Spectral Domain Optical Coherence Tomography Findings in a Pediatric Case with Type II Membranoproliferative Glomerulonephritis

Tip II Membranoproliferatif Glomerülonefritli Pediatrik Bir Olguda Spektral Domain Optik Koherens Tomografi Bulguları

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ABSTRACT

Membranoproliferative glomerulonephritis type II (MPGN II) is a rarely encountered disease that leads to accumulation of "dense deposits" in the kidneys and chorioretinal tissues. We present the ophthalmologic and spectral domain optical coherence tomography (SD-OCT) findings of a 13-year-old boy with pathologically proven MPGN II followed up at the pediatric nephrology clinic. The patient did not have any ocular symptoms, fundus examination showed yellowish deposits and the SD-OCT demonstrated an influence on the retinal pigment epithelium – choriocapillary complex. As shown in this study, the evaluation of the findings on SD-OCT, which allows non-invasive visualization of the pathologies involving chorioretinal tissues, may be helpful in the diagnosis of the MPGN II.

Key Words: Biopsy, membranoproliferative glomerulonephritis, electron microscopy, retina, optical coherence tomography.

ÖZET

Tip II membranoproliferatif glomerülonefrit (MPGN II) böbrekler ve koryoretinal dokularda "yoğun birikintilerin" gelişimine yol açan nadir görülen bir hastalıktır. Biz pediatrik nefroloji kliniğinde patolojik olarak kanıtlanmış MPGN II tanısı bulunan 13 yaşındaki erkek çocuğa ait göz ve spektral domain optik koherens tomografi (SD-OKT) bulgularını sunuyoruz. Hastanın herhangi bir göz şikayeti bulunmamaktaydı. Fundus muayenesinde sarımsı birikintileri bulunan olguda SD-OKT incelemesi retina pigment epiteli – koryokapiller komplekste etkilenmeyi ortaya çıkardı. Bu çalışmada gösterildiği gibi, koryoretinal dokuları içeren patolojilerin non-invaziv bir biçimde görüntülenmesini sağlayan SD-OKT ile bulguların değerlendirilmesi MPGN II tanısında yardımcı olabilir.

Anahtar Kelimeler: Biyopsi, membranoproliferatif glomerülonefrit, elektron mikroskobu, retina, optik koherens tomografi.

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INTRODUCTION

Membranoproliferative glomerulonephritis type II (MPGN II) is a rare disease which is characterized by the deposition of an abnormal electron-dense material on the basement membranes of renal glomeruli.[1] The preliminary findings of the disease generally include proteinuria, hematuria and renal impairment.[2] Another tissue that might be influenced during the course of the disease, however, is the retinal pigment epithelium (RPE). Histopathologic changes in the RPE occurring as part of the disease spectrum of MPGN II have been reported by Duvall-Young et al3 for the first time. The authors reported that the dense deposits accumulating in the glomerular capillary basement membrane and the Bruch's membrane have a similar structure.[3]

There are a few studies in the literature that document the retinal changes seen in adult cases with MPGN II.[1,8,9] However, to the best of our knowledge, there are no reports that describe the high-definition morphological changes affecting the chorioretinal vasculature in pediatric MPGN II cases. Herein, we evaluate a pediatric MPGN II case by means of spectral domain optical coherence tomography (SD-OCT).

CASE REPORT

A 13-year-old boy without any ocular symptoms consulted the ophthalmology clinic for hypertensive retinopathy screening. He was being followed up at the pediatric nephrology clinic with a biopsy proven diagnosis of MPGN II for approximately five years. The histopathologic examination of his renal biopsy samples showing changes consistent with MPGN II are depicted in Figure 1.

A comprehensive ophthalmic examination revealed that his best-corrected visual acuity was 10/10 in both eyes. The anterior segments were unremarkable, and his intraocular pressures were within normal limits in both the eyes. The fundus examination revealed yellowish deposits, which were irregularly distributed within the macular area in both eyes. Because of the renal insufficiency associated with MPGN II, a fundus fluorescein angiography (FFA) could not be performed. SD-OCT (RTVue-100, Optovue Corp., Fremont, CA) imaging showed irregularities and undulations in the contour of the RPE, multiple serous elevations of the RPE and small deposits within the RPE/Bruchs membrane complex in his both eyes (Figure 2). The ganglion cell complex and the retinal nerve fiber layer appeared to be normal.

DISCUSSION

MPGN II, a rarely encountered disease, forms approximately 20% of all MPGN types diagnosed in childhood. Although the etiopathogenesis of the disease is yet to be determined precisely, it is thought that uncontrolled systemic activation of the alternative pathway of the complement cascade plays a role in the development of the disease. The pathognomonic morphological characteristic of the disease is the presence of electron dense deposits within the glomerular basement membrane on electron microscopy. In many cases, deposits having similar composition and structure are also seen in the choriocapillaris-Bruch's membrane-RPE interface. This ocular area morphologically resembles the capillary tuft of the glomerular basement membrane-glomerular epithelial interface. In this disease, spontaneous remissions are quite rare, end-stage renal disease might develop in most of the cases and deterioration in visual acuity develops in some of the cases.[2,4]

Among MPGN, it was reported that the ocular findings develop only in subjects with type II disease. Accordingly, it is also extremely important to determine the clinical and pathological features specific to MPGN II in the chorioretinal vasculature. Deposits developing in the eye in MPGN II ophthalmoscopically resemble drusen and can be associated with RPE alterations. In the advanced stages of the disease, however, RPE detachments and choroidal neovascularization that might lead to vision loss might occur.[5]

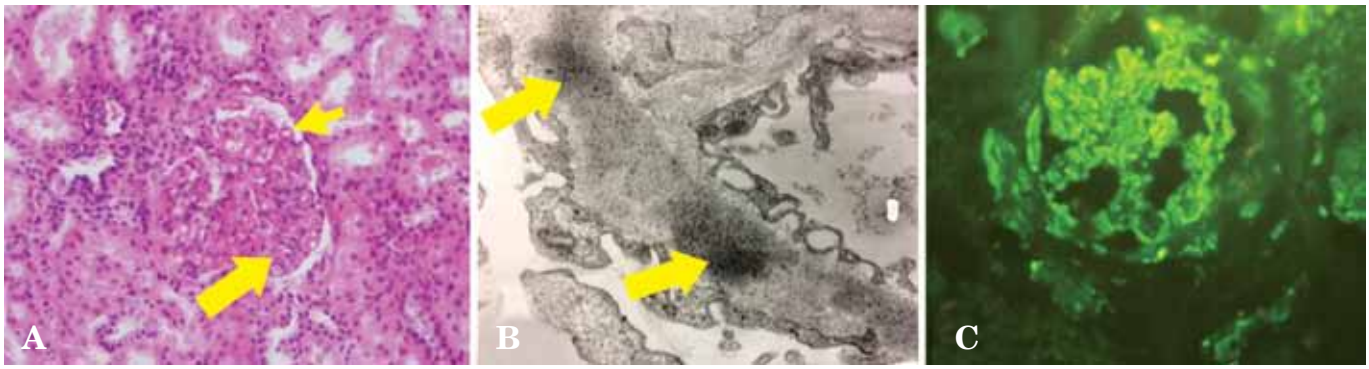

Figure 1a-c: *Renal biopsy samples of the case diagnosed with MPGN II: A. HE X 400: Endocapillary cell proliferation (arrow) and diffuse global thickening in the capillary basement membrane (small arrow) of the glomerulus: B. EM x 10.000: Electron dense deposits in the mid zone of the capillary basement membrane of the glomerulus. Their locations are neither subepithelial nor subendothelial (arrows). C. Immunofluorescence x anti-C3 ab: Intense staining for complement (C3) in the glomerulus.*

Figure 2: *Fundus images of both eyes (those at the top) and spectral domain optical coherence tomography examination images (those at the bottom) of the case diagnosed with MPGN II (The right fundus image has stain artifacts). Irregularities and undulations in the contour of the retinal pigment epithelium (RPE) and multiple serous elevations of the RPE (arrows) with small deposits (arrowheads).*

In a study describing ocular findings from 20 cases of MPGN II, drusenoid images were found in the fundus examination of only one out of the four pediatric cases, whereas ocular symptoms were not found in any of four pediatric patients between the ages of 9-17.5 It was found that the ocular findings in MPGN II are related to the duration of the disease rather than the severity of the renal involvement.[5] For this reason, visual acuity and visual fields can be preserved in the initial part of the disease. However, FFA and specialized tests of retinal function, such as electroretinography and electrooculography, might reveal early damage, and clinical worsening may develop in the progressive periods of the disease.6 However, there are difficulties in the application of the above-mentioned tests to pediatric patients. Although there were no ocular symptoms or visual findings in the case that we presented, RPE changes ophthalmoscopically resembling drusen were noticed. To understand such changes better, FFA and the other special tests mentioned could not be performed, considering the current clinical condition and the age of our patient.

SD-OCT is a noninvasive and objective cross-sectional tissue imaging technique that shows promise in early detection of the pathologic changes that occur in the retinal layers. The application of this imaging technique in the pediatric age group is highly successful as well.[7] In our study, SD-OCT was also successful in revealing the ocular features related to MPGN II.

In the literature, several OCT studies document the retinal morphological changes in eyes with MPGN II.[1,8,9] Batioglu et al.,[8] found irregularities and modulation in the contour of the RPE, along with multiple small, serous elevations of the RPE in areas of retinal drusenoid lesions. In the same study, it was reported that the lack of optical shadowing below these contour changes might be compatible with drusenoid deposits occurring under the neurosensory retina and the Bruch's membrane.[8] In another study, Venkatesh et al.[9] reported RPE irregularities, indicating affection of the RPE-choriocapillary complex, excrescences most likely representing electron dense deposits and findings of small serous RPE elevations in a case with MPGN II. In the same study, the finding of a slit-like optically clear area adjacent to the area of RPE detachment, suggesting subretinal fluid collection, was also reported.[9] Similar findings were also detected by Ritter et al.,[1] Though different from the above-mentioned studies, similar OCT findings were also obtained in our study in which a pediatric case is addressed.

In conclusion, SD-OCT is beneficial in the early diagnosis of ocular findings of pediatric patients with MPGN II and in the monitoring and treatment of ocular complications, such as RPE detachments and choroidal neovascularization, that might develop in the future. Furthermore, SD-OCT can be successfully used in the monitoring of MPGN II as an alternative to FFA examination, which is an invasive operation that is difficult to apply in the pediatric age group and especially in such patients with renal insufficiency.

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