

Chronic Severe Uveitis

Etiology and Visual Outcome in 927 Patients from a Single Center

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Introduction

There is a significant under-registration of blind and visually impaired patients attending ophthalmic departments. Blindness is defined by the World Health Organization (WHO) as the best possible corrected visual acuity in the better eye of less than 3/60 (20/400 or 0.05) or corresponding visual field $\leq 10^\circ$. Severe visual impairment is defined as the best-corrected visual acuity in the better eye of 3/60 (20/400 or 0.05) or more, but less than 6/60 (20/200 or 0.1) (40, 49). In 1995, the WHO estimated that nearly 38 million people in the world were blind and 110 million had severe visual impairment. In 1997, the total number of estimated blind people was 44,800,000 (47). Causes of blindness can be classified according to different age-groups and geographic distribution (49). The major causes of blindness in the middle age-group in the Western world are tapetoretinal degeneration (20%), congenital abnormalities (20%), diabetes (20%), uveitis (10%), and accidents (5%) (17, 21, 25, 44, 48). Approximately 5%–20% of legal blindness in developed countries is due to uveitis (7, 17, 21, 46).

Uveitis is a potentially sight-threatening intraocular inflammation. Because of a variety of diagnostic criteria used by different centers, the epidemiology and etiologic distribution of uveitis have not been clearly defined. As a result, its incidence is reported to range from 17 per 100,000 to 24 per 100,000 residents, and its prevalence from 38 per 100,000 to 204 per 100,000 residents (7, 23, 24, 38, 51). Diagnostic workup determines an infectious etiology or an associated condition in about 60%–70% of cases of uveitis (13, 15, 16, 22, 30, 41, 42, 50, 52). Primary in-

flammatory disorders potentially associated with systemic manifestations or inflammatory conditions associated with infectious agents are most frequently reported. Infectious conditions are often associated with uveitis and blindness, especially in developing countries (6, 31, 32, 49, 53). Usual cases of uveitis, such as B27-associated disease or herpetic uveitis, are easily handled and are not usually referred to a tertiary eye care center. Most of these cases have a favorable evolution. In a number of complicated cases, the diagnosis is not easy to determine. The use of systemic antiinflammatory and immunosuppressive drugs is often justified and has considerably lowered the incidence of complications in these patients. These therapeutic applications are generally performed in specialized centers under the supervision of internists and ophthalmologists (35).

We analyzed a series of 927 consecutive patients referred to our center between 1991 and 1996 for diagnostic and/or therapeutic management of severe uveitis. Therapeutic strategy was evaluated according to the level of intraocular inflammation, and the final visual outcome was determined.

Patients and Methods

The clinical records of all patients with severe or sight-threatening uveitis referred to the Ophthalmology Department of the Pitié-Salpêtrière Hospital, Paris, France, between January 1991 and January 1996 were retrospectively analyzed. Severe uveitis was defined based on the following criteria at the time of presentation: 1) visual acuity less than 20/100, 2) chronic uveitis (duration >3 months), 3) undiagnosed or unclassified cases after more than 2 different workups, and/or 4) uveitis requiring systemic antiinflammatory or immunosuppressive drugs. In all other cases, uveitis was not considered severe, and those patients were excluded. Furthermore, patients presenting with masquerade syndromes (intraocular lymphoma and postoperative or endogenous endophthalmitis) or infection in an immunosuppressed setting (cytomegalovirus retinitis) were excluded. In the latter, infectious or malignant conditions are highly predominant, and ocular inflammation is a secondary problem. On the other hand, cases of uveitis associated with other infectious diseases, such as toxoplasmic retinochoroiditis, Lyme

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disease, and syphilis or viral infections in immunocompetent patients, were included in this study. Immune disorders seem to play a major role in these conditions and should be treated in association with antiinfectious drugs. A minimal follow-up of 2 years was required for inclusion in the study to estimate final visual outcome.

Classification of uveitis

The International Uveitis Study Group (IUSG) classification (2), based on the ocular site of inflammation, was used throughout the study. *Anterior uveitis* was defined as the presence of anterior chamber inflammatory flare or cells associated with keratic precipitates and/or iris lesions, such as synechiae or partial atrophy. Since June 1991, anterior chamber inflammation was objectively quantified with a Laser Flare Cell Meter (KOWA 1000). *Intermediate uveitis* was defined as the presence of cells in the vitreous with or without the presence of inflammatory cell condensations in the anterior vitreous (snowballs) or the peripheral retina. The term "pars planitis" was used to describe a subgroup of patients with intermediate uveitis presenting with white exudates (snowbanks) over the pars plana. *Posterior uveitis* was defined as the presence of inflammatory signs involving the retina, choroid, retinal vessels, and/or posterior vitreous humor. Finally, *panuveitis* was defined as a combination of inflammation involving the 3 previously described sites. Further classifications were established according to other morphologic features. The presence of keratic precipitates visible on slit lamp examination and accumulating on the endothelial surface of the cornea was included. According to their size and their appearance, keratic precipitates were classified as granulomatous and nongranulomatous. Presence of hypopyon, elevated intraocular pressure (>22 mmHg), cystoid macular edema (confirmed by fluorescein angiography), band keratopathy, keratouveitis, cataract, and papillitis was also noted.

Diagnostic criteria

Based on a detailed clinical history, complete laboratory workup and additional ophthalmologic or specialized medical consultations were performed when the origin of uveitis was uncertain, and always before the institution of intravenous steroids or immunosuppressive drugs. Evaluation of patients with uveitis potentially associated with spondylarthropathies was based on the European Spondylarthropathy Study Group criteria (10). HLA-B27 typing was performed in all nongranulomatous cases of uveitis and was not limited to patients with typical HLA-B27-associated anterior uveitis. It is well known that severe, atypical, corticoid-dependent, or corticoid-resistant presentations of uveitis may be associated with seronegative or HLA-B27+ spondylarthropathies; uveitis associated with psoriatic arthritis and inflammatory bowel disease is usually the severe form. Behçet disease was diagnosed according to criteria of the International Study Group for Behçet Disease (5). For granulomatous uveitis associated with sarcoidosis, results of serum angiotensin-converting enzyme and serum lysozyme tests, chest X-ray, and gallium scintigraphy were considered. Evidence of a noncaseating granuloma was required for the diagnosis of sarcoidosis, except when uveitis presented with typical bilateral hilar or mediastinal adenopathy. Vogt-Koyanagi-Harada syndrome was diagnosed according to the classification established by the American Uveitis Society (43). Ocular toxoplasmosis was diagnosed based on the presence of a typical fundus change of focal retinochoroiditis and positive IgG anti-*Toxoplasma* on serology. Syphilis was diagnosed when treponemal hemagglutination (TPHA)

and nontreponemal (VDRL) tests were positive. For tuberculous uveitis, 2 different diagnostic groups were determined. A positive culture for mycobacteria in any fluid or tissue sample was necessary for diagnosis of an active infection by the tubercle bacillus. However, a positive purified protein derivative (PPD) skin test associated with a personal or familial history of tuberculosis, abnormalities on chest X-ray, and granulomatous uveitis without positive culture for mycobacteria was associated with putative hypersensitivity to the bacillus. Herpetic keratouveitis or uveitis was diagnosed based on the presence of corneal scars compatible with a previous herpetic keratitis and/or sectorial iris atrophy and/or ocular hypertension and positive IgG serologic findings. For all potentially infectious uveitis presenting with atypical clinical presentations, anterior chamber or vitreous taps were performed with subsequent local antibody titer determination and, when necessary, specific polymerase chain reactions (PCR) to detect *Toxoplasma gondii*, different herpes viruses, or bacterial agents such as *Tropheryma whippelii* (3, 9). Cases of uveitis that were not associated with a detectable infectious agent or the above systemic disorders were classified as idiopathic.

Statistical analysis and evaluation of visual outcome

Data were entered into a FileMaker Pro (FileMaker, Inc.) database for descriptive statistical analysis. State of initial and final visual acuity was reviewed in each anatomic group of uveitis. For each group, visual acuity was classified in 4 different subgroups: 1) less than 20/400, 2) between 20/400 and 20/200, 3) between 20/200 and 20/40, and 4) more than 20/40. Initial and final visual acuity were compared in each anatomic group using the chi-square test. Statistical significance was defined as an alpha of less than 0.05, showing an improvement after a minimal follow-up of 2 years.

Results

Patient characteristics

Among 1,080 patients presenting with 1 of the 4 criteria required for severe uveitis as described in the Patients and Methods section, 927 were finally included. We analyzed 1,310 eyes. One hundred fifty-three patients were excluded due to short follow-up (<24 months). The mean age was 37.1 years at the time of presentation, but 34.5 years at the onset of disease. The female to male sex ratio was 1.05. Most patients (92%) were Caucasian; the cohort comprised 80% Europeans and 12% North Africans. Blacks represented 6% and Asians 2% of patients. Uveitis was unilateral in 58.7% and bilateral in 41.3% of cases. At the time of referral, 29% of the patients were undergoing medical treatment (Table 1). Examples of different aspects of uveitis are provided in Figures 1–6.

Anatomic and etiologic distribution

Panuveitis was the most frequent form (35%) followed by anterior uveitis (28.5%), posterior uveitis

TABLE 1. Clinical characteristics of 927 patients with severe or sight-threatening uveitis

	No.	(%)
Initial visual acuity < 20/200 (time of referral)	196	21.1
Unilateral	180	19.4
Bilateral	60	6.5
Chronic uveitis	751	81
Systemic treatment at time of referral	269	29
Mean age (yr)		
Onset of disease	34.5	
Time of referral	37.1	
Sex ratio (F/M)	1.05	
Race*		
Caucasian	853	92
European	741	79.9
North African	112	12
Black	56	6.0
Asian	18	1.9
Previous episodes of uveitis $\pm$ SD	5 $\pm$ 0.9	
Mean evolution time of uveitis (yr) $\pm$ SD	3.34 $\pm$ 2.5	
Type of uveitis at time of referral*	927	100
Anterior	264	28.5
Intermediate	139	15
Posterior	200	21.6
Panuveitis	324	35

*Percentages do not equal 100% due to rounding.

(21.6%), and intermediate uveitis (15%). At the end of the study, a noninfectious condition was established in 34.2% of cases and an infectious etiology in 31.8% of cases; uveitis remained idiopathic in 34% of cases. The distribution of cases according to the anatomic site of inflammation and associated conditions of uveitis is shown in Table 2. Excluding the idiopathic cases (37.7%), Behçet disease was the most frequent diagnosis in the panuveitis group (13.9%). Herpes virus infections (31%) and HLA-B27-associated uveitis (17.4%) were the most frequent diagnoses in the anterior uveitis group. Toxoplasmic retinochoroiditis was the most frequent in patients presenting with posterior uveitis (39%), but was also noted in a large proportion of patients with panuveitis (9.9%). In the intermediate

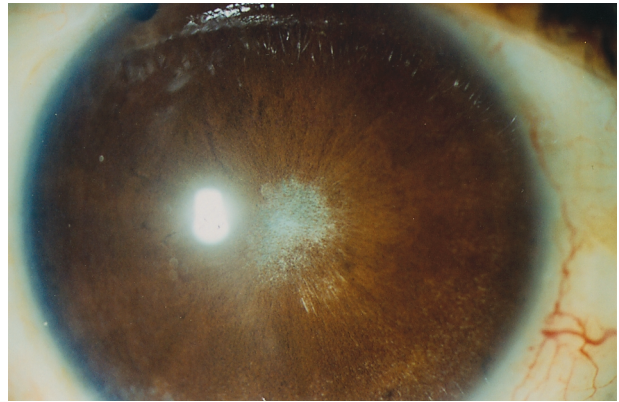


FIG. 2. Posterior synechiae with totally occluded pupil in a patient with tuberculous uveitis.

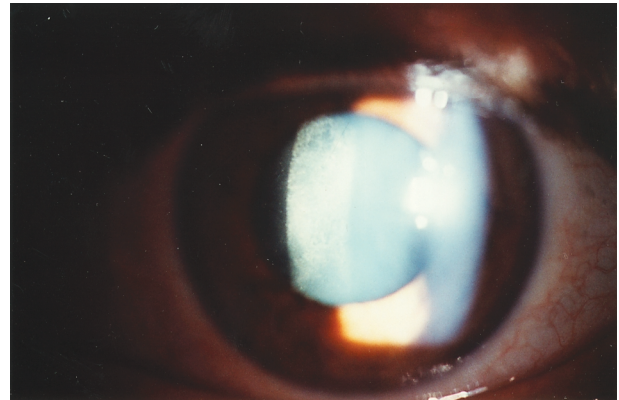


FIG. 3. Posterior subcapsular cataract in a patient with juvenile chronic idiopathic uveitis.

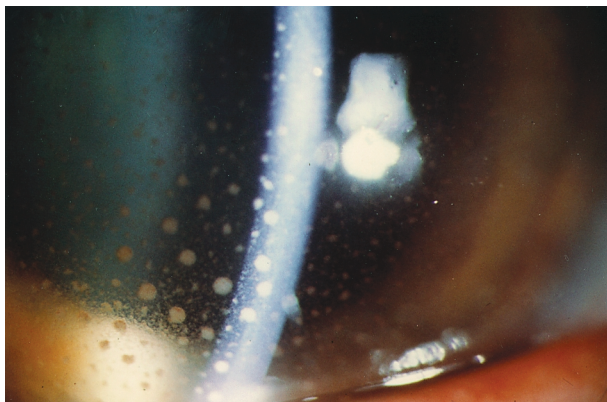


FIG. 1. Mutton-fat keratic precipitates associated with systemic sarcoidosis.

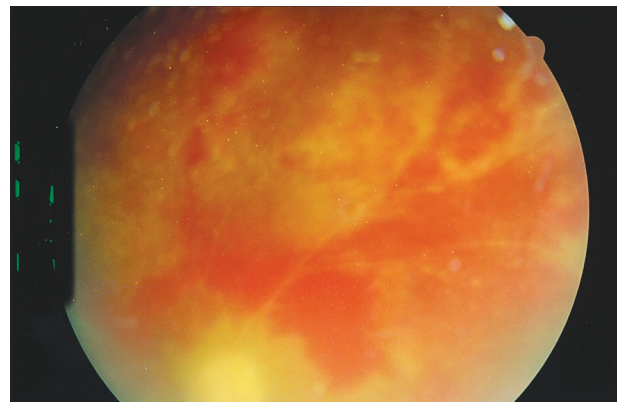


FIG. 4. Viral retinal necrosis in an immunocompetent patient.



FIG. 5. Retinal hemorrhages, optic nerve infiltration, and vasculitis in a patient with a severe case of Behçet disease.

uveitis group, most cases were idiopathic (75.5%). More than 24.9% of noninfectious cases of uveitis were associated with a systemic disease (Table 3).

Therapeutic strategies

Twenty-nine percent of patients were on systemic steroid or immunosuppressive therapy at the time of presentation. The general aspects of our therapeutic management are given in Table 4. The majority of patients received systemic steroids (68%). Intravenous methylprednisolone pulses were administered in 53.8% of cases. Systemic steroids were used in 399 of 632 (63.1%) patients with noninfectious and 231 of 295 (78.3%) patients with infectious uveitis. The best results were obtained in cases of toxoplasmosis involving the zone 1 of Holland, sparing the fovea and associated with a major intraocular inflammation; in complicated cases of sarcoidosis; and in Behçet disease. In severe cases, such as birdshot retinochoroiditis and other corticoreistant or corticodependent types of posterior uveitis or panuveitis, an immunosuppressive regimen was necessary (9.6% of cases). Azathioprine and cyclosporine were most frequently

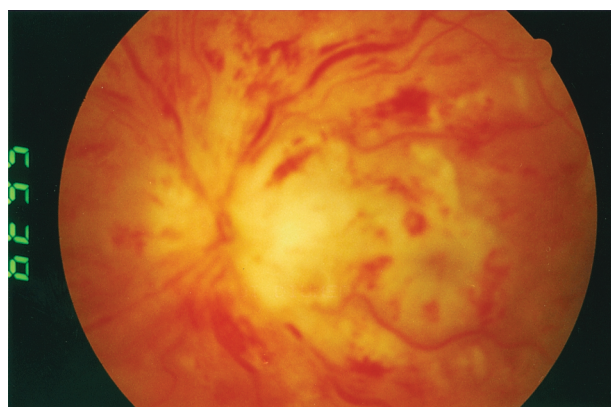


FIG. 6. Toxoplasmic papillitis associated with a central vein occlusion.

used. In all confirmed or presumed infectious conditions, antimicrobial, antiviral, or antiparasitic treatment was combined with the antiinflammatory regimen in order to avoid local or systemic complications. Intravenous polyclonal immunoglobulin (IVIg) treatment was used successfully in 8 cases of birdshot retinochoroidopathy. IVIg was given at 1.6 g/kg every 4 weeks for 6 months, followed by injections of 1.2–1.6 g/kg at 6- to 8-week intervals (18).

Evolution and visual outcome

The mean follow-up was 3.5 years (range, 2–7 yr). Clinical evaluation of intraocular inflammation at the end of the follow-up period confirmed improvement in 59%, stabilization in 22%, and impairment in 19% of cases. Mean values of initial and final anterior chamber inflammation evaluated by laser flare photometry were 27.5 ± 24 ph/ms and 14 ± 8.5 ph/ms, respectively. Anterior chamber inflammation was significantly reduced at the end of the observation period ($p < 0.01$). In terms of visual acuity, values less than 20/200 in unilateral ocular involvement were observed in 19.5% of patients before treatment, but in only 9.7% of patients at the final stage. In bilateral cases, 6.5% of patients presented to our center with initial visual acuity less than 20/200, and only 3.1% had a final visual acuity below 20/200. The etiologic distribution of these uveitis cases associated with severe outcome is summarized in Table 5. We compared the initial and final states of visual acuity in different anatomic subgroups of uveitis in order to evaluate the efficacy of therapeutic strategies. There was a statistically significant improvement in all except the intermediate uveitis group. Treatment did not seem to change the spontaneous evolution of visual acuity in these patients. It is noteworthy that more than 75% of these cases were idiopathic.

Ocular complications

Cystoid macular edema was the main factor influencing visual outcome in this study. Although we noted a significant improvement of cystoid macular edema after treatment, 18.3% of patients still presented this complication at the end of the observation period. Cataract was present in 14.3% of cases. Cataract extraction was performed in 47% of the latter patients during the period of observation. The other 2 principal complications were secondary glaucoma (6.8% of cases with a mean intraocular pressure of 31 ± 1.6 mmHg) and epiretinal membrane (4.2% of cases).

Discussion

Panuveitis was the most common presentation in this study, followed by anterior, posterior, and

TABLE 2. Etiologic distribution by anatomic classification of uveitis

Etiology of Uveitis	No. of Patients (n = 927)	(%)*
Anterior uveitis		
Infectious		
Herpetic infections (herpes simplex and varicella zoster viruses)	82	31
Tuberculosis	13	4.9
Bacterial infection (Lyme disease, rickettsiosis)	6	2.3
Uveitis associated with a systemic disease		
HLA-B27-associated disease	46	17.4
Juvenile idiopathic arthritis	25	9.5
Sarcoidosis	18	6.8
Behçet disease	2	0.8
Multiple sclerosis-associated uveitis	1	0.4
Eye-limited, presumed immune-mediated disorder		
Complicated Fuchs heterochromic iridocyclitis	25	9.5
Idiopathic eye-limited disorder not conforming to a defined syndrome	36	13.6
Other	10	3.8
Subtotal	264	100
Intermediate uveitis		
Idiopathic eye-limited disorder not conforming to a defined syndrome (including pars planitis syndrome)	105	75.5
Uveitis associated with a systemic disease		
Multiple sclerosis-associated uveitis	15	10.8
Sarcoidosis	4	2.9
Infectious		
Lyme disease	4	2.9
Tuberculosis	2	1.4
Rickettsiosis	2	1.4
Other	7	5
Subtotal	139	100
Posterior uveitis		
Infectious		
Toxoplasmic retinochoroiditis	78	39
Syphilis	6	3
Other bacterial infection (Whipple disease)	6	3
Tuberculosis	1	0.5
Idiopathic eye-limited disorder not conforming to a defined syndrome	33	16.5
Uveitis associated with a systemic disease		
Behçet disease	10	5
Sarcoidosis	3	1.5
Eye-limited, presumed immune-mediated disorder		
Birdshot retinochoroidopathy	41	20.5
Multiple evanescent white dot syndrome	3	1.5
Sympathetic ophthalmia	2	1
Other	17	8.5
Subtotal	200	100
Panuveitis		
Idiopathic eye-limited disorder not conforming to a defined syndrome	122	37.7
Uveitis associated with a systemic disease		
Behçet disease	45	13.9
Sarcoidosis	34	10.5
Vogt-Koyanagi-Harada syndrome	19	5.9
Infectious		
Toxoplasmic retinochoroiditis	32	9.9
Tuberculosis	22	6.8
Acute retinal necrosis	14	4.3
Bacterial infection	9	2.8
Eye-limited, presumed immune-mediated disorder		
Sympathetic ophthalmia	9	2.8
Other	18	5.6
Subtotal	324	100
Total	927	100

*Percentages do not equal 100% due to rounding.

TABLE 3. Systemic diseases associated with noninfectious uveitis

Etiology	No. of Patients	(%/927)
Sarcoidosis	59	6.4
Behçet disease	57	6.1
Spondylarthropathies	46	5
Juvenile idiopathic arthritis	25	2.7
Vogt-Koyanagi-Harada syndrome	19	2
Multiple sclerosis-associated uveitis	16	1.7
Other	9	0.9
Total	231	24.9

intermediate uveitis. Because of the reference bias in this patient population, these data cannot be considered for the general population of uveitis patients. These results are different from other recently published surveys, which is partly explained by the analysis of a subgroup of patients suffering from severe uveitis in our series (15, 22, 30). Patients included in this study were those who had the highest risk of ocular complications leading to a poor visual outcome. Under such circumstances, collaboration between ophthalmologists and internists is quite important, requiring extensive workup followed by systemic therapy. The tertiary character of our department could explain the relatively low level of anterior uveitis, which is seen frequently in the general practice of ophthalmology, in this study. This result has

TABLE 4. Medical management of patients with severe uveitis

Medical Treatment	No. of Patients	(%/927)
Immune-mediated uveitis		
Systemic steroids		
Oral	102	11
Intravenous pulse + oral	297	32
Immunosuppressive drugs		
Azathioprine	50	5.4
Cyclosporine	31	3.3
Cyclophosphamide	8	0.9
Intravenous immunoglobulins (exclusively birdshot retinochoroidopathy)	8	0.9
Infectious uveitis		
Systemic steroids		
Oral	29	3.1
Intravenous pulse + oral	202	21.8
Antiparasitic		
Sulfadiazine + pyrimethamine	99	10.7
Antivirals		
Intravenous acyclovir or foscarnet	79	8.5
Antibiotics		
Antituberculous (rifampin, isoniazid, ethambutol, pyrazinamide)	38	4
Pefloxacin	19	2
Trimethoprim + sulfamethoxazole	5	0.5
Tetracycline	8	0.9
Penicillin	9	1
Cephalosporin	33	3.6
Antiglaucomatous		
Local	33	3.6
Local + systemic (acetazolamide)	17	1.8

TABLE 5. Conditions associated with severe uveitis with poor visual outcome (final visual acuity < 20/200)

Type of Uveitis	Associated Conditions	No. of Patients	(%/119)*
Panuveitis	Behçet disease	17	14.3
	Tuberculosis	8	6.7
	Vogt-Koyanagi-Harada	5	4.2
	Sympathetic ophthalmia	4	3.4
	Sarcoidosis	3	2.5
	Bacterial infection	2	1.7
	Toxoplasmosis	3	2.5
	Acute retinal necrosis	3	2.5
	Idiopathic	6	5
	Total	51	42.8
Posterior uveitis	Toxoplasmic retinochoroiditis	19	16
	Birdshot retinochoroidopathy	11	9.2
	Tuberculosis	3	2.5
	Serpiginous choroiditis	4	3.4
	Syphilis	3	2.5
	Idiopathic	9	7.6
	Total	49	41.2
Intermediate uveitis	Multiple sclerosis	2	1.7
	Tuberculosis	2	1.7
	Idiopathic	9	7.6
	Total	13	11.0
Anterior uveitis	HSV or VZV uveitis	2	1.7
	Juvenile chronic arthritis	4	3.4
	Total	6	5.1
Total		119	100

Abbreviations: HSV = herpes simplex virus; VZV = varicella zoster virus.

*Percentages do not equal 100% due to rounding.

been discussed in a previous study reported by McCannel et al (22). Because of their relatively favorable visual prognoses, we have not included patients with uncomplicated HLA-B27 or herpetic uveitis, Fuchs heterochromic cyclitis, and Posner Schlossman disease. However, a subgroup of HLA-B27-associated uveitis has a poor outcome mainly due to posterior segment involvement (29, 39, 45). The systemic disease is frequently diagnosed after the onset of ocular involvement (19, 26, 27, 33, 34, 45).

Associated conditions were defined in 67.5% of cases. The diagnostic rate is higher than that obtained in 1966 in a relatively comparable French population (14). Our study correlates closely with other studies regarding the percentage of cases in which the etiology was determined (57.8%–67%) (16, 22, 30, 42, 50, 52). Patterns of uveitis similar to those reported by Banares et al (1) were present in most of our patients. However, anterior chamber tap, diagnostic vitrectomy, and other organ biopsies allowed us to confirm atypical presentations such as Whipple disease or unusual conditions with longer clinical evolution (3, 28). These cases were considered as uveitis associated with a systemic disease, and patients were on immunosuppressive regimens. Internists play a major role by ruling out a systemic infection such as tuberculosis, Lyme disease, or Whipple disease associated with an ocular inflammation. Moreover, the etiologic diagnosis was often established during the longitudinal follow-up period. Therefore, repeated clinical and

laboratory evaluation of uveitis should be performed during the follow-up. This is the best way to increase the diagnostic efficacy in patients with chronic uveitis.

As previously reported (8), the highest proportion of idiopathic uveitis was found among patients with the intermediate type. The proportion of noninfectious and infectious etiologies of uveitis is nearly the same in this study. Uveitis associated with tuberculosis or herpetic infections is still an important cause of complication with poor visual outcome. Ocular manifestations associated with tuberculosis are caused by either an active infection or an immunologic reaction in the absence of any infectious agent (4, 11). Molecular tools such as conventional or nested PCR are now applied to ocular specimens to detect the presence of *Mycobacterium tuberculosis* (20). Until the last 2 years, isolation of *Mycobacterium tuberculosis* from the eye was particularly difficult. Hematogenous dissemination of the bacillus is usually associated with choroiditis, and the bacillus is isolated in culture. This type of choroidal involvement happened quite rarely in our surveys. Toxoplasmosis remains the leading cause of posterior uveitis in the French population. For noninfectious uveitis, 75.53% of cases were associated with systemic disorders, and associated conditions were identified (36). These results emphasize the importance of a close collaboration between internists and ophthalmologists in managing patients with severe uveitis.

Systemic steroids and/or immunosuppressive regimens were used in 77.5% of cases. Every time an infectious etiology was suspected, an antiviral, antiparasitic, or antimicrobial treatment was initiated, usually 2 days before the use of antiinflammatory drugs. Immunosuppressive and immunomodulatory agents were principally required in patients with Behçet disease, birdshot retinochoroidopathy, Vogt-Koyanagi-Harada syndrome, sympathetic ophthalmia, and severe idiopathic uveitis. In anterior and panuveitis, anterior chamber inflammation evaluated by laser flare photometry during treatment was significantly reduced.

Initial and final visual acuity were compared statistically. It is important to highlight that there was a significant difference after treatment in all but the group of patients with intermediate uveitis. This might be related to the high level of idiopathic cases and a steroid-independent evolution in this group. These results are similar to those reported by Deane and Rosenthal (8). The principal complications reported in our patients were cystoid macular edema and cataract (37). However, the prevalence of cystoid macular edema is lower than in other reports. This could be partially explained by the rapid and intensive antiinflammatory treatment initiated as soon as infectious conditions were ruled out. Cystoid macular edema responded partially to steroids and required immunosuppressive drugs especially when associated with Behçet disease and birdshot retinochoroidopathy. Cataract required surgical extraction in most patients. The prognosis has dramatically changed since the phacoemulsification era, leading to a minimal and transient rupture of the blood-aqueous barrier. However, the surgical procedure should be performed in a quiet eye, after a period of 3 months and with a special antiinflammatory protocol. In critical situations, the use of laser flare photometry could help the ophthalmologist to taper steroid doses progressively and to detect a relapse occurring at a subclinical level, in order to increase the antiinflammatory regimen without delay (12).

The incidence of blindness among patients with uveitis is poorly defined. The incidence of total blindness due to uveitis in the United States was estimated to be 10%–15%. Previous statistics published in 1962 (7), based on the epidemiology of uveitis (excluding trauma and keratitis) in the small urban community of Rochester, Minnesota, estimated the prevalence of uveitis as a cause of blindness to be in the range of 10 per 100,000 population. This corresponds to nearly 5% of the prevalence of clinically identifiable uveitis determined in Rochester (204 per 100,000 population). More recently, similar data from different European surveys have been published (17, 21, 37, 46). In the present study we retrospectively evaluated the associated conditions and the prevalence of visual

impairment in patients with severe uveitis referred to a tertiary eye care center with a minimum follow-up of 2 years (mean, 3.5 yr). Close collaboration between ophthalmologists and internists is proposed, especially in sight-threatening cases of uveitis. Molecular tools adapted to ocular fluids could improve the diagnostic procedure by confirming infectious conditions to be treated specifically (9, 20). In other cases, systemic steroids or immunosuppressive regimens should be undertaken more rapidly to prevent irreversible visual damage or general complications. Randomized studies with long-term follow-up are necessary to validate different therapeutic protocols and to determine the prevalence of blindness in patients with severe uveitis.

Summary

To determine the infectious etiologies, associated conditions, and visual prognosis of patients with uveitis, we retrospectively analyzed the records of 927 consecutive patients with severe ocular inflammation defined as chronic or sight-threatening who were referred for diagnostic and/or therapeutic management between 1991 and 1996. A minimal follow-up of 2 years was required. The mean age at onset of uveitis was 37.1 years, and the female to male ratio was nearly 1. Most patients were Caucasian (92%). Uveitis was chronic in 81% of cases, associated with an initially reduced visual acuity in 21.1% of cases. The disease was unilateral in 58.7% and bilateral in 41.3% of cases. Panuveitis was the most common presentation (34.9%), followed by anterior (28.5%), posterior (21.6%), and intermediate uveitis (15%). An associated condition was determined in 67.5% of cases. Noninfectious uveitis and systemic diseases were associated in 24.9% of patients. Most patients required systemic high-dose steroids and/or immunosuppressive regimens (77.5%). Cystoid macular edema and cataract were the most common complications. Intraocular inflammation was controlled in 81% of the patients. Final visual acuity was less than 20/200 in 3.1% of bilateral and 9.7% of unilateral cases. The management of patients with severe and sight-threatening forms of uveitis is efficiently performed in collaboration with internal medicine physicians and depends on a complete diagnostic procedure and well-adapted treatment. As a result, the rate of blindness due to uveitis can be decreased.

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