Bilateral Posterior Uveitis with Retinitis Revealing Primary Sjögren's Syndrome

Salem Bouomrani[1,2,*] and Nesrine Regaïeg[1]

[1]*Department of Internal medicine, Military Hospital of Gabes, Gabes 6000, Tunisia*

[2]*Sfax Faculty of Medicine, University of Sfax, Sfax 3029, Tunisia*

Abstract: *Introduction:* Ocular manifestations are frequent during primary Sjögren's syndrome (PSS) and largely dominated by the signs of xerophthalmia. The extra-glandular ocular involvement is, on the contrary, exceptional and unusual. We are reporting an original case of bilateral posterior uveitis with retinitis inaugural of PSS.

Case report: A 48-year-old Tunisian woman, with no notable pathological history, was explored for progressive decline in visual acuity, bilateral visual blur, and moderate eye pain.

The examination noted a visual acuity at 6/10 on the left and 5/10 on the right, without redness of the eyes or irritative signs. The anterior segment of both eyes was normal.

Ophthalmologic exam (slit lamp, fundus, and retinal angiography) revealed bilateral, non-granulomatous posterior uveitis associated with retinitis and retinal vasculitis.

Further investigations concluded to PSS and treatment with intravenous pulse methylprednisolone followed by oral prednisone was introduced. The evolution was rapidly favorable and ophthalmological exam with eye fundus was totally normal at two months.

Conclusion: PSS remains an exceptional and unusual cause of uveitis. It is therefore appropriate to evoke it in front of any uveitis that does not prove itself, especially if recidivating.

Keywords: Posterior uveitis, Uveitis, Retinitis, Primary Sjögren's syndrome, Sicca syndrome, Retinal vasculitis.

INTRODUCTION

Primary Sjögren's syndrome (PSS), also known as primary sicca syndrome, is a fairly common connective tissue disease, particularly in the elderly. Its prevalence is estimated at 0.1-3% of the general population according to series [1, 2]. His clinic is mainly dominated by glandular lesions with major symptoms of xerostomia and xerophthalmia [1-3].

Despite its relative frequency and ease of diagnosis, this disease remains enigmatic, not well known, and often difficult to diagnose, especially if atypical or unusual presentations [2, 3].

As a result, several criteria have been proposed for the diagnosis of this disease; the most recent are those developed jointly by the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) in 2016, recognizing the systemic clinical manifestations of this connectivitis and the specific immunological signature by anti-Ro antibodies [4].

The extra glandular manifestations of this disease are noted in more than 30% of cases and can occur at any time in the evolution, sometimes even reveal the disease [5, 6].

We are reporting an original case of exceptional and unusual extra glandular ocular involvement inaugural of PSS.

CASE REPORT

A 48-year-old Tunisian woman, with no notable pathological history, was transferred to our department by his city ophthalmologist for the management of bilateral non granulomatous posterior uveitis with retinitis.

Eye complaints started fifteen days ago, with a progressive decline in visual acuity, bilateral visual blur, and moderate eye pain.

The examination noted a visual acuity at 6/10 on the left and 5/10 on the right, without redness of the eyes or irritative signs. The anterior segment of both eyes was normal.

Ophthalmologic investigations (slit lamp, fundus, and retinal angiography) concluded to bilateral, non-granulomatous posterior uveitis, associated with retinitis, and retinal vasculitis.

*Address correspondence to this author at the Department of Internal medicine, Military Hospital of Gabes. Gabes 6000. Tunisia;
Tel: +00216 98977555;
E-mail: salembouomrani@yahoo.fr

E-ISSN: 2309-6136/18

The somatic examination in our department was without abnormalities. Biology revealed an erythrocyte sedimentation rate at 65mmH1 and polyclonal hypergammaglobulinemia at 23g/l. The other basic biological tests were within normal limits: total blood count, C-reactive protein, creatinine, ionogram, glycaemia, liver enzymes, muscle enzymes, and urine analysis.

Biopsy of the minor salivary glands revealed a chronic lymphocytic sialadenitis (grade 3 of Chisholm), with no evidence of granulomatosis, caseous necrosis, or neoplastic cells.

The immunoassay showed positive anti-nuclear autoantibodies at 1/164 with speckled immuno-fluorescence pattern, and strongly positive anti-SSA (anti-Ro) antibodies.

Other investigations for infections (particularly tuberculosis), autoimmune diseases, neoplasms, and systemic angiitis were negative.

Similarly, there were no evidence for cardiac, neurological, pulmonary or renal involvement thus; the diagnosis of PSS complicated with bilateral posterior uveitis, retinitis, and retinal vasculitis was retained.

The patient was treated with intravenous pulse methylprednisolone at a dose of 1g/day for three days, followed by oral prednisone at a dose of 1mg/kg/day with adjuvant therapy. After four weeks, corticosteroids were gradually tapered.

The evolution was rapidly favorable with progressive disappearance of ocular complaints and totally recuperation of visual acuity. At two months, ophthalmological exam with eye fundus was normal and without sequel.

DISCUSSION

Ocular manifestations may be indicative of PSS in 24% of cases [7]. They are largely dominated by the signs of xerophthalmia, namely dry eye, foreign-body sensation, ocular pruritus, photophobia, eye redness, visual blur, and decreased visual acuity [1, 2, 5, 8]. Keratoconjunctivitis sicca remains the most specific and characteristic feature of ocular involvement of PSS [2, 8].

On the other hand, extra glandular ocular manifestations are exceptionally reported during this connectivitis [7, 9, 10]. These events include; ocular hypertonia, cataract, corneal ulcer, corneal perforation, scleritis/episcleritis, conjunctivitis, retinitis, optic neuropathies, and uveitis [1, 7]. These manifestations appear to be largely underestimated in current medical practice, because often drowned in the global clinic of the disease. Indeed, their systematic search revealed them in 57 of the 163 patients with PSS of Mathews PM *et al.* (34.9%) [7].

These extra glandular ocular complications appear to be significantly more common in men [7].

Among the extra glandular ocular manifestations of this connective tissue disease, uveitis were exceptionally reported as sporadic cases [9]; they are classically chronic, total and bilateral [9]. They are often diagnosed at a late stage with complications such as synechiae or ocular hypertension [9].

Similarly, PSS remains an exceptional and unusual cause of uveitis [7, 9, 10-12]. Indeed, in the Rosenbaum JT *et al.* series of 185 patients referred in ophthalmology for uveitis of undetermined origin, only eight had Sjögren's syndrome as etiology (4.32%) [9], and only five patients had this syndrome in the series of 163 patients with uveitis of Mathews PM *et al.* (3%) [7].

These findings were also noted for childhood uveitis where only one case was related to PSS in the pediatric series of Stoffel PB *et al.* of 70 patients with uveitis (1.42%) [10].

In our country, this syndrome seems to be even more unusual and more exceptional as etiology of uveitis. Indeed no case was noted in the series of 424 cases of uveitis of Chebil A *et al.*, of which 25.3% was related to systemic diseases [11], and no case was also noted in the series of 472 adult patients with uveitis of Khairallah M *et al.*, of which 20.1% were related to systemic diseases [12].

Our observation is further characterized by its association with retinal vasculitis, an association not found in the previous literature. Indeed, no case of the seven bilateral, total, and chronic uveitis of Rosenbaum JT *et al.*, reported during PSS, had associated retinal vasculitis [9].

CONCLUSION

Primary Sjögren's syndrome remains an exceptional and unusual cause of uveitis. It is therefore appropriate to evoke it in front of any uveitis that does not prove itself, especially if recidivating. Early diagnosis and adapted management are the only guarantors for a better prognosis of these disorders.

CONFLICTS OF INTEREST

No conflicts.

REFERENCES

[1] Choi W, Lee SS, Park YG, Yoon KC. A case of necrotizing keratoscleritis in primary Sjogren's syndrome. Korean J Ophthalmol. 2011; 25(4): 275-7. https://doi.org/10.3341/kjo.2011.25.4.275

[2] Maślińska M, Przygodzka M, Kwiatkowska B, Sikorska-Siudek K. Sjögren's syndrome: still not fully understood disease. Rheumatol Int. 2015; 35(2): 233-41. https://doi.org/10.1007/s00296-014-3072-5

[3] Luciano N, Valentini V, Calabrò A, Elefante E, Vitale A, Baldini C, *et al*. One year in review 2015: Sjögren's syndrome. Clin Exp Rheumatol. 2015; 33(2): 259-71.

[4] Shiboski CH, Shiboski SC1, Seror R2, Criswell LA1, Labetoulle M2, Lietman TM1, *et al*. 2016 American College of Rheumatology/European League Against Rheumatism classification criteria for primary Sjögren's syndrome: A consensus and data-driven methodology involving three international patient cohorts. Ann Rheum Dis. 2017; 76(1): 9-16. https://doi.org/10.1136/annrheumdis-2016-210571

[5] Tincani A, Andreoli L, Cavazzana I, Doria A, Favero M, Fenini MG, *et al*. Novel aspects of Sjögren's syndrome in 2012. BMC Med. 2013; 11: 93. https://doi.org/10.1186/1741-7015-11-93

[6] Ramos-Casals M, Brito-Zerón P, Solans R, Camps MT, Casanovas A, Sopeña B, *et al*. Systemic involvement in primary Sjogren's syndrome evaluated by the EULAR-SS disease activity index: analysis of 921 Spanish patients (GEAS-SS Registry). Rheumatology (Oxford). 2014; 53(2): 321-31. https://doi.org/10.1093/rheumatology/ket349

[7] Mathews PM, Hahn S, Hessen M, Kim J, Grader-Beck T, Birnbaum J, *et al*. Ocular complications of primary Sjögren syndrome in men. Am J Ophthalmol 2015; 160(3): 447-452.e1. https://doi.org/10.1016/j.ajo.2015.06.004

[8] Patel SJ, Lundy DC. Ocular manifestations of autoimmune disease. Am Fam Physician. 2002; 66(6): 991-8.

[9] Rosenbaum JT, Bennett RM. Chronic anterior and posterior uveitis and primary Sjögren's syndrome. Am J Ophthalmol 1987; 104(4): 346-52. https://doi.org/10.1016/0002-9394(87)90223-6

[10] Stoffel PB, Sauvain MJ, von Vigier RO, Beretta-Piccoli BC, Ramelli GP, Bianchetti MG. Non-infectious causes of uveitis in 70 Swiss children. Acta Paediatr 2000; 89(8): 955-8. https://doi.org/10.1111/j.1651-2227.2000.tb00416.x

[11] Chebil A, Baroudi B, Slim M, Chaker N, Lamloum M, Bouladi M, *et al*. Epidemiology of uveitis in the Tunis area. J Fr Ophtalmol 2013; 36(9): 764-8. https://doi.org/10.1016/j.jfo.2013.05.006

[12] Khairallah M, Yahia SB, Ladjimi A, Messaoud R, Zaouali S, Attia S, *et al*. Pattern of uveitis in a referral centre in Tunisia, North Africa. Eye (Lond). 2007; 21(1): 33-9. https://doi.org/10.1038/sj.eye.6702111

Received on 30-09-2018 Accepted on 07-10-2018 Published on 13-12-2018

DOI: https://doi.org/10.12974/2309-6136.2018.06.04