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Bilateral Serous Retinal Detachment in a Case with Nephrotic Syndrome

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Abstract

Purpose: *To present a case with nephrotic syndrome who developed serous retinal detachment in bilateral eyes.*

Case Report: *A 38-year-old man went to nephrology department due to bilateral leg edema and foamy urine. A diagnosis of focal segmental glomerularsclerosis with nephrotic syndrome was made and proved by kidney biopsy. Bilateral blurred vision was noted 3 months later with best corrected visual acuity (BCVA) 20/125 in right eye and 20/100 in left eye. Anterior segment and intraocular pressure were normal in both eyes. No intraocular inflammation was observed. Fundus examination showed bilateral submacular fluid. The patient was treated with diuretic, albumin supplement and steroid therapy and BCVA improved to 20/100 and 20/32 one month later. Optical coherence tomography (OCT) still revealed massive submacular fluid in both eyes. Steroid was replaced by cyclosporin A owing to intolerance of the side effect of steroid. Another one month later, BCVA improved to 20/40 and 20/25. No subretinal fluid was found by OCT in both eyes. Almost restoration of retinal anatomy was demonstrated except for some retinal pigment epithelial undulation.*

Conclusion: *Bilateral serous retinal detachment can rarely occur in patients with nephrotic syndrome. Serous retinal detachments can resolve with good visual prognosis with early recognition and proper medical treatment of nephrotic syndrome. Residual retinal pigment epithelial undulation can be found after the episode.*

Keywords: *serous retinal detachment; nephrotic syndrome; retinal pigment epithelial undulation; optical coherence tomography*

Introduction

Nephrotic syndrome is a clinical complex characterized by a number of renal and extra renal features secondary to disorders of the basement membrane of the kidney's glomerulus including proteinuria (> 3.5 gm/24hr), hypoalbuminemia (< 3.5 g/dL), edema, and hyperlipidemia. Serous retinal detachment can occur due to underlying ocular or systemic diseases, such as posterior uveitis, panuveitis, preeclampsia, etc. Permanent vision loss may develop if absence of proper medical treatmen for serous retinal detachment. However the occurrence of ocular abnormalities related to nephrotic syndrome is rarely reported. We described a case developing bilateral serous retinal detachment secondary to underlying nephrotic syndrome and focal segmental glomerularsclerosis.

Case Presentation

A 38-year-old man went to nephrology department due to bilateral leg edema and foamy urine. A diagnosis of focal segmental glomerularsclerosis with nephrotic syndrome was made and proved by kidney biopsy. Bilateral blurred vision was noted 3 months later with best corrected visual acuity (BCVA) 20/125 in right eye

and 20/100 in left eye. Anterior segment and intraocular pressure were normal in both eyes. No intraocular inflammation was observed. Fundus examination showed bilateral submacular fluid. Systemic evaluation revealed severe hypoalbuminemia (albumin, 1.3 gm/dL), and urinalysis showed 4+ proteinuria. The patient was quite healthy without any remarkable systemic disease history. He had normal HbA1c, autoimmune profiles, and not receiving any medications recently. The patient was treated with diuretic (Furosemide 40 mg per day), albumin supplement and steroid therapy (prednisolone 70 mg per day) and BCVA improved to 20/100 and 20/32 one month after medication. The spectral-domain optical coherence tomography revealed massive submacular fluid in both eyes (Figure 1). Steroid was replaced by cyclosporin A (200 mg per day) owing to intolerance of the side effect of steroid. Another one month later, BCVA improved to 20/40 and 20/25. No subretinal fluid was found by OCT in both eyes (Figure 2). Almost restoration of retinal anatomy was demonstrated except for some retinal pigment epithelial undulation.

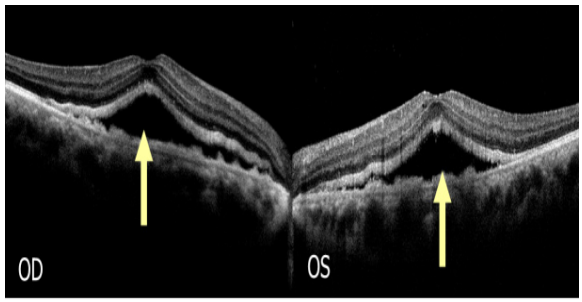

Figure 1. *The spectral-domain optical coherence tomography revealed massive submacular fluid (arrow)*

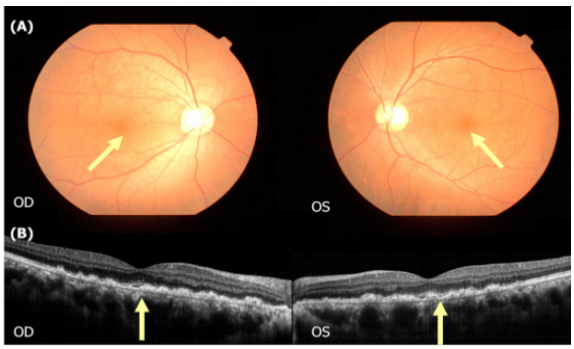

Figure 2. (A) *Color fundus showed no obvious submacular fluid in bilateral eyes under proper medical treatment 2 months later. (arrow) (B) The spectral-domain optical coherence tomography showed no submacular fluid in both eyes. Almost restoration of retinal anatomy was demonstrated except for some retinal pigment epithelial undulation.*

Discussion

Nephrotic syndrome is a group of signs and symptoms secondary to disorders of the basement membrane of the kidney's glomerulus. Classification of nephrotic syndrome can be distributed to primary or secondary causes. Primary causes are being a disease specific to the kidneys including minimal-change nephropathy, focal glomerulosclerosis, membranous nephropathy, mesangial proliferative glomerulonephritis, and rapidly progressive glomerulonephritis. Secondary causes are being a renal manifestation of a systemic general illness including diabetes mellitus, lupus erythematosus, amyloidosis, viral infections, and preeclampsia and drug toxicity. In adults, the most common cause of nephrotic syndrome is membranous glomerulonephritis, followed by focal glomerulosclerosis. Diabetic nephropathy is emerging as a major cause of nephrotic syndrome. In our patient, the cause of nephrotic syndrome should be primary because lacking of other secondary involvements.

Nephrotic syndrome is principally defined with on the basis of the presence of proteinuria (> 3.5 g/1.73 m^2/day), hypoalbuminemia (< 3.5 g/dL), hyperlipidemia, lipiduria, edema, and hypertension. Glomerular capillaries are lined by a fenestrated endothelium that sits on the glomerular basement membrane covered by glomerular epithelium, or podocytes which envelops the capillaries with cellular extensions called foot processes. In between the foot processes are the filtration slits. These three structures are the glomerular filtration barrier. Therefore in healthy individual, less than 0.1% of plasma albumin may traverse the glomerular filtration barrier. In the nephrotic syndrome, protein loss is due to glomerular proteinuria, characterized by increased filtration of macromolecules across the glomerular capillary wall. The podocyte appears to be the major target of injury in diseases that cause idiopathic nephrotic syndrome in adults and children including membranous nephropathy, minimal change disease, and focal segmental glomerulosclerosis. Because of the loss of different plasma proteins into the urine, hypoproteinemia and the reduction of plasmatic oncotic pressure with hypovolemia and sequestration of interstitial fluid in different tissues is observed in patients with nephrotic syndrome. Subsequently, secondary sodium retention induces new plasma volume expansion via an overflow mechanism that leads classic generalized edema and the interstitial accumulation of fluid in the retinal layers[1].

Ocular abnormalities directly related to nephrotic syndrome is rarely reported. Nephrotic syndrome with specific eye involvement mentioned before including Pierson syndrome, Wilms tumor-Aniridia syndrome, and congenital nephrotic syndrome. The prevalence of subretinal detachment is unknown.

Serous retinal detachments are common and may be seen in ocular as well as systemic conditions. Ocular causes include central serous chorioretinopathy, posterior scleritis, coats disease, Vogt-Koyanagi-Harada disease, sympathetic ophthalmia, focal choroidal tumor (such as malignant melanoma, hemangioma, or metastasis), and sudden hypotony due to perforation of the globe or intraocular operations. Systemic inflammatory or infectious diseases, such as sarcoidosis, cytomegalovirus infection, acute occlusion of the precapillary choroidal arterioles by fibrin-platelet thrombi, collagen vascular diseases, disseminated intravascular coagulopathy, preeclampsia, malignant hypertension, hypercortisolism, and hypoalbuminemia. The mechanisms of serous retinal detachment are thought related to choroidal vascular perfusion and permeability changes with increased choroidal interstitial fluid and further extension into the subretinal space. The diagnosis of neurosensory retinal detachment secondary to nephrotic syndrome in our case was supported by the marked improvement with oral steroids. It is essential in distinction between this case from central serous chorioretinopathy since the treatment is completely different. Treatment with systemic steroids in central serous chorioretinopathy would worsen the condition of serous detachment and the vision. In contrast, serous retinal detachment related to nephrotic syndrome would respond very well to systemic steroids.[2]. Besides, the origin of serous retinal detachment of the patient can be excluded as uveitis, choroidal tumor, or retinitis, because there was no anterior chamber or vitreous inflammation and focal choroidal or retinal abnormalities.

Table1. *Characteristics of Nephrotic Syndrome patients with retinal Detachment*

Reference	Age/ sex	Past history	At the onset of RD			Diagnosis	Treatment	RD outcom
			Edema	Protein loss	SAlb (g/dL)			
Hager A	52F	Hyper tension	Pheriphieral	Pu, 10.5g/ day	2.35	NS/ Plasmo cytoma	Steroid	Resolusion with in 6 days
De Benedetto	24F	None	Anasarca	Pu, 6.97g/ day	NA	NS/MCN	Diuretic	(OD) 20/32→20/20 (OS) 20/40→20/20
							Steroid+	Resolusion with in 5 days
Wong MHY	60F	None	Ankle	Pu, 4.8g/ day	1	NS/MCN	Diuretic+ Cyclosporin	(OD)20/100→20/40 (OS)20/70→20/30
								Resolusion with in 2 Months
	70M	VKH, Steroid	NA	Pu, 8.2 g/ day	1.4	NS/FSGS	Cyclosporin	(OD)20/400→20/200 (OS) HM 20/40 Resolusion with in 2 Months
Hasan	48/M	None	Ankle	Pu. 17g/ day	1.7	NS/AL Amyloidosis	Diuretic Steroid+	(OD)20/125→20/20 (OS) 20/50→20/20 Resolusion with in 2 Months
Current case	38M	GN	Anasarca	Pu, 3.5g/ day	1.3	NS/FSGS	Diuretic+ Cyclosporin	(OD)20/125→20/40 (OS)20/100→20/25

SAlb: *serum albumin, NA: not available, NS: nephrotic syndrome, MCN: minimal change nephropathy, VKH: Vogt-Koyonagi-Harada, FSGS: focal segmental glomerulosclerosis,*

The force that drives a net outward movement of fluid across the retina is another important contributing factor. The intraocular pressure, osmotic pressure and the active ion-fluid transport pump of the retinal pigment epithelial cells comprises this mechanism[3,4,5]. Any disruption between the osmotic pressure and intraocular pressure may result in fluid being drawn from the choroid through the retina and into the vitreous leading to retinal separation from the retinal pigment epithelial, such as Vogt-Koyanagi-Harada disease and posterior scleritis[5].

Direct damage to the ion channels on the surfaces of the retinal pigment epithelial cells in conditions like malignant hypertension and pre-eclampsia also leads to accumulation of sub-retinal fluid and exudative detachment[6]. Serous retinal detachment secondary to renal conditions has been reported among patients with hypertensive retinopathy, hemodialysis or corticosteroid therapy[7,8,9,10]. However, the patient did not have systemic hypertension.

Only 5 cases of serous retinal detachment related to nephrotic syndrome were reported (Table 1).[6-13] among these cases, the average age was 50.8 year-old (range, 24 to 70). The median levels of serum albumin and proteinuria at Serous retinal detachment presentation were 1.6 g/dL (range, 1.0 to 2.15) and 9.4g per day (range, 4.8 to 17), respectively. The pathological findings included plasmocytoma, minimal change nephropathy, focal segmental glomerulosclerosis and amyloidosis. Subretinal fluid usually resolved within 20 days in average (range, 5 days to 2 months) after treatment with steroids/ immunosuppressants / diuretics. There are 2 cases reported by De Benedetto and Hassan respectively that the subretinal fluid resolved completely with visual acuity total recovery to 20/20 within 10 days with diuretic only for treatment.[6,13] This suggests the accumulation of fluid into the choroid and supports the pathophysiologic mechanism of gradient pressure distress.[6,13] The other 2 cases required steroid or immunosupressent to resolve clinical condition.[12] Following the treatment, unexpected wavy reduction in serous retinal detachment and remodeling of the macular edema was alao reported after systemic interstitial fluid decreased.[6,13] Animal studies reported have also provided evidence of albumin and IgG in the retinal, choriocapillaris and larger choroidal vessels[14]. In conditions involving an overall decrease in serum albumin such as nephrotic syndrome, osmotic pressure in the choroidal vessels decreased, allowing the transudation of fluid into subretinal space and interstitial accumulation of fluid in the retinal layers.

We describe a case of bilateral serous retinal detachment in a young man which is an ocular complication of nephrotic syndrome, confirmed by kidney biopsy with the diagnosis of focal glomerulosclerosis. After the treatment of steroid, diuretic and cyclosporine A, general edema and subretinal fluid gradually improved within 2 months. OCT showed unique undulation of the photoreceptor's ellipsoid zone and the external limiting membrane.

Bilateral serous retinal detachment can rarely occur in patients with nephrotic syndrome. Serous retinal detachments can resolve with good visual prognosis with early recognition and proper medical treatment of nephrotic syndrome. Ophthalmologists should conduct a thorough systemic evaluation for this condition in patients with sudden-onset serous retinal detachments. Residual retinal pigment epithelial undulation can be found after the episode.

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