Posterior Scleritis

Clinical Features, Systemic Associations, and Outcome in a Large Series of Patients

Peter J. McCluskey, FRACO,[1] Peter G. Watson, FRCOphth,[1,2] Susan Lightman, FRCOphth,[1] John Haybittle, PhD,[3] Marie Restori, MSc,[1] Michael Branley, FRACO[1]

Objective: To document the clinical features, systemic associations, and visual outcome in a large number of patients with posterior scleritis.

Design: Retrospective, noncomparative case series.

Participants: There were 137 patient records showing patients with a diagnosis of posterior scleritis who were attending or had attended the scleritis clinic at Moorfields Eye Hospital between 1974 and 1996. Ninety-nine records were suitable for detailed analysis.

Methods: The medical records and B-mode ultrasound examinations were reviewed.

Main Outcome Measures: The clinical features, systemic associations, treatment, and outcome of each patient were determined.

Results: Posterior scleritis occurred at all ages. The mean age at onset was 49.3 years. Posterior scleritis began before age 40 in 30% of patients and was twice as common in women as in men. The B-mode ultrasound examination showed diffuse and nodular changes in the posterior sclera. Necrotizing posterior scleritis was not identified. Twenty-nine percent of patients had an associated systemic disease that included systemic vasculidites, autoimmune diseases, and lymphoma. Such patients more commonly had nodular changes on B-mode ultrasound examination. Early treatment controlled posterior scleral inflammation and limited visual loss. Thirty-one percent of patients lost two or more lines of vision. Statistical analysis revealed that patients older than age 50 had an increased risk of having an associated systemic disease and were more likely to experience visual loss. Patients with associated systemic disease required more aggressive immunosuppressive therapy and more frequently had accompanying anterior scleritis. There was no association between unilateral, bilateral, or recurrent disease and the presence of systemic disease or visual loss from posterior scleritis.

Conclusions: The B-mode ultrasound examination reveals that posterior scleritis occurs far more often than previously thought and can lead to rapid and permanent visual loss. All patients with posterior scleritis must be assumed to be at risk of visual loss. Forty percent of patients had no anterior scleral inflammation, and 9% had no detectable physical signs. All patients need to be investigated for an associated systemic disease and all require early treatment to minimize loss of vision. *Ophthalmology 1999;106:2380–2386*

Posterior scleritis is an uncommon and under-recognized form of scleral inflammation. Because of its low incidence, it has been difficult to ascertain the clinical features, systemic associations, and outcome of this disease. It may present with a range of clinical findings, and its clinical features may be confused with those of intraocular inflammation, ocular tumors, and orbital inflammation.[1–3] Posterior scleritis may be idiopathic or may be associated with systemic diseases such as rheumatoid arthritis and Wegener granulomatosis.

Increasing awareness that unexplained posterior segment and orbital inflammation might be caused by posterior scleritis and the more widespread availability of B-mode ultrasound examination has led to an increase in the number of patients treated by the Scleritis Clinic at Moorfields Eye Hospital. These patients have been predominantly young or middle-aged adults who have presented with severe symptoms and profound visual loss without associated systemic disease. The increased number of patients with this form of scleritis prompted the current study, which was performed to more fully characterize the systemic associations, clinical features, treatment, and outcome of the large group of patients with posterior scleritis attending the Scleritis Clinic at Moorfields Eye Hospital, London, England.

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[1] Department of Clinical Ophthalmology, Institute of Ophthalmology and Moorfields Eye Hospital, London, England.

[2] Addenbrookes Hospital, Cambridge, England.

[3] MRC Cancer Trials Office, Cambridge, England.

Address correspondence to Peter J. McCluskey, FRACO, Diseases and Surgery of the Eye, 357 Beamish Street, Campsie NSW 2194, Australia.

Table 1. Ultrasonographic Classification of Posterior Scleritis*

Ultrasonographic Abnormality	Classification
Increased eye wall thickness	Diffuse
Scleral nodules	Nodular

* The upper limit of posterior scleral wall thickness varies slightly. For this study, eye wall thickness of greater than 2.0 mm was defined as increased eye wall thickness. Any of the following additional ultrasonographic signs may be seen in either diffuse or nodular posterior scleritis: fluid in Tenon's capsule, swelling of the optic disc, distended optic nerve sheath, and retinal detachment.

Patients and Methods

A retrospective review of the medical records of patients attending the Scleritis Clinic at Moorfields Eye Hospital between 1974 and 1996 was performed to identify all patient records with a patient diagnosis of posterior scleritis. Of the 137 records identified, 38 were excluded because of insufficient clinical detail to ensure a correct diagnosis or lack of satisfactory ultrasonography data. The first patients in this series had ultrasonography with early equipment. The resolution of some of these images was so poor that it was impossible to be certain of the diagnosis in some of these patients, and they were excluded from the analysis. Ninety-nine medical records were therefore suitable for inclusion in the study. Detailed information regarding the onset, clinical features, ultrasound features, systemic associations, treatment, complications, and outcome was recorded. At the time of the study, 54 patients continued to be followed in the clinic, and 18 patients were being treated for active disease. The patient data were collected over two 6-month periods in 1994 and 1996. The final date of study for patients included in this review was December 31, 1996.

In this study, the patients were classified into two groups using the ultrasonographic features found at the time of their first B-mode ultrasound examination. Table 1 summarizes the ultrasonographic abnormalities used in this classification.

To look for associations between the various clinical features, two-way or multiway tables were constructed and tested by chi-square analysis with Yates correction in the 2×2 tables. A P value of 0.05 or less was considered statistically significant. The numbers given after values of odds ratios (OR) are 95% confidence limits. Forward stepwise logistic regression was undertaken to determine whether any of the recorded variables had predictive value for patient outcome in terms of visual loss.

Results

The main characteristics of the patients studied are given in Table 2. There were 64 female and 35 male patients, with an age range of 11 to 84 years and a mean age of 49.3 years. All ethnic groups were represented. Posterior scleritis occurred in 40 patients without associated anterior scleritis and occurred with anterior scleritis at the time of presentation in 36 patients. Anterior scleritis occurred at some time during the period of follow-up in 59 of the 99 patients. Unilateral involvement occurred in 64 patients and bilateral disease in 35 patients. At the time of the study, 18 patients had active disease and were taking medication, in 4 patients the disease state was uncertain, and in the remaining 77 patients, the scleritis was inactive. Fifty patients had a single episode of posterior scleritis, and 49 patients had had multiple episodes of posterior scleritis.

Table 2. Characteristics of Patients with Posterior Scleritis

Characteristic	%	95% Confidence Limits
Mean age (yrs)	49.3	46.0–52.6
Male	35.4	25.8–44.8
Unilateral	64.6	55.2–74.0
Anterior scleritis at presentation	36.4	26.9–45.9
Anterior scleritis at some time	59.6	49.9–69.3
Idiopathic posterior scleritis	70.7	61.7–79.7
Associated systemic disease	28.3	19.4–37.2
Visual loss	31.3	22.2–40.4

Table 3. Systemic Associations Seen in Patients with Scleritis

Systemic disease	No. of Patients
Rheumatoid arthritis	5
Systemic vasculitis	4
Wegener granulomatosis	4
Vogt–Koyanagi–Harada disease	2
Relapsing polychondritis	3
Thyroid disease	2
Sarcoidosis	2
Primary biliary cirrhosis	1
Systemic lupus erythematosus	1
Multiple myeloma	1
Lymphoma	1
Carcinoma of pancreas	1
Ankylosing spondylitis	1
Polyarteritis nodosa	1

Idiopathic posterior scleritis was seen in 68 patients, and there was an associated systemic disease in 29 patients. The systemic disease associations are detailed in Table 3. Two patients developed the syndrome of surgically induced scleritis involving the posterior segment after retinal detachment surgery complicated by infected scleral buckles.[4]

Periocular pain and headache, visual loss, and symptoms associated with anterior scleritis were the common symptoms of patients with posterior scleritis, with 55 patients reporting pain, 31 reporting reduced vision, and 59 reporting symptoms related to anterior scleritis. Physical signs of posterior scleritis varied, and there were no signs in 17 patients. A serous retinal detachment was the most frequent abnormality among the range of clinical signs that occurred. The physical signs seen in patients with posterior scleritis are summarized in Table 4. There was no correlation between any physical sign and the type of posterior scleritis.

Table 4. Signs of Posterior Scleritis

Physical Sign	No. (%) of Patients
Associated anterior scleritis	34 (34)
Serous retinal detachment	21 (21)
Swollen optic disc	18 (18)
No abnormalities	17 (17)
Subretinal localized granuloma	13 (13)
Elevated intraocular pressure	12 (12)
Choroidal effusion	4 (4)
Uveitis*	2 (2)
Retinal vasculitis	2 (2)
RPE changes	2 (2)

RPE = retinal pigment epithelium.

* Uveitis is defined as cells in the anterior chamber, posterior chamber, or both chambers.

Table 5. Visual Outcome in Patients with Posterior Scleritis

Loss of Visual Acuity	No. of Patients
No loss of vision	68
Visual loss	31
6/12–6/18	7
6/24–6/60	10
Counting fingers only	5
Hand motions only	5
Perception of light	1
Blindness	3

Loss of vision is the most important complication of posterior scleritis. Loss of vision of 2 or more lines of visual acuity occurred in 31 (31%) of the 99 patients. Permanent loss of vision occurred in only one eye in almost all of these patients, but three patients became legally blind as a result of posterior scleritis. The visual outcome data are summarized in Table 5. Most patients lost vision from either macular changes (17 patients) or optic atrophy related to posterior scleritis (13 patients). A number of abnormalities were seen involving the macula. Retinal pigment epithelial changes were the most common change seen. Epiretinal membrane formation, macular edema, and macular cyst/hole were also observed in patients in this study. Cataract (four patients) and retinal detachment (three patients) also resulted in loss of vision. Multiple causes of visual loss were often present in patients with severe disease.

Statistical analysis of the results revealed that patients older than 50 years had an increased risk of associated systemic disease (OR = 3.1: 1.2–7.7) and that this disease was most frequently a connective tissue disease. There was no association between unilateral, bilateral, or recurrent posterior scleritis and the presence of systemic disease or development of visual loss from posterior scleritis. Patients older than 50 years were also more likely to experience visual loss (OR = 4.6: 1.6–10.1). Patients who required treatment with systemic corticosteroids and additional immunosuppressive drugs tended to have a higher risk of visual loss, but the association was not statistically significant (OR = 3.0: 0.1–11.1). Logistic regression with visual loss as the dependent variable showed that when age was included, no other factor was a significant predictor of visual loss. Posterior scleritis associated with an identifiable systemic disease is associated with the need for more aggressive immunosuppressive therapy (OR = 5.3: 2.0–14.0). Posterior scleritis associated with anterior scleritis has a higher risk of associated systemic disease (OR = 3.4: 1.2–9.3).

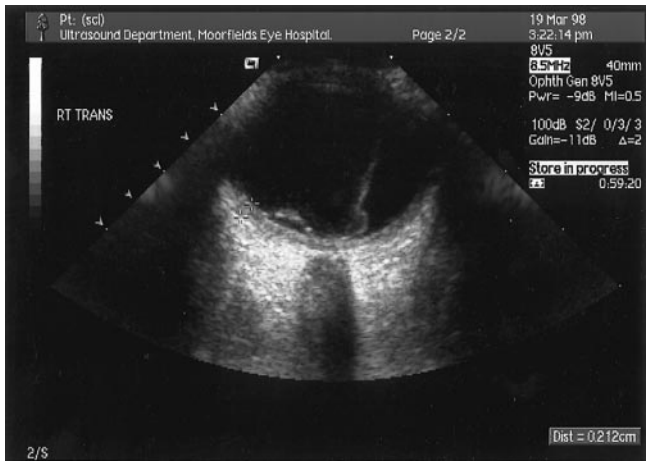

Figure 2. Transverse section B-mode ultrasound scan from a 50-year-old white man showing diffuse thickening of the posterior coats of the eye accompanied by an exudative retinal detachment.

Discussion

In this study, 137 medical records of patients with posterior scleritis were reviewed. Ninety-nine patients seen over a 22-year period who had been followed clinically and with B-mode ultrasonography were selected and analyzed in detail. The age range of patients was 11 to 84 years with a mean age of 49.3 years. Posterior scleritis began before age 40 in 30% of patients and was twice as common in women as in men. The majority of patients had unilateral disease, but 35% had bilateral involvement. The frequency of posterior scleritis in young adults and children was also higher than expected, although posterior scleritis has been recognized in children as young as age 5.[5,6] Patients older than age 50 had a higher risk of having an associated systemic disease and of experiencing visual loss. There was no as-

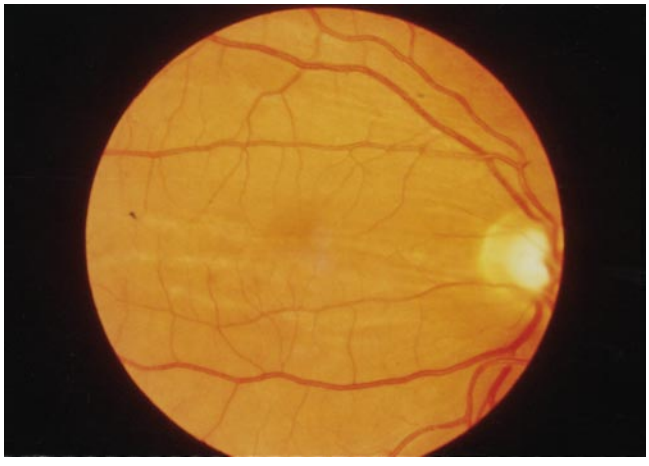

Figure 1. Choroidal folds in a 63-year-old white woman induced by posterior scleritis. The presenting feature was reduced vision. The ultrasound appearances were similar to those seen in Figures 7 and 8.

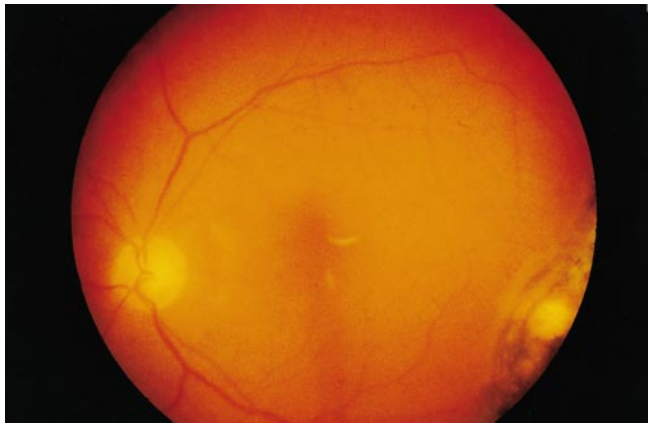

Figure 3. A peripheral visible subretinal mass seen in a 60-year-old white woman because of a large peripheral scleral nodule similar to that in Figure 4. Nodules do not always produce fundus lesions, but when the choroid is involved in the inflammatory process, this is the typical appearance.

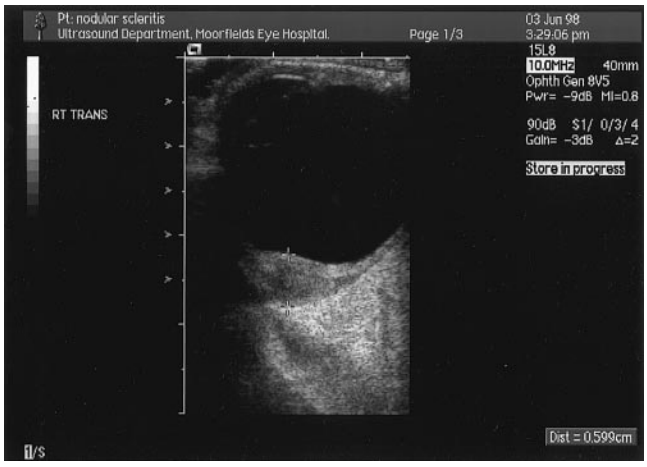

Figure 4. Transverse B-mode image of the right eye of a 50-year-old white man deviated temporally. There is a large posterior temporal scleral nodule that produced a similar fundus lesion to that seen in Figure 3.

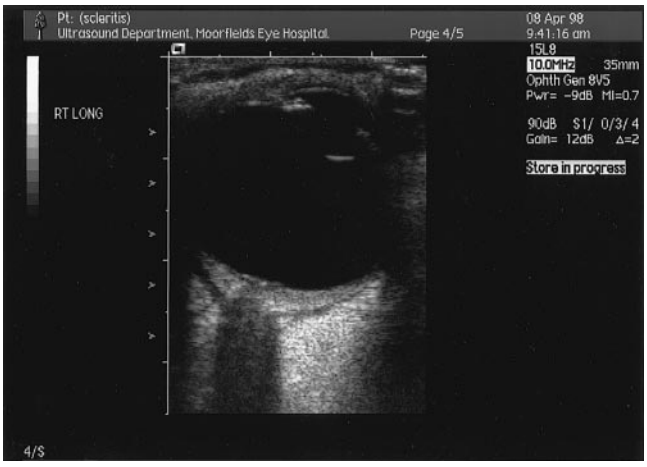

Figure 6. Longitudinal B-mode ultrasound scan showing diffuse thickening of the posterior coats of the eye (2.5 mm; normal, 0.8–2.0 mm) together with fluid in the Tenon capsule and the nerve sheath.

sociation between unilateral, bilateral, or recurrent disease and the presence of systemic disease or visual loss.

Thirty-six percent of the patients presented with combined anterior and posterior scleritis, and 59% developed clinically apparent anterior scleritis at some time during the period of follow-up. This incidence of posterior scleritis combined with anterior scleritis is far higher than previously documented, illustrates the presumed underdiagnosis of posterior scleritis in the past, and highlights the value of sensitive B-mode ultrasonography.[7] Patients who developed anterior scleritis had a higher risk of having an associated systemic disease.

All patients in this series were symptomatic or presented with symptoms or signs that led to a provisional diagnosis of posterior scleritis, which was confirmed with B-mode ultrasonography. The common presentations of patients with posterior scleritis were periocular pain and headache with or without reduced vision and anterior scleritis combined with posterior scleritis. Periocular pain, pain on movement, and decreased vision were the most common symptoms. Physical signs were diverse and variable with no abnormal physical signs in 17% of patients. In this series, signs of anterior scleritis, a swollen optic disc, choroidal folds (Fig 1), serous retinal detachment (Fig 2), visible subretinal mass lesion (Figs 3,4), and elevated intraocular pressure were the most common physical signs (Fig 5). The symptoms and signs depend on the location and severity of the inflammation, its extent, and its relationship to surrounding structures such as the choroid, retinal pigment epithelium, and extraocular muscles. Peripheral inflammation deep in the sclera produces uveal effusions and secondary angle-closure glaucoma (Fig 5).[2,8] Localized nodular inflammation may produce circumscribed subretinal mass lesions and overlying retinal detachment (Figs 2–4). Inflammation surrounding the optic disc or macula results in visual loss, optic disc edema, retinal folds, macular exudation, or

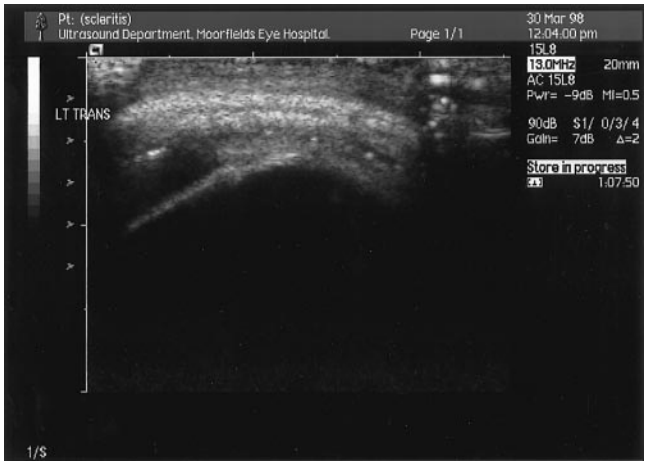

Figure 5. Transverse B-mode ultrasound image (gaze deviated nasally) from a 55-year-old black woman showing gross thickening of the anterotemporal sclera. The thickening extends into the posterior sclera. This patient had an elevated intraocular pressure that returned to normal after the scleritis was treated.

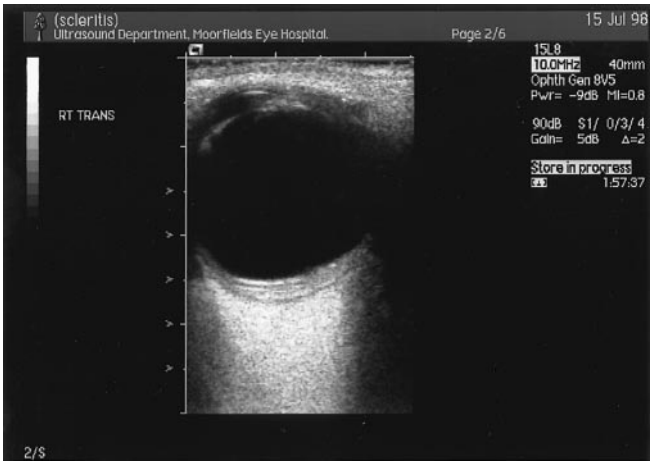

Figure 7. Transverse B-mode section from a 53-year-old white woman with anterior and posterior scleritis showing the striated appearance associated with choroidal folds similar to those seen in Figure 1. There is also fluid in the Tenon capsule.

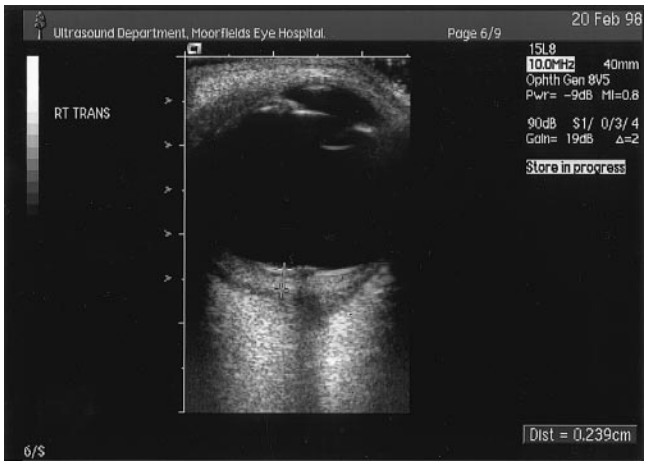

Figure 8. Transverse B-mode image from a 15-year-old black girl patient before treatment. The coats of the eye are thickened at 2.4 mm. The disc is swollen, and there is fluid in the Tenon capsule.

retinal pigment epithelial defects (Fig 1). Low-grade uveitis is uncommon in posterior scleritis and was seen in only 2% of the patients in this series; severe uveitis is not a feature of any type of scleritis and, if present, the diagnosis of scleritis should be reconsidered. The diagnosis of posterior scleritis is often one of exclusion as 17% of the patients had no detectable physical signs of the disease.

Ultrasonography is the key investigation necessary to make the diagnosis of posterior scleritis and in this study documented a range of abnormalities in patients with posterior scleritis including increased thickness of the ocular coats (Figs 2, 6–9), fluid in the Tenon capsule (episcleral space) (Figs 6), swelling of the optic disc, distended optic nerve sheath (Figs 1, 6), retinal detachment, and scleral nodules (Figs 2–4). Eyewall thickness of greater than 2.0 mm was considered abnormal. It has been possible with ultrasonography to confirm that the clinically recognized forms of diffuse anterior scleritis and nodular anterior scleritis are reproduced in the posterior segment. It is known from histopathologic studies that necrotizing posterior scleritis does rarely occur, but at this time, ultrasonography is unable to detect the early changes of necrotizing scleritis such as local ischemia and scleral thinning.[9] Scleral thinning was detected in only one B-mode examination during this study. Follow-up data from ultrasound performed after the first visit were incomplete and not included in this study. However, study of the available data shows that the clinical response to treatment and the ultrasound appearances do not always correlate. In some patients, there is rapid, complete resolution of the ultrasound abnormalities, but in other patients, residual mass or swelling remains for a considerable period after all symptoms and signs have resolved (data not shown and unpublished observations). Fluorescein angiography and CT scanning may add useful additional information in selected patients (Figs 10 and 11), but in this series, CT scanning was performed only if the ultrasound images were unsatisfactory or to confirm the diagnosis in difficult cases.[10] The CT scanning has no advantages over B-mode ultrasonography in this condition. Similarly, fluo-

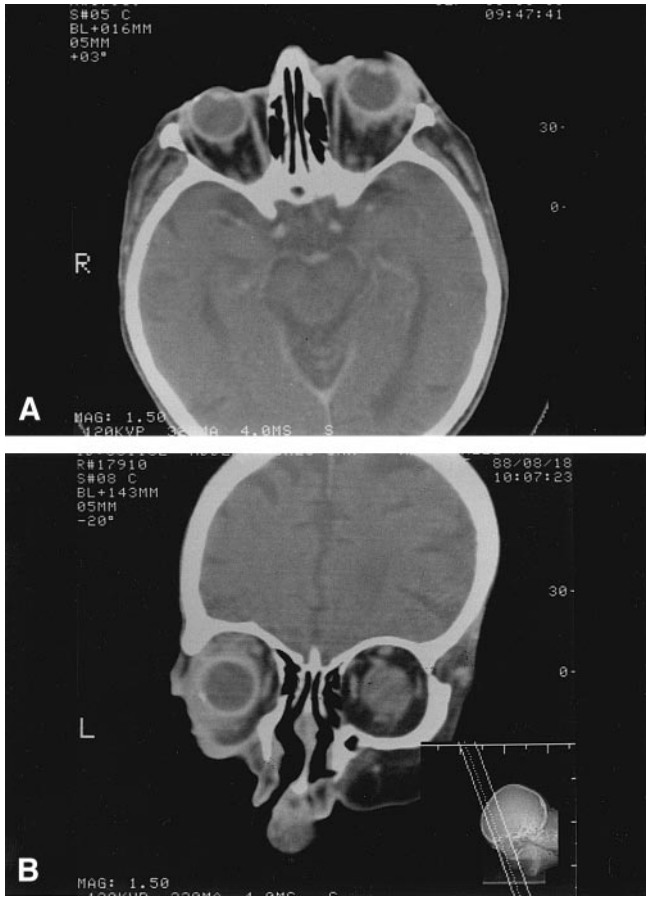

Figure 10. A, axial computerized tomography of a 22-year-old white man who presented with severe proptosis, reduction of vision, and slight swelling of the optic disc. The scan shows proptosis, scleral thickening, and diffuse opacification within the orbit. The B scan confirmed the scleral thickening but did not show any orbital infiltration. **B,** coronal computerized tomography of the same patient. The scan shows proptosis, scleral thickening, and diffuse opacification within the orbit.

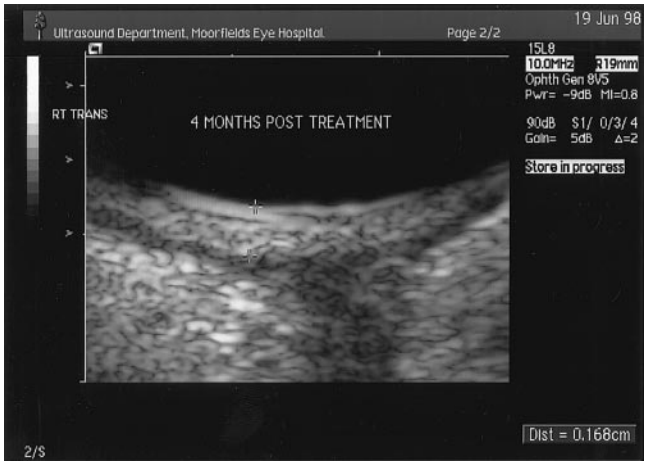

Figure 9. The same eye as Figure 8 of patient 4 months after treatment with systemic corticosteroids. At the time this zoomed image was taken, she was receiving no treatment, and vision had returned to normal. There is some residual fluid in the Tenon capsule, but the thickness of the coats was now 1.7 mm.

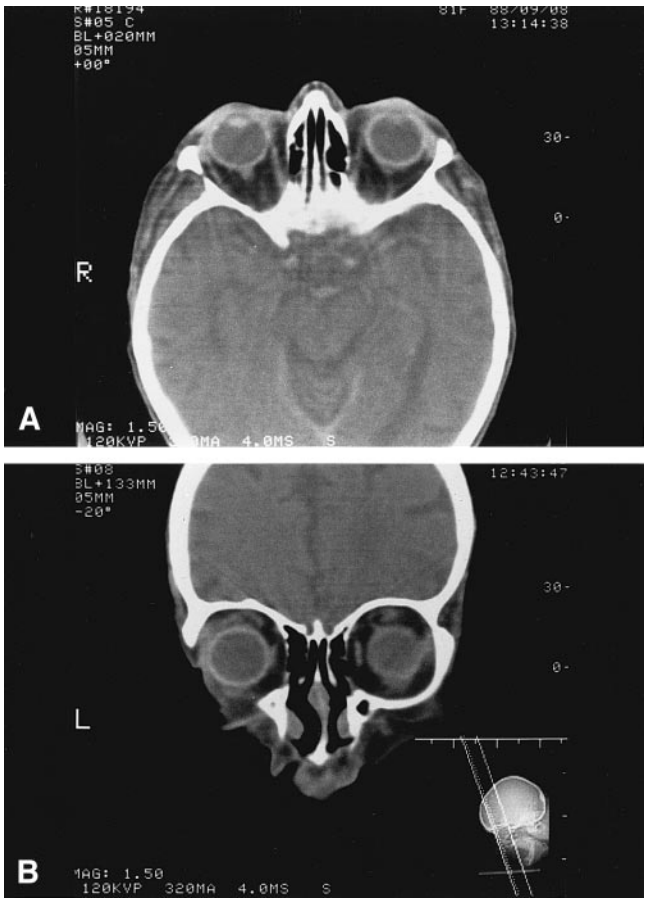

Figure 11. A, appearance 10 days later of scans from the same patient as in Figures 10A andB after two doses of 500 mg of intravenous methylprednisolone and a tapering dose of oral prednisolone. Vision had returned to 6/6, and the scleral and orbital edema have almost completely resolved. The proptosis has regressed. This figure is the coronal view. **B,** axial scan from the same patient as in (**A**). The scleral and orbital edema have almost completely resolved. The proptosis has regressed.

rescein angiography was not found to be helpful in making a diagnosis of posterior scleritis, because the inflammatory changes are deep to the retinal pigment epithelium.

Posterior scleritis was associated with systemic disease in 29% of patients, and 24% of the 28 patients were older than age 40. The systemic associations were similar to those seen in anterior scleritis and included rheumatoid arthritis, Wegener granulomatosis, systemic vasculitis, relapsing polychondritis, and other autoimmune disease. Typical posterior scleritis was seen in a patient with systemic lymphoma and a further patient with multiple myeloma. This association is rare but important and has been reported previously.[11]

Most patients with idiopathic posterior scleritis respond well to treatment with nonsteroidal anti-inflammatory drugs. Patients with loss of vision, evidence of optic nerve involvement, and associated systemic disease usually need more aggressive anti-inflammatory therapy. In this series, all patients with posterior scleritis and systemic disease needed more aggressive treatment with systemic immunosuppressive therapy, but despite this therapy, 31% lost vision with three patients (3%) becoming legally blind. Patients were treated initially with systemic corticosteroids and, when necessary, with additional immunosuppressive therapy including cyclophosphamide or cyclosporin A.

Patients with idiopathic posterior scleritis were not as clearly identifiable a group as those with associated systemic disease, and they presented with variable clinical and B-scan features. Some developed severe painful inflammation clinically and had diffuse scleral thickening and fluid in the Tenon space on B scan. Others presented with relatively little pain and inflammation despite severe visual loss and widespread gross scleral thickening on B scan. Those with nodular posterior scleritis typically had extremely large scleral nodules on B scan. There was no association between diffuse or nodular disease and the risk of visual loss.

The histologic features and systemic associations of scleritis suggest that it is an immune-mediated process. Idiopathic forms may represent ocular autoimmunity. Scleritis responds to therapy with anti-inflammatory and immunosuppressive medication.

In this study, patients older than age 50 and those who developed anterior scleritis in association with posterior scleritis had a higher risk of having an identifiable systemic disease associated with their posterior scleritis. Patients who required treatment with systemic corticosteroids and additional immunosuppressive drugs had a higher risk of visual loss. Statistical analysis was unable to reveal any other specific features in the presentation of posterior scleritis, the clinical features of the patients, or the course of the disease that would identify those patients likely to lose vision or those more likely to have an associated systemic disease. All patients must therefore be assumed to be at risk of visual loss, and all need investigation to search for an associated systemic disease as well as treatment to minimize the risk of developing sight-threatening complications.

Posterior scleritis is a common form of scleritis that should be suspected in patients who present with periocular pain, visual loss, or any of the signs of posterior scleritis. Up to 9% of patients have no physical signs on examination. Occasional patients may present with minimal or no pain but with physical signs typical of posterior scleritis. Careful clinical assessment combined with B-mode ultrasonography is therefore essential for diagnosis. Posterior scleritis responds to therapy with systemic nonsteroidal anti-inflammatory drugs, corticosteroids, and immunosuppressive drugs (Figs 8–11). It is most often unilateral and recurs frequently. A high index of suspicion is necessary to detect this potentially destructive disease early in its course, so that effective therapy can be instituted to control scleral inflammation and limit visual loss.

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