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Membranoproliferative glomerulonephritis in a patient with primary Sjögren's syndrome:Report of a new case and review of the literature

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ABSTRACT

Glomerular involvment in patients with primary Sjögren's syndrome (SS) is extremely rare, with few cases of membranoproliferative glomerulonephritis reported in the literature . It's essentially a tubulointerstitial nephritis with some cases of glomerular nephritis reported in the literature. Membranoproliferative glomerulonephritis has been reported rarely.

Keywords : Sjogren's syndrome; membranoproliferative glomerulonephritis; nephrotic syndrome.

INTRODUCTION

Sjogren's syndrome (SS) is an autoimmune disease characterized by mononuclear cell infiltration of exocrine tissues and autoantibodies against the ribonucleoprotein particles SS-A (Ro) and SS-B (La). Primary SS (pSS) is distinguished from secondary SS by the absence of other autoimmune diseases, such as rheumatoid arthritis, systemic sclerosis, systemic lupus erythematosus[1]. Because of the different criteria for diagnosing pSS and the small groups of patients studied for renal involvement in the previous literature, the reported prevalence of renal manifestations in pSS varies widely from 2% to 67% [2,3]. Among them, tubulointerstitial nephritis resulting in tubular abnormalities compromised major renal involvement in pSS and may manifest as Type 1 renal tubular acidosis, nephrogenic diabetes insipidus, hypokalemia without renal tubular acidosis, or Fanconi syndrome [4,5]. However, glomerular involvement in patients with pSS has rarely been reported (Table 1) [6,7,8,9,10,11,12,13,14,15,16,17,18,19]. In the report, we present a case of membranoproliferative glomerulonephritis (MPGN) associated with pSS.

Table 1: : Summary of glomerulonephritis with Sjogren's syndrome reported in the literature

Reference	Glomerulonephritis	Number of cases
Chen et al. [6]	IgAnephropathy	2
Guillot et al. [7]	Membranousnephropathy	1
Stefanidis et al.[8]	Membranousnephropathy	1
Ren et al.[9]	Membranousnephropathy MPGN Focal segmental glomerulosclerosis	1 5 2
Tsai et al. [10]	IgAnephropathy	1
Watanabe et al. [11]	Membranousnephropathy	1
Bossini et al. [12]	Membranousnephropathy MPGN	1 2
Cortez et al.[13]	MPGN	1
Dussol et al. [14]	Crescentricglomerulonephritis	1
Khan et al. [15]	MPGN	1
Siamopoulos et al. [16]	Membranousnephropathy MPGN	1 1
Moutsopoulos et al. [17]	Membranousnephropathy MPGN	1 1
Yang et al. [18]	Minimalchangedisease	1
Ghannouchiet al. [19]	Focal segmentalglomerulosclerosis	1

MATERIALS AND METHODS

Case report:

A 42-year-old woman was admitted on February2012for the exploration of a generalized edema with mixed arthralgia interesting knees, shoulders and wrists and sparing the small joints and spine. She also reported a clear dry eye and mouth for 1 year. She had no history of systemic diseases and denied specific medication taken previously, including analgesics and traditionnal herbs. The general examination revealed a blood pressure of 140/90 mm Hg, pulse rate of 91 / min, and a body temperature of 37.2 ° C. Physical examination revealed obvious edema over the bilateral lower limbs with free joints. Laboratory tests revealed a normochromic anemia with an erythrocyte sedimentation rate of 120 mm the first hour, hypoprotidemia to 51g / l with hypoalbuminemia to 22 g / L without monoclonal peak or hypergammaglobulinemia in electrophoresis of plasma proteins. The daily urine protein was 5.22 g / 24h without hematuria. Serum creatinine was 11 mg / l (creatinine clearance of 89.6 ml / min according to the Cockcroft andGault) and blood urea nitrogen was 1.02 g / l. The evaluation of serum complement showed discrete low rates of C3 and C4. The immunological assessment found antinuclear antibodies (ANA) positive to 1/800 (homogeneous and speckled type) with anti-SSA Ac / Ro and anti-SSB / La positive (anti-SSA> 240 U / ml, anti SSB = 35.6 U / mL).The native anti-DNA, soluble antigens antibodies, cryoglobulinemia and rheumatoid factor were negative. Serological tests for viral hepatitis B, C and HIV were also negative. The dry eye syndrome was confirmed by a positive Schirmer test and an ophthalmologic

consultation (test with green of Lissamine). The histological examination of the salivary glands biopsy showed chronic sialadenitis stage III of Chisholm. Renal biopsy of 16 glomeruli objectified in optical microscopy MPGN type I with tubular atrophy and interstitial edema without fibrosis. The immmunofluorescent study showed diffuse deposits of C3, IgG and IgM without reaction to IgA or C1q. The treatment of dry eye syndrome consisted on artificial tears and a prescription of 10 mg of pilocarpine per day (anticholinergic). The patient was given three doses of intravenous methylprednisolone (MTP) 500 mg for three days and relay with prednisone at a dose of 1 mg / kg / day. The persistence of the nephrotic syndrome I the third week required the use of cyclophosphamide (Cyc) at a dose of 750 mg.

RESULT AND DISCUSSION

Renal involvement is one of the extraglandular manifestations of pSS, although the true prevalence and types of renal diseases are unclear. Some previous studies on renal involvement did not distinguish primary from secondary SS, making it difficult to clarify whether renal involvement contributed to pSS or to other associated disorders.

Our patient had a SS according to agreed criteria 2002 European-Americans (five out of six criteria) [1]. This syndrome is probably primitive because no other autoimmune disease associated was found.

Renal involvement in the SS is generally limited to tubules and interstitium, with a reported incidence between 5% and 70% [2]. The glomerular disease is not common and demonstrations could be hematuria, proteinuria or nephrotic syndrome. In a study by Bossini et al. [3], the incidence of glomerular lesions was 5%. In 62 patients reported by SSP Aasarod et al. [4], only one case (1.6%) of glomerular involvement was found. A higher incidence of 13.6% of glomerular lesions associated with the SSP was reported by Ren et al. [5]. It is essentially membranous nephropathy and MPGN [6]. In addition, some cases of crescentric glomerulonephritis, IgA nephropathy and focal segmental glomerulosclerosis have also been

reported [7]. The pathogenesis of glomerular damage in pSS remains uncertain. It has been reported that the clearancy the mechanisms of the glomerular mesangium and/or the reticuloendothelial system are overwhelmed by the increased levels of circulating immune complexes with consequent glomerular deposition [13].Cryoglobulins also could deposit in glomeruli and play a role in fixing complement leading to tissue injury [8]. Our patient had undetectable cryoglobulins in two laboratory tests.

Information on the treatment of MPGN in Sjogren's syndrome (Table 2), is limited and anecdotal. Moutsopoulos et al [17] reported efficacy with moderate dose steroids, 40 mg of prednisone every other day initially and 30 mg every other day for 2 years. A beneficial effect of combined steroid and cyclophosphamide (Cyc) on proteinuria and renal function has been previously described [15,16].Cortez et al [13] reported a complete and spontaneous resolution of the nephrotic syndrome of her patient, despite a very severe and aggressive-looking lesion in renal biopsy. Our patient was treated aggressively with steroids and immunosuppressive agent and yet had a complete remission of her nephrotic syndrome.

CONCLUSION

MPGN is rare. Nevertheless, we must know because a rapidly established treatment can avoid the occurrence of renal failure.

Table 2. The treatment of MPGN in Sjogren´s Syndrome as reported in the literature

Reference	Creatinine clearance (ml/m)	TUPg/24 h	Treatment	Outcome
[17]	25	1,8	Prednisone 40 mg every other day	↓TUPand ↑ creatinineclairance
[15]	60	4,9	Prednisone 60 mg /jour +Cyc 150 mg	↓TUP to 0,26g/d and ↑ creatinine clairance to85ml/mn
[16]	50	>3,5	Prednisone+Cyc	Responded well
[13]	69	5,19	No treatment	Remission
Notre cas	89,1	5,22	Bolus de MTP+Cyc	Remission

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