Bilateral Serous Retinal Detachment as a Presenting Sign of Nephrotic Syndrome

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Abstract

We herein present a case of bilateral serous retinal detachment (SRD) as a presenting sign of nephrotic syndrome (NS). A 48-year-old man complained of decreased vision related to bilateral SRD. Laboratory tests revealed NS (serum albumin, 17 g/L; proteinuria, 15.40 g over 24 hours). Following treatment for edema with a diuretic, the bilateral SRD resolved completely, with a full recovery of the patient’s vision. A kidney biopsy disclosed glomerular and vascular amyloid deposits; the amyloid stained strongly with anti-λ antiserum. Therefore, a diagnosis of AL amyloidosis was made. The sudden appearance of SRD should raise suspicion of a diagnosis of NS. Prompt recognition of this symptom is important for early treatment and restoration of the visual function.

Key words: retinal detachment, nephrotic syndrome, edema, diuretic

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Introduction

Ocular involvement is not uncommon in the practice of nephrology. It most often occurs in patients with systemic conditions (infectious or vasculitic) (1, 2) or genetic diseases (3). However, the occurrence of ocular abnormalities directly related to nephrotic syndrome (NS) is rarely reported (4-6). We herein report the case of a patient who presented with bilateral serous retinal detachment (SRD) as the initial sign of NS.

Case Report

A 48-year-old healthy man presented to the Ophthalmology service with bilateral severe visual loss lasting for six months. This symptom had been preceded by a one-month history of headache. The patient’s best corrected visual acuity was 20/125 in the right eye and 20/50 in the left eye. A slit lamp examination revealed no obvious inflammation in the anterior chamber or vitreous, while a fundus examination of both eyes showed bilateral multiple SRD in the macular and superotemporal macular regions (Fig. 1). An image of late-phase fluorescein angiography is shown in Fig. 2. Fluorescein angiography of both eyes demonstrated multifocal hyperfluorescence beneath the area of detachment in the early phase and diffuse subretinal accumulation of fluorescein in the superior area of the posterior pole in the late phase (Fig. 2). Spectral domain optical coherence tomography (OCT) indicated SRD with an increased thickness of the choroid layer in both eyes (Fig. 3). In addition, an increased choroidal thickness was clearly observed on mode B ultrasound echography. The findings of a clinical examination were unremarkable, with the exception of bilateral ankle edema. Neither neurologic, dermatologic nor dysmorphic signs were detected on a physical examination. Systemic investigations revealed severe hypoalbuminemia (albumin, 17 g/L), and a urinalysis showed 3+ proteinuria. The patient was therefore admitted to the Nephrology department for further investigations.

Upon admission, the patient’s blood pressure was 140/80 mmHg. His laboratory data were as follows: white blood cell count=5,100/μL, red blood cell count=450×10⁴/μL, hemoglobin=13.8 g/dL, hematocrit=46.1%, platelet count=
20.8×10³/μL, total serum protein=4.2 g/dL, albumin=1.7 g/dL, creatinine=1.38 mg/dL, sodium=144 mEq/L, potassium=3.8 mEq/L, chloride=109 mEq/L. The erythrocyte sedimentation rate (ESR) (10 mm/h) and C-reactive protein (CRP) (0.4 mg/dL) values were normal. The patient’s urine was 4+ for protein, and 30 red blood cells were observed per high-power field. The 24-hour urinary protein excretion was 17 g; Bence-Jones proteinuria was not present. An autoimmune screen blood profile and ultrasound of the kidneys were unremarkable. Symptomatic treatment of the patient’s edema involving a low-salt diet and the administration of a diuretic (lasix, 40 mg per day) allowed the edema to resolve after a loss of 5 kg (4.25 percent of the patient’s body weight). Within 10 days of treating the edema with the diuretic, the patient’s visual acuity improved markedly to 20/20 in both eyes. In addition, a fundus examination and OCT showed that the central macular bilateral SRD had resolved completely, although SRD remained visible in the superotempo-
Figure 4. Spectral domain OCT performed one week after the start of furosemide treatment. a: right eye, b: left eye. The central macular bilateral SRD resolved, while SRD remained visible in the superotemporal area of the posterior pole in the left eye.

Discussion

The ocular changes observed in nephrology patients are varied, occurring secondarily to direct genetic links between the ocular and kidney structures, systemic diseases and infections or the treatment of nephropathy itself. In the majority of cases, the disease is diagnosed prior to the development of ocular symptoms.

NS is a clinical syndrome with specific eye involvement, such as that involving Pioser syndrome, Wilms tumor- Aniridia syndrome and congenital NS. Strabismus, nystagmus, hypertelorism, myosis, buphthalmos and congenital glaucoma have been reported as steroid-independent ocular findings (7-16). Meanwhile, ocular abnormalities (SRD or progressive outer retinal necrosis syndrome) are classic features of NS secondary to diabetes or neoplastic disorders, such as plasmacytoma (17). However, in the present case, SRD was directly related to the intensity of NS, irrespective of its cause.

The prevalence of SRD associated with NS is unknown. The underlying mechanisms of SRD are thought to include choroidal vascular perfusion and permeability changes, which subsequently result in increased choroidal interstitial fluid with further extension into the subretinal space. These changes mostly develop during the course of systemic inflammatory or infectious diseases, such as sarcoidosis, Vogt-Koyanagi-Harada disease and cytomegalovirus infection, and in association with disorders resulting in the acute occlusion of the precapillary choroidal arterioles by fibrin-platelet thrombi. Collagen vascular diseases, disorders associated with disseminated intravascular coagulopathy, preeclampsia and malignant hypertension fall into this category. Hypercortisolism and, very rarely, malignant disease have also been implicated in the development of SRD. The causes of SRD may be classified as involving idiopathic central serous chorioretinopathy, scleritis of various etiologies (infection or autoimmune disease), diffuse choroiditis (Vogt-Koyanagi-Harada disease), focal choroidal tumoral lesions (metastasis, melanoma) or acute choriocapillaris ischemia (toxemia of pregnancy).

The occurrence of SRD secondary to renal conditions has been widely described among patients with hypertensive retinopathy and those under hemodialysis or corticosteroid therapy (18-21). However, cases of SRD related to hypoalbuminemia, like protein-losing enteropathy or NS, have been sparsely reported, with even fewer such cases with SRD as the initial finding (Table) (4-6, 22). Among these cases, the median levels of serum albumin and proteinuria at SRD presentation were 1.55 g/dL (range, 1.0 to 2.3) and 7.58 g per day (range, 4.8 to 17), respectively. The pathological findings included minimal change nephropathy, focal segmental glomerulosclerosis and AL amyloidosis, and the SRD usually resolved within weeks to months after treatment with steroids/immunosuppressants/diuretics. As for the cases reported by De Benedetto (5), the SRD resolved completely with a full recovery of visual acuity under treatment with a diuretic only. This suggests the accumulation of fluid into the choroid. Animal studies have also provided evidence of albumin in the retinal, choriocapillaris and larger choroidal vessels (23). In conditions involving an overall decrease in serum albumin, a common pathology in patients with protein-losing enteropathy and NS, the osmotic pressure in the choroidal vessels is decreased, allowing for the
transudation of fluid into the subretinal space, leading to the interstitial accumulation of fluid in the retinal layers.

The decrease in plasma colloid osmotic pressure observed in patients with NS, related to the loss of plasma proteins into the urine, results in a reduction in the plasma volume, with functional hypovolemia and the sequestration of interstitial fluid in different tissues (5). Subsequently, secondary sodium retention induces new plasma volume expansion via an overflow mechanism that induces classic generalized sodium retention. This results in a reduction in the plasma volume, related to the loss of plasma proteins in patients with NS, leading to the transudation of fluid into the subretinal space, leading to the interstitial accumulation of fluid in the retinal layers.

Following the initiation of diuretic therapy, an unexpected wavy reduction in SRD and remodeling of the macular edema may be effects of the rapid decrease in systemic interstitial fluid (5). The complete clinical and anatomic resolution of all signs and symptoms achieved with diuretic therapy alone supports this pathophysiologic mechanism of gradient pressure distress (5). However, not all patients with renal failure or gastrointestinal problems have associated complications of SRD. Indeed, NS and/or hypoalbuminuria are frequently reported in patients with other diseases and/or conditions, including diabetes, liver cirrhosis, malignancy/tumors and chronic inflammation, albeit with only very few reports of SRD (24-26).

Therefore, providing an early diagnosis and appropriate treatment is essential for securing a favorable prognosis, as SRD may be worsened by the administration of systemic corticosteroids that should be considered in eye involvement. Posterior subcapsular cataracts, glaucoma, increased intraocular pressure, pioroidodysplasia, eyelid skin atrophy, keratitis, thinning of the cornea and sclera, macular pigmentation changes, epiblapharon with inverted eyelashes and repeated hordeolum exacerbation by bacterial and viral infection are each associated with corticosteroid use (28-31).

In conclusion, bilateral SRD should be recognized as a complication of NS. Therefore, ophthalmologists should conduct a thorough systemic evaluation for this condition in patients with sudden-onset SRD.

The authors state that they have no Conflict of Interest (COI).

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