UVEITIS AND CELIAC DISEASE: ABOUT TWO CASES

LM. Hmidchat, M. Laghmari, M. Tijan, O. Cherkaoui
Departement of Ophthalmology, University Hospital Center of Rabat, Morocco.

ABSTRACT

Case Reports: A 25-year-old woman, suffering from chronic diarrhea since childhood, was referred to our Ophthalmology Department for blurred vision; clinical and angiographic examinations revealed bilateral panuveitis with chorio-retinal involvement. The diagnosis of celiac disease was made through jejunal biopsy. Laboratory investigations showed no evidence of other causes of uveitis.

A 11-year-old girl who had a history of familial celiac disease confirmed by bowel biopsy; she has been on a gluten-free diet for several years. She complained of visual loss in her left eye. Ocular examination disclosed a pars planitis with severe macular edema. Etiologic investigations of uveitis were negative.

In both cases, the ophthalmologic features improved significantly under gluten-free diet and general steroid therapy.

Conclusion: The association of celiac disease and uveitis seems to have a certain morbid linkage. Ophthalmologic evaluations must be done in all patients with celiac disease.

Keywords: Celiac disease; Uveitis.

INTRODUCTION

Celiac disease is an enteropathy due to gluten intolerance. It is characterized by complete atrophy of the villi of the small intestine resulting in malabsorption. It may present with a variety of symptoms including diarrhea, steatorrhea, abdominal pain, weight loss, or anemia. Ophthalmologic findings are rarely reported in celiac disease: progressive external ophtalmoplegia, retinitis pigmentosa, keratoconjunctivitis, and uveitis [1-3]. We report here two patients with uveitis and celiac disease.

CASE REPORTS

Case 1
A 25-years-old female patient was referred to our department because of blurred vision in her left eye. She had a several year history of malabsorptive syndrome with chronic diarrhea, sideropenic anemia, dyspepsia, and weight loss. She also suffered form recurrent oral ulceration. Visual acuity was 20/30 in the right eye (OD) and 20/200 in the left eye (OS). Intraocular pressure was normal in both eyes. Slit lamp biomicroscopy showed anterior chamber flare and vitreous cells more distinct in the left. Fundus examination revealed a peri-papillary atrophic area with macular edema in the OS confirmed with optical coherence tomography (OCT) at 532 microns thickness, multiple small deep white lesions alternating with pigmented small lesions in both eyes (Fig.1). Those white dots showed early hyperfluorescence and late staining in the fluorescein angiography (Fig.2). Physical examination showed conjunctival paleness, abdominal inflation, and edema of the lower limbs. A tuberculin skin test was negative.

The laboratory investigations including determination of the blood count showed sideropenic anemia; electrolytes were altered and confirmed the malabsorption syndrome. Determination of antibodies against treponemapaalidum, Toxoplasma, Toxocara, and human immunodeficiency virus was negative.

IgA and Ig G anti-gliadin antibodies (AGA) and IgG anti endomyisal antibodies (EMA) were strongly positive. Pre-endoscopic jejunal biopsy revealed a severe atrophy of villi (grade IV).
compatible with fully clinically expressed adult celiac disease. Treatment consisted of a gluten-free diet and oral prednisone (1 mg/kg/day and tapered slowly over months). This therapy induced clinical recovery and marked weight gain. Visual acuity improved to 20/20 OD and 10/20 OS.

Figure 1. A. Right peripheral fundus from case 1 shows pigmented and white lesions. B. Left fundus photography of case 1 shows peripheral multiple white lesions. Note that dense vitritis blurs the photograph.

The anterior chamber became clear and vitreous opacities decreased. Ophthalmoscopy revealed improvement of the macular edema with the same previous findings in the mid periphery of both fundi.

Case 2

A 12-year-old girl who had suffered from celiac disease since she was 5 was referred to the Ophthalmology Department at the hospital of specialties of Rabat because of a severe OS visual loss. Celiac disease had been confirmed by jejunal biopsy and the patient was on a gluten-free diet. Her visual symptoms and diarrhea occurred after her going off the gluten-free diet. The screening of close relatives showed that the patient’s mother and elder sister were suffering from a less expressive celiac disease.

Visual acuity was OD 20/20 and OS 20/200. Slit lamp examination disclosed a pars planitis with vitreous cells, a mild optic nerve hyperemia and macular edema (Fig.3).

Tests for converting enzyme, erythrocyte sedimentation rate, and antinuclear antibodies, rheumatoid factor, serum angiotensin-converting enzyme syphilis serology, gave normal or negative results. A tuberculosis skin test was negative, and x-rays of sacroiliac, wrist, knees, and chest were normal. Because of the severity of the case, a pulse therapy was given: 660 mg/day of methylprednisolone for three consecutive days. Then the patient was treated with 1mg/kg/day of prednisone and slowly tapered.

One month later, visual acuity had dramatically improved to OS 20/20, vitreous inflammation had decreased with spectacular reduction of macular edema at ophthalmoscopy and fluorescein angiography. The gluten-free diet was maintained and there was no relapse of ocular symptoms during 12 months after the end of steroid treatment.

DISCUSSION

Celiac disease is an intolerance of the gluten of wheat, barley, rye and oats characterized by atrophy of the intestinal villi with different degrees of bowel malabsorption[2]. It typically affects children and young adults. Diagnosis is made through jejunal biopsy; treatment consists of eliminating gluten from the diet [4].

Despite numerous studies, the pathogenesis of celiac disease remains uncertain; it is thought to be caused by both immunologic pathogenesis and genetic predisposition [5] with preferential association with some alleles of the major complex of histocompatibility of class II : DQG*0201 and DQA1*0501. Currently, we think that the toxic fraction of gluten, the gliadin, is the antigenic cause of this disease [6, 7].

A large number of diseases have been reported to be associated with celiac disease, many with a probably immunologic pathogenesis [8]. This disease has been described associated with celiac...
disease, many with a probably immunologic pathogenesis. This disease has been described associated with dermatitis, cutaneous vasculitis, glomerulonephritis, pericarditis, polyarthritis, chronic hepatic diseases, T large granular lymphocytes leukemia [9] and sarcoidosis [5]. Celiac disease has also been associated with some rare ocular features such as uveitis [1,2]. To our knowledge, this association has been reported in only 6 cases; five of these were in the same circumstances as our second case report where celiac disease was already diagnosed and the uveitis occurred at the same time as the digestive symptoms [1, 5, 9]. In another case [2], uveitis was the revealing sign of celiac disease (case 1). The mechanism of this association remains unknown. Different hypotheses have been suggested; an immune reaction against an abnormal permeability to the gliadin leading to a lymphocytic antigenic stimulation seems the most plausible. Our two cases support the association between celiac disease and uveitis. The linkage seems certain in our patients; we could find no evidence of other reasons for their ocular inflammation and in case 2, uveitis occurred at the same time as the symptom of diarrhea. On the other hand, about 25% of patients with celiac disease also have a history of oral ulceration, and the incidence of celiac disease in patients with oral ulceration is 2%-4% [10]. Some authors have reported a possible association between celiac disease and Behcet’s disease: these two diseases share the feature of recurrent oral ulceration -the hallmark of Behcet’s disease-; it may be the only presenting feature of celiac disease [8, 11]. They suggested that looking for evidence of celiac disease screening might be useful in Behcet’s disease and individuals may have typical mucosal change without overt symptoms [8]. The 1st patient had a history of many years of relapsing oral aphthous but did not meet the diagnosis criteria of the Behcet’s disease International Study Group [12]. Ig A AGA and Ig G EMA, which are the most specific combination of serologic screening tests for celiac disease, were strongly positive in our patient.

CONCLUSION

Our two cases illustrate the association between uveitis and celiac disease. Genetic and immunologic mechanisms seem to underlie both affections. We think that this linkage is underestimated because it is not often searched for or identified. It seems interesting to consider patients with celiac disease for thorough ophthalmological evaluation.

REFERENCES