and GPA.[1] Prompt diagnosis and treatment of the underlying condition may halt the progression of both ocular and systemic disease, preventing further organ damage. Treatment of scleritis depends on the severity and underlying systemic illness. In GPA, this includes high-dose

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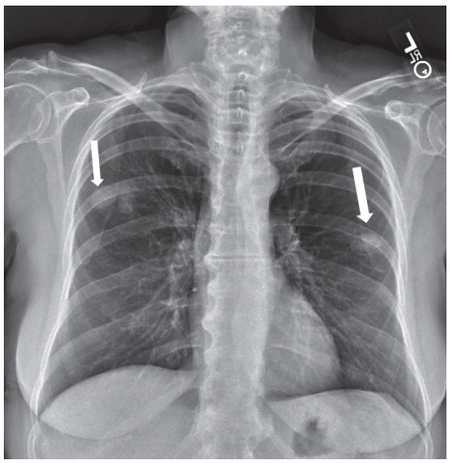

Figure 2. Chest X-ray showing bilateral upper lobe pulmonary nodules (arrows).

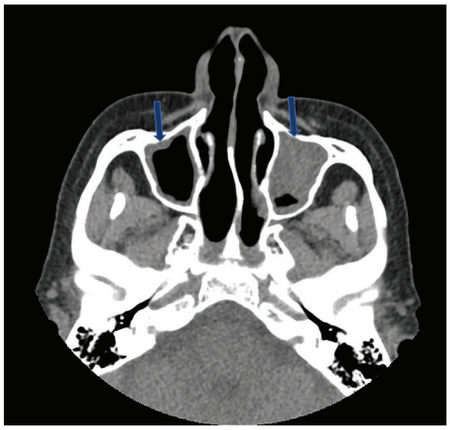

Figure 3. Maxillofacial computed tomography showing moderate right and severe left maxillary sinus mucosal thickening (arrows).

glucocorticoids coupled with either rituximab or cyclophosphamide.[2] Even with the treatment, necrotizing scleritis can have devastating complications, with approximately 75% of the patients experiencing some degree of permanent vision loss and other complications, including keratitis, cataracts, and glaucoma.[3]

Written informed consent was obtained from the patient.

Informed Consent: Written informed consent was obtained from the patient.

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