Epidemiological, etiological and therapeutic profile of intermediate uveitis in adults: a review of 42 cases

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Abstract :

Background :

Uveitis is the third leading cause of legal blindness worldwide. Intermediate uveitis is the least frequent anatomical form (4 to 20% of all uveitis). They pose the problem of their etiological diagnosis requiring numerous and costly biological and radiological investigations that provide etiological diagnostic certainty in less than 30% of cases.

The aim of this study is to describe the epidemiological and etiological characteristics of intermediate uveitis through the experience of our structure.

Patients and methods :

Retrospective study of 42 patients followed for intermediate uveitis in the adult ophthalmology department, Hospital 20 August 1953 of Casablanca, between January 2017 and December 2021. All patients had a complete ophthalmologic examination followed by a clinical and paraclinical workup in collaboration with internists. Diagnoses were made according to recent international criteria.

Results :

The average age was 31.66 ± 10 years (extremes 16 and 74 years) with a male predominance and a sex ratio *M/F* of 2. The main reason for consultation was decreased visual acuity (84%) and/or myodesopia (26%). The damage was bilateral in 63% of cases. The initial visual acuity was less than 1/10 in 73.6% of cases. The most frequent etiologies were Behçet's disease (23.6%), followed by sarcoidosis (18.4%), tuberculosis (13%), multiple sclerosis (5.3%) and idiopathic disease (40%). The main complications found were macular edema (26.3%), cataract (23.6%) and ocular hypertonia (13%). All patients received corticosteroid treatment (100%), immunosuppressive treatment (21%) and antibacillary treatment (13%). Visual acuity at 6 months of treatment was superior to 3/10 in 31%.

Conclusion :

Intermediate uveitis mainly affects young adults with an average age of 30 to 40 years without gender predominance. They are idiopathic in 70 to 85% of cases. The etiologies most frequently found in the literature are sarcoidosis, tuberculosis and multiple sclerosis. In our series, Behçet's disease was the most frequent etiology, which can be explained by its predominance in our country and in the Mediterranean basin, as the first etiology of uveitis of all types. A well established interdisciplinary cooperation between ophthalmologists and internists is essential for the optimization of the management process and a better etiological diagnostic yield. **Keywords :** Intermediate uveitis, young adults, idiopathic origin, Behçet's disease, corticosteroid therapy.

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I. Introduction :

Intermediate uveitis is a chronic and insidious disease. Children and young adults are preferentially affected. Vision loss and floaters are commonly reported in a quiet eye. The inflammation predominates in the vitreous and some characteristic peripheral exsudates may be observed. Intermediate uveitis is generally idiopathic but may sometimes be associated with a systemic disease that must be researched and treated. The treatment of the idiopathic form is only necessary in case of complications and will consist in corticosteroids eventually combined with immunosuppressive drugs. Surgery (cryotherapy, vitrectomy) will be reserved for complications non responding to medical therapy.

Our Objective was to describe the epidemiological and etiological characteristics of intermediate uveitis through the experience of our structure.

II. Materials And Methodes :

This is a retrospective study conducted between January 2017 and December 2021 in our ophthalmology department, gathering 42 adult patients followed in our specialized uveitis consultation with intermediate uveitis. All patients underwent a detailed interview and a complete ophthalmologic examination, including a measurement of the best corrected visual acuity, a measurement of ocular tone, a slit lamp examination, and a fundus examination with a Goldmann 3-mirror lens. Complementary examinations (fluorescein retinal angiography, optical coherence tomography (OCT), ocular ultrasound) were requested whenever their indications arose. All patients received an internal medicine consultation, and other specialized examinations were requested if necessary in collaboration with the internists. The diagnoses were established according to recent international criteria.

III. Results :

The total number of patients was 42, the average age was 31.66 ± 10 years (extremes 16 and 74 years) with a male predominance and a sex ratio M/F of 2. The average time between the diagnosis of uveitis and the appearance of the first ocular disease was 5 years. In our series, 40% of the patients had at least 2 episodes of recurrent red eye per year, and 63% of the cases were bilateral. The main reason for consultation was a decrease in visual acuity (84%) and/or myodesopsia (26%). Clinically, the initial visual acuity was greater than 5/10 in 2.4% of cases, between 1/10 and 5/10 in 18% of cases, less than 1/10 in 73.6% of cases, and bright perception in 6% of eyes. A perception of floating bodies was noted in the majority of patients (85%), some patients were asymptomatic and the external appearance of the eye was normal in 10% of cases and rarely an inflammation of the anterior segment, translated by pain, redness and photophobia was noted (only in 8.79%). The appearance of snowballswas found in 30% of the patients, snowbankin 11.5% and engulfment of the small peripheral veins in 5.2% of the cases. The most frequent etiologies were represented by Behcet's disease (23.6%), followed by sarcoidosis (18.4%), tuberculosis (13%), plaque sclerosis (5.3%) and idiopathic (40%) (Table 2). While the complications were dominated by macular edema (26.3%), cataract (23.6%) and ocular hypertonia (13%), followed by intravitreal hemorrhage and epi-retinal membrane in (1.7%) (Table 1). In our series, all patients received corticosteroid treatment (100%), immunosuppressive treatment (21%) and antibacillary treatment (13%). 50% of the patients were on colchicine, and 5.8% on antivitamin K. After a mean follow-up of 6 months, 31% of the patients maintained stable visual acuity under treatment of more than 3/10, 23% showed a significant decrease in visual acuity, including 5% of cases of absolute blindness. Only 6.6% showed an improvement in visual acuity with treatment and 39.4% of patients were lost to follow-up.

IV. Discussion :

The distribution of uveitis types and etiologies in a given population is strongly influenced by a variety of genetic, geographic, environmental, and socioeconomic factors. In Western and American countries, the incidence of uveitis is reported to be 17-52 cases per 100,000 population per year and its prevalence 38-204 cases per 100,000 population [1,2]. Anterior uveitis accounts for 35.24%, posterior uveitis 10.48% and panuveitis 48.57%. While intermediate uveitis represents only 5.71%. In Western and American studies, anterior uveitis is the predominant anatomical type 28.5% to 62% followed by posterior uveitis 13 to 26%, then intermediate uveitis and panuveitis [2,6]. In our Mediterranean region, the frequency of intermediate uveitis is estimated at 8-18% of uveitis in general. Children and young adults are preferentially affected. There does not appear to be a predilection for race or gender. In addition, hereditary and environmental factors appear to be involved with the observation of familial cases (3.5%). The association with HLA antigens, in particular HLA DR2 also linked to multiple sclerosis, has been determined (13.17%). In our series, the average age was $31.66 \pm$ 10 years (extremes 16 and 74 years) with a male predominance and a sex ratio M/F of 2. The characteristic complaints of intermediate uveitis are visual disturbance and perception of floating bodies. Sometimes patients are asymptomatic and it is then a chance discovery during a routine ophthalmologic examination. The external appearance of the eye remains normal in most cases. The onset is therefore particularly insidious. Rarely, the symptomatology can take a brutal character by posterior detachment of the vitreous or by the appearance of an inflammation of the anterior segment, resulting in pain, redness and photophobia. The anterior segment is usually normal but there are slight inflammatory signs. The diagnosis of intermediate uveitis is based on clinical findings. The diagnosis of intermediate uveitis is based on clinical findings. The diagnosis of intermediate uveitis is based on clinical findings and is necessary to exclude a specific etiology and to determine the idiopathic origin. Thus, it is idiopathic in 70 to 85% of cases. The most frequent etiology found in our study is Behçet's disease, followed by sarcoidosis, tuberculosis and multiple sclerosis (Table 2). The first one is explained by its predominance in our country and in the Mediterranean basin, as the first etiology of uveitis of all types. Similar frequencies have been reported in several series from the Near and Far East, and countries around the Mediterranean basin [8]. Chebil et al. found Behcet's disease first 14.7% followed by toxoplasmosis 10.2% then VKH 3.7% [9]. When there is an etiological orientation, infectious uveitis is in the majority (30% to 50% of the etiologies) [10]. Concerning complications, in the literature, cystoid or non-cystoid diffuse macular edema is the most frequent complication of the disease, it conditions the long-term visual prognosis and can lead to the formation of a macular hole [10]. Cataract is the most frequent complication in our study, it is secondary to uveitis or to topical or general corticosteroid treatment, the most frequent aspect is that of posterior subcapsular cataract [11]. Other complications may involve the anterior segment:posterior synechiae, ocular hypertonia, secondary glaucoma; or the posterior segment: retinal vein branch occlusions, central retinal vein occlusions, retinal arterial branch occlusion, preretinal or prepapillary neovascularization associated or not with peripheral retinal ischemia [12]. Other less frequent complications have been reported: intravitreal hemorrhage, retinal dehiscence, retinal detachment and globe phthisis [1] (Table 1) [9, 13, 14]. In case a specific cause is found, its specific treatment is required and may allow resolution of the eye disease. When an idiopathic pathology is found, treatment is necessary when vision is less than or equal to 5/10 or when macular edema is observed. The treatment of choice for idiopathic intermediate uveitis is periocular cortisone injections, and systemic corticosteroids are usually reserved for severe, bilateral disease or when local treatment is insufficient. If local and systemic corticosteroids are not sufficient, immunosuppressive treatment can be combined with good results. Cyclosporine, azathioprine and methotrexate are the most suitable products. These treatments should be followed by an experienced internist to reduce iatrogenic complications. Surgically, two options can be proposed in intermediate uveitis but their indications must be restricted. Ice pack cryotherapy can be successful in one or more treatments. Vitrectomy is generally reserved for complications of intermediate uveitis: dense vitritis not responding to medical treatment, vitreous hemorrhage not resolving spontaneously, retinal detachment and epimacular membrane. In our series, all patients received corticosteroid treatment (100%), immunosuppressive treatment (21%) and antibacillary treatment (13%). 50% of the patients were on colchicine, and 5.8% on antivitamin K. At a mean follow-up of 6 months, 31% of patients maintained stable visual acuity under treatment greater than 3/10, 23% showed a significant decrease in visual acuity, and 6.6% of patients showed an improvement in visual acuity under treatment. Classically, blindness occurs in approximately 50% of cases within 5 years of the first ocular sign, however, due to therapeutic advances in the management of intraocular inflammation, the visual prognosis has improved in series of patients presented after the 1990s [9]. In our series, absolute blindness was reported in 5% of patients.

V. Conclusion :

The most frequent etiologies of intermediate uveitis in our context are Behçet's disease and sarcoidosis. Knowledge of the epidemiological profile in a given population is a valuable aid in the orientation of the etiological approach. A well-functioning interdisciplinary cooperation between ophthalmologists and internists is essential for the optimization of the management process and a better etiological diagnostic yield.

CONFLICT OF INTEREST :

No potential conflict of interest selevant to this article was reported.

PATIENT CONSENT :

Patients provided written informed consent for publication and the use of their images.

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| Anatomic Site | Number of cases | Percentage |
|-------------------------|-----------------|------------|
| Macular edema | 11 | 26,3 % |
| Cataract | 10 | 23,6% |
| Ocular hypertonia | 5 | 13% |
| Intravitreal hemorrhage | 1 | 1,7% |
| Epi-retinal membrane | 1 | 1,7% |
| Other | 14 | 33,7% |

Table 1 : Complications of intermediate uveitis.

| Etiologies | Number of cases | Percentage |
|--------------------|-----------------|------------|
| Idiopathic | 17 | 40% |
| Behçet's disease | 10 | 23,6% |
| Sarcoidosis | 8 | 18,4% |
| Tuberculosis | 5 | 13% |
| Multiple sclerosis | 2 | 5,3% |

 Table 2 : Distribution of etiologies of intermediate uveitis.

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