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### **Scleral inflammatory disease.**

M H Johnson, G J DeFilipp, R A Zimmerman and P J Savino

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# Scleral Inflammatory Disease

Michele H. Johnson<sup>1</sup>  
 Gary J. DeFilipp<sup>2</sup>  
 Robert A. Zimmerman<sup>3</sup>  
 Peter J. Savino<sup>4</sup>

**Scleral inflammatory disease, a relatively common disease of the eye, is demonstrable on CT. Because of its unilateral and sometimes focal involvement, it may be mistaken for more serious diseases, both clinically and by CT. In particular, the clinical difficulties in diagnosing posterior scleritis have led to enucleations when such entities as melanoma and metastasis were suspected. We report the CT appearance and differential diagnosis of scleritis and correlate them with its clinical manifestations, both within the eye and systemically.**

**We found CT to be reliable in differentiating scleritis from true choroidal and retrobulbar masses.**

Scleral inflammatory disease, a relatively common ophthalmologic entity, is demonstrable on CT. Involvement of the sclera and/or episclera is unilateral in the majority of cases; however, bilateral disease may occur [1]. When the inflammation is unilateral and nodular or focal in distribution, it may be mistaken for more serious diseases, such as melanoma or metastasis, both by clinical and CT evaluation [2]. The clinical diagnosis of posterior scleritis is often difficult and has led to enucleation when malignant diseases have been suspected [3].

Scleral inflammatory disease is associated with systemic illnesses in as many as 46% of cases [4]. These include rheumatoid arthritis [5], Wegener's granulomatosis [6-9], sarcoidosis [10], inflammatory bowel disease [11], and a variety of other entities [12]. Scleral involvement may be the initial manifestation of these systemic illnesses.

## Materials and Methods

All five patients were referred for orbital CT from the ophthalmologic services of Temple University Hospital (TUH), Hospital of the University of Pennsylvania (HUP), or Wills Eye Hospital with clinical signs of scleral mass or inflammation. Scans of the orbits were obtained in both axial and coronal planes at 2- to 5-mm intervals. Images were obtained on a Siemens DR3 at TUH (cases 1 and 5), a GE 9800 (case 4), or EMI 5005 (case 3) at HUP, and a GE 8800 at Children's Hospital of Philadelphia (case 2). In all cases scans were obtained with IV contrast agents. Several cases were also scanned without enhancement.

## Results

The clinical and CT data for all five cases are provided in Table 1. Three of the five patients presented initially with unilateral eye symptoms, all of which exhibited signs of anterior and posterior scleral involvement. The other patients (cases 2 and 4) presented with bilateral symptoms and predominantly posterior scleral findings. In all cases of scleritis the entire sclerouveal rim was thickened. In most cases this thickening was shaggy and irregular (Fig. 1). In one case (case 5) the posterior sclera was thicker than the remaining sclerouveal rim (Fig. 2). No cases exhibited

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<sup>1</sup> Department of Diagnostic Imaging, Temple University Hospital, 3401 N. Broad St., Philadelphia, PA 19140. Address reprint requests to M. H. Johnson.

<sup>2</sup> Department of Radiology, University of Michigan, Ann Arbor, MI 48109.

<sup>3</sup> Department of Radiology, Hospital of the University of Pennsylvania, 3400 Spruce St., Philadelphia, PA 19104.

<sup>4</sup> Department of Ophthalmology, Wills Eye Hospital, Ninth and Walnut Sts., Philadelphia, PA 19107.

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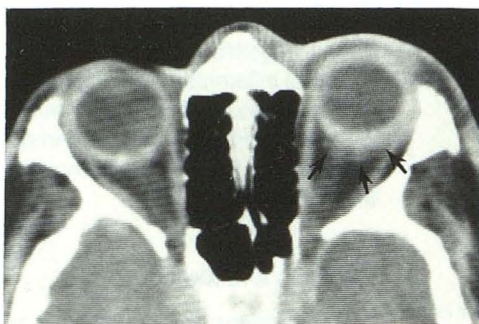
**TABLE 1: CT and Clinical Features of Scleritis**

Case No.	Age	Gender	Clinical Symptoms	CT Appearance	Diagnosis/Treatment	Clinical Course	Systemic Illness
1	8	M	Unilateral proptosis; red, painful eye—posterior scleritis vs retrobulbar mass	Irregular thickening and enhancement of sclerouveal rim, L eye	Anterior and posterior scleritis/oral prednisone	Prompt resolution of symptoms with therapy	None
2	11	M	5 days of L conjunctival injection; R pain; examination bilateral uveitis and disk edema—scleral infection vs inflammation (scleritis)	Preenhancement: irregular thickening of sclerouveal rim bilaterally; postenhancement: marked enhancement of thick sclerouveal rim bilaterally	Bilateral anterior and posterior scleritis/oral prednisone	Resolution of pain within hours	None
3	13	F	Red, painful eye; proptosis—scleritis vs retrobulbar mass	Irregular thickening and enhancement of sclerouveal rim	Anterior and posterior scleritis	Prompt symptom relief with therapy	None
4	31	F	Bilateral eye pain; decreased visual acuity—clinical diagnosis: posterior scleritis; fluorescein angiography and sonography consistent	Bilateral diffuse, smooth sclerouveal rim; thickening and enhancement	Bilateral posterior scleritis/oral corticosteroids	Initial prompt resolution of symptoms with therapy; relapse over past 8 months with anterior and posterior scleritis and acute glaucoma	None found to date
5	69	M	L proptosis, painful eye anteriorly and posteriorly—scleritis vs retrobulbar mass	L proptosis with irregular thickening and enhancement of sclerouveal rim, especially posteriorly	Posterior scleritis/oral prednisone	Prompt resolution of symptoms with therapy; 3 months after therapy scleritis developed in R eye	None noted to date

Note.—L = left; R = right.



1



2

Fig. 1.—Case 1: unilateral scleritis. Contrast-enhanced CT scan through orbits exhibits typical shaggy sclerouveal rim seen in scleritis.

Fig. 2.—Case 5: unilateral scleritis with focal posterior involvement. Contrast-enhanced CT scan through orbits shows shaggy sclerouveal rim thickening, most marked in posterior sclera (arrows).

abnormalities of the extraocular muscles, lacrimal glands, optic nerves, or orbital apices. No masses were seen. Overlying soft-tissue swelling was not a feature of scleritis in these cases. Despite the large percentage of scleritis cases associated with systemic illness in the literature (46%) [4], none of our patients have had documented associated illness to date. However, two patients (cases 4 and 5) had relapsing courses after discontinuance of steroid therapy. The glaucoma seen in one case (case 4) may be a complication of scleritis itself or of the corticosteroid treatment [13].

## Discussion

Scleritis and episcleritis refer to nonpyogenic inflammatory disease involving the outer coats of the eye [14]. The cornea forms the outer surface of the anterior one-sixth of the globe while the sclera covers the posterior five-sixths. The sclera is composed of three parts. The episclera is the vascular outermost layer of the sclera. It is most prominent anterior to the equator of the eye and becomes quite thin posterior to the equator. The scleral stroma is a relatively avascular composite



of collagen fibers in a glucoaminoglycan matrix. Posteriorly, these stromal fibers blend into the dura mater surrounding the optic nerve. The lamina fusca is the innermost layer of the sclera and provides an interface between the sclera and the very vascular choroid. The scleral stroma derives its vascular supply predominantly from the episcleral and choroidal vessels. Thus, the sclera can be divided into a more vascularized portion anterior to the equator (anterior sclera) and a relatively avascular portion posterior to the equator (posterior sclera) (Fig. 3).

Isolated episcleritis is generally a benign, self-limited process characterized by eye pain, redness, and photophobia. It lasts 7–21 days and frequently follows a viral illness. Pathologically, the episclera is hyperemic and edematous, but the underlying sclera is spared [4]. These patients seldom come to the attention of a radiologist.

Scleritis is characterized by edema of the sclera and episclera along with hyperemia of the episclera. The anterior sclera is most commonly involved in scleral inflammatory disease (90%) [1, 4]. The clinical diagnosis of scleral inflammation anterior to the equator is readily made; however, the clinical diagnosis of inflammation posterior to the equator is much more difficult. Posterior scleritis is most often seen in association with anterior scleritis, but may occur in isolation. Alterations in the vascular supply to the sclera by the inflammatory process ultimately lead to focal or diffuse scleral ischemia. The anterior sclera, by virtue of its ample episcleral

and choroidal vascular networks, is more resistant to the complications of ischemia than is the less vascular sclera posterior to the equator. When the posterior sclera is inflamed, ischemia often results in scleral thinning, necrosis, and perforation. Focal, nodular, and diffuse scleral inflammation may occur in either compartment.

Exudation may occur in posterior scleritis. Exudation may result in uveal effusion, retinal detachments, and/or proptosis. Posterior scleritis, particularly when focal and unilateral, may mimic choroidal or retrobulbar mass lesions [2, 3]. Metastatic disease to the choroid [15] (Fig. 4), melanoma (Fig. 5), and vascular malformations (Fig. 6) may all look clinically similar to scleritis. CT scans of these other entities demonstrate focal mass lesions of the sclerouveal rim that are variably enhancing; however, the uniform thickening and enhancement of the sclerouveal rim demonstrated in our cases of scleritis is not seen. True retrobulbar mass lesions causing secondary scleral edema are readily diagnosed by CT (Fig. 7).

Complications of scleritis [1] include scleral thinning and ischemic necrosis, secondary glaucoma [13], retinal detachment [16], disk edema, uveitis, and uveal effusion. Severe pain and decreased visual acuity may be present in the acute stages. *Scleromalacia perforans* is a term applied to a severe form of scleritis with perforation of the sclera secondary to ischemic necrosis. In our patient group, disk edema and uveitis were seen in case 2, and secondary glaucoma was seen in case 4.

Fig. 3.—Scleral anatomy.

A, Drawing of globe shows multiple layers enclosing vitreous, seen as sclerouveal rim on CT.

B, Normal contrast-enhanced CT scan of orbits shows sclerouveal rim (arrow).

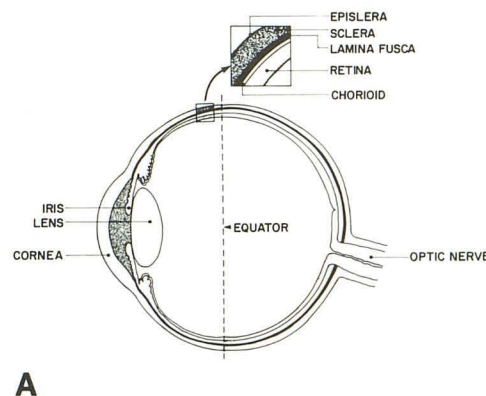
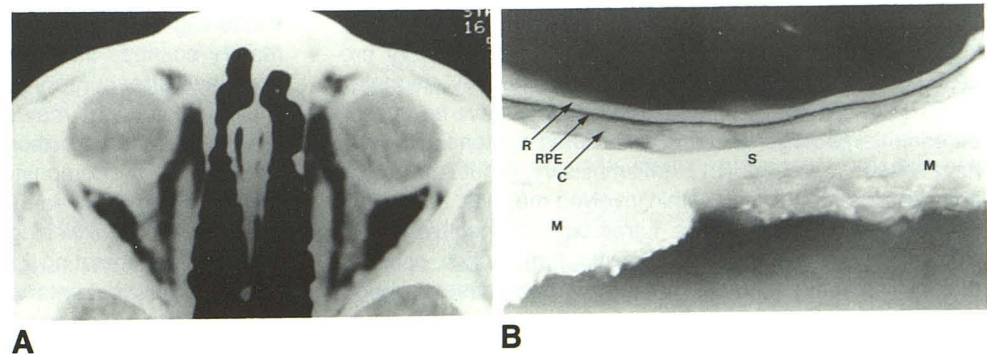


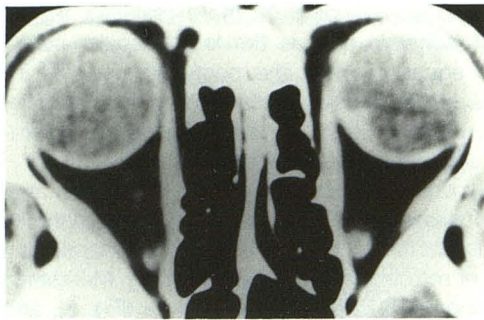
Fig. 4.—Scleral metastasis from lung.

A, Contrast-enhanced CT scan shows focal sclerouveal rim thickening posterior to equator in patient with lung carcinoma.

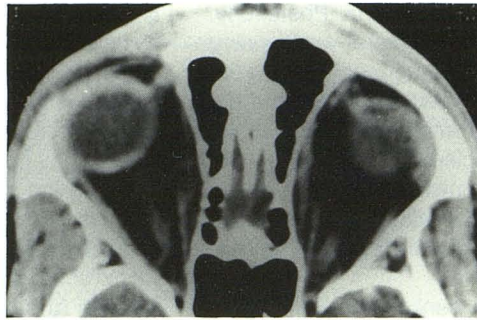
B, Axial pathologic section through globe shows fluffy tumor implants along posterior scleral margin. R = retina; RPE = retinal pigment epithelium; C = choroid, abnormally thickened; S = sclera; M = metastatic deposits.







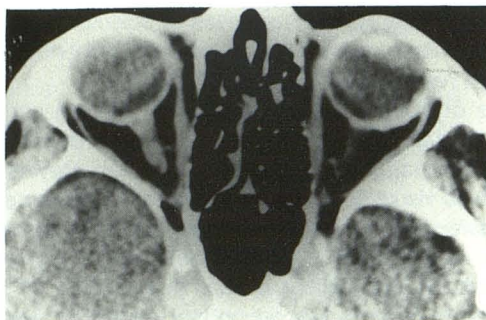
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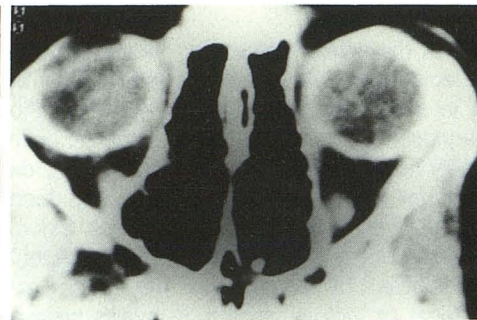
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Fig. 5.—Choroidal melanoma in 46-year-old man. Focal enhancing lesion involving sclerouveal rim posteriorly extends into vitreous.

Fig. 6.—Hemangioma. Focal posterior sclerouveal rim enhancement is from vascular malformation of retina and choroid.



A



B

Fig. 7.—Metastatic disease vs scleritis.

A, Patient with lung cancer. Sclerouveal rims are studded bilaterally with implants characterized by focal thickening and contrast enhancement.

B, Case 2. Diffuse, shaggy thickening and enhancement of sclerouveal rims are typical of bilateral scleritis.

The association of scleral inflammatory disease with systemic illnesses is common, particularly in the collagen vascular disorders [17]. In a study by Watson and Hayreh [1], 21 of 89 patients with associated systemic illnesses and scleritis had rheumatoid arthritis. Scleritis in these cases may be nodular or diffuse in nature and is pathologically similar to typical subcutaneous rheumatoid nodules. Systemic lupus erythematosus, periarteritis nodosa, IgA nephropathy, inflammatory bowel disease, and gout have all been reported in association with scleritis. In some cases, scleritis may be the initial presentation of systemic illness [17].

Ocular involvement with Wegener's granulomatosis [6–9] occurs in 40–50% of patients with the generalized form. Manifestations may include granulomatous inflammatory masses (22%), orbital cellulitis, scleritis/episcleritis (12%), vasculitic-ischemic injury to the retina or optic nerve (15%), and nasolacrimal duct obstruction (6%) [8] (Fig. 8).

Orbital pseudotumor is a nonpyogenic inflammatory process involving the orbital contents and commonly diagnosed by CT [18–20] (Fig. 9). Immune mechanisms have been implicated in its pathogenesis and associated systemic diseases have been described [21]. Inflammatory cellular infiltrates have been demonstrated within involved muscles, sclera, and episclera as well as in the intraorbital connective tissue, forming focal tissue masses. Myositic, tumefactive, episcleritic, and infiltrative (diffuse) forms have been described both alone and in combination. Prompt clinical and radiologic response to systemic corticosteroid therapy is the rule. Isolated scleral inflammation, as seen in our patients, characterized by

thickening and enhancement of the sclerouveal rim, probably represents a type of orbital pseudotumor. Bernardino et al. [18] reviewed 350 normal patients and patients with orbital pseudotumor. In the normal patients, the sclerouveal rim was 3.2–4 mm thick. In the patients with pseudotumor, the sclerouveal rim was 1.5–3.5 times thicker than the normal. In three of eight patients with sclerouveal rim thickening and orbital pseudotumor studied by Bernardino et al., the scleral changes were the predominant feature. Assessment of other orbital structures involved by orbital pseudotumor is readily accomplished by CT.

Sclerouveal rim thickening on CT and scleral inflammation need not always be scleritic or caused by pseudotumor. Thick CT sections through the globe may result in volume averaging of the sclerouveal rim and the scleral surface of the globe, which is parallel to the plane of section. This causes apparent thickening of the sclerouveal rim in a normal patient. Inflammatory changes after trauma (especially foreign-body reactions) and after surgery exhibit similar CT findings [18].

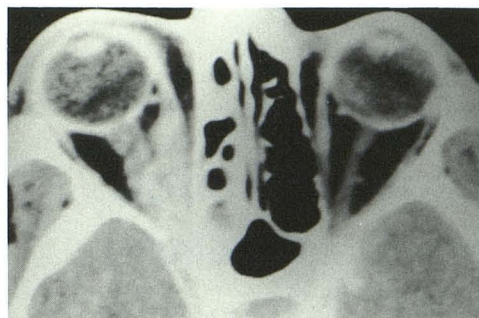
Orbital infections may also involve the sclera; however, in the vast majority of these cases other evidence of orbital cellulitis will be demonstrated both clinically and by CT. A careful history is needed when evaluating a globe showing sclerouveal rim thickening on CT.

Scleral inflammation is a common ophthalmologic entity; however, only patients with unclear clinical and ophthalmologic findings are referred for CT.

In our series, scleritis is recognized as diffuse scleral thickening and enhancement, in contrast to other scleral and uveal

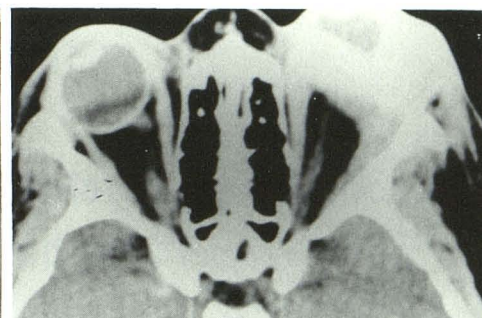


Fig. 8.—Wegener's granulomatosis in 47-year-old woman. Proptosis of right globe, sclerouveal rim thickening, and medial rectus thickening and enhancement. Note ipsilateral ethmoid sinus involvement.



8

Fig. 9.—Left orbital pseudotumor, extreme case. Marked proptosis, thickening of sclerouveal rim with encroachment into vitreous, involvement of extraocular muscle and lacrimal gland.



9

Fig. 10.—Scleritis vs metastatic disease.

A, Case 3: 13-year-old girl with scleritis has diffuse sclerouveal rim thickening.

B, Metastatic disease to sclera from primary lung carcinoma does not confine itself to sclerouveal rim but extends into vitreous and is more focal in its involvement.



A



B

disease processes such as choroidal melanoma or metastatic disease, which either involve the sclera locally or extend beyond the sclera into adjacent structures (Fig. 10). CT is excellent for characterizing true choroidal and retrobulbar masses and for differentiating them from the clinically similar exudative collections of posterior scleritis. An understanding of the scleral anatomy and pathologic features of scleritis should facilitate its recognition on CT and allow the exclusion of more ominous disease processes from the differential diagnosis.

## REFERENCES

1. Watson PG, Hayreh SS. Scleritis and episcleritis. *Br J Ophthalmol* 1976;60:163-191
2. Benson WE, Shields JA, Tasman W, Crandall AS. Posterior scleritis: a cause of diagnostic confusion. *Arch Ophthalmol* 1979;97:1482-1486
3. Fraunfelder FT, Watson PG. Evaluation of eyes enucleated for scleritis. *Br J Ophthalmol* 1976;60:227
4. Watson PG. The diagnosis and management of scleritis. *Ophthalmology* 1980;87:716-720
5. Barr CC, Davis H, Culbertson WW. Rheumatoid scleritis. *Ophthalmology* 1981;88:1269-1273
6. Vermess M, Haynes BF, Fauci AS, Wolff SM. Computer assisted tomography of orbital lesions in Wegener's granulomatosis. *J Comput Assist Tomogr* 1978;2:45-48
7. Spalton DJ, Graham EM, Page NGR, Sanders MD. Ocular changes in limited forms of Wegener's granulomatosis. *Br J Ophthalmol* 1981;65:553-563
8. Haynes BF, Fishman ML, Fauci AS, Wolff SM. The ocular manifestations of Wegener's granulomatosis: fifteen years experience and review of the literature. *Am J Med* 1977;63:131-141
9. Straatsma BR. Ocular manifestations of Wegener's granulomatosis. *Am J Ophthalmol* 1957;44:789-799
10. Henkind P. Sarcoidosis: an expanding ophthalmic horizon. *J R Soc Med* 1982;75:153-159
11. Petrelli EA, McKinley M, Troncale FJ. Ocular manifestations of inflammatory bowel disease. *Ann Ophthalmol* 1982;14(4):356-360
12. Nomoto Y, Sakai H, Endoh M, Tomino Y. Scleritis and IgA nephropathy. *Arch Intern Med* 1980;140:783-785
13. Wilhelmus KR, Grierson I, Watson PG. Histopathologic and clinical associations of scleritis and glaucoma. *Am J Ophthalmol* 1981;91(6):697-705
14. Cobo M. Inflammation of the sclera. *Int Ophthalmol Clin* 1983;23(1):159-171
15. Yeo JH, Jakobiec FA, Iwamoto T, Brown R, Harrison W. Metastatic carcinoma masquerading as scleritis. *Ophthalmology* 1983;90(2):184-194
16. Berger B, Reeser F. Retinal pigment epithelial detachments in posterior scleritis. *Am J Ophthalmol* 1980;90:604-606
17. Watson PG, Hazleman BL. *Major problems in ophthalmology*, vol 2. The sclera and systemic disorders. Philadelphia: Saunders, 1976
18. Bernardino ME, Zimmerman RD, Citrin CM, Davis CO. Scleral thickening: a CT sign of orbital pseudotumor. *AJR* 1977;129:703-706
19. Harr DL, Quencer RM, Abrams GW. Computed tomography and ultrasound in the evaluation of orbital infection and pseudotumor. *Radiology* 1982;142:395-401
20. Trokel SL. Computed tomographic scanning of orbital inflammations. *Int Ophthalmol Clin* 1982;22(4):81-98
21. Blodi FC, Gass JDM. Inflammatory pseudotumor of the orbit. *Br J Ophthalmol* 1968;52:79-92