Bilateral Acute Retinopathy with Vitreous Hemorrhage as a First Manifestation of Sarcoidosis: Case Report

Sarkoidozun İlk Belirtisi Olarak Vitreus Kanamalı Bilateral Akut Retinopati

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ABSTRACT Acute retinopathy and vitreous hemorrhage (VH) are very rare ocular manifestations of sarcoidosis. A 42-year-old male patient complained of blurred vision and floaters in his left eye for three weeks. On the ophthalmologic examination, bilateral acute retinopathy and VH were observed. Superotemporal branch retinal vein occlusion (BRVO), retinal fibrovascular membrane and VH in the left eye were noted via fundus fluorescein angiography. Mediastinal lymphadenopathies were detected during systemic evaluation of the patient. Transthoracic lymphoid biopsy from the mediastinal region, confirmed the diagnosis of sarcoidosis histologically. Although the patient was treated with oral steroids for sarcoidosis, he underwent pars plana vitrectomy and membranectomy due to progressive VH and tractional fibrovascular membrane. Sarcoidosis should be considered within the differential diagnosis list in patients with bilateral acute retinopathy, BRVO, and VH. Improved outcomes can be achieved through early recognition of the underlying disease and appropriate therapy.

Key Words: Retinal vein occlusion; sarcoidosis; vitreous hemorrhage; vitrectomy

ÖZET Akut retinopati ve vitreus kanaması (VK), sarkoidozis' in oldukça nadir oküler belirtilerindendir. Kırk iki yaşında erkek hasta, sol gözünde 3 haftadır bulanık görme ve uçuşmadan şikâyet etmekteydi. Oftalmolojik muayenede, bilateral akut retinopati ve sol VK izlendi. Sol gözünde üst temporal ven dal tıkanıklığı ve fibrovasküler membran, VK floresein anjiyografi ile belirlendi. Bu olgunun sistemik değerlendirmesi sonunda, mediyastinal lenfadenopatiler saptanmış olup, mediyastinal bölgeden transtorasik yaklaşım ile lenfadenopati örnekleri alınmış ve böylece histopatolojik olarak sarkoidoz tanısı konmuştur. Hastaya sarkoidoz için sistemik steroid tedavisi uygulanmasına rağmen, ilerleyen VK ve traksiyonel fibrovasküler membran sebebi ile pars plana vitrektomi gerçekleştirilmiştir. Sarkoidoz, bilateral akut retinopati, retina dal tıkanıklığı ve VK olduğu durumlarda etyolojide düşünülmeli ve gözardı edilmemelidir. Altta yatan hastalığın erken tanınması ve uygun tedavisi ile iyi sonuçlar alınabilmektedir.

Anahtar Kelimeler: Retina ven tıkanıklığı; sarkoidoz; vitreus kanaması; vitrektomi

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Sarcoidosis is a multisystem granulomatous disease of unknown etiology, described for the first time in 1878 by Jonathan Hutchinson.[1] It often affects multiple organs, including the skin, nervous system, lung, liver, and eye.[2] Ocular manifestations are frequent in 26-50% of sarcoidosis patients and may occur without apparent systemic involvement.[3] Uveitis is the most familiar ocular manifestation (30-70%) and is potentially vision threatening.[4] Posterior segment manifestations of sarcoidosis are periphlebitis cuffing, diffuse infiltrates, perivenous sheath-

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ing, posterior uveitis, and optic neuropathy.[5-7] Rarely, capillary occlusion and ischemia lead to neovascularization and vitreous hemorrhage (VH).[8,9]

In this study, we presented a case of sarcoidosis with a rare etiology that had acute retinopathy findings, VH due to retinal vein occlusion, and a tractional epiretinal membrane.

CASE REPORT

A 42-year-old patient complained of blurred vision and floaters in the left eye for about three weeks. Ophthalmological examination revealed that the eyes were orthotropic, and the eye movements were free in every direction. Visual acuities were 20/20 in the right eye and 30/200 in the left eye. Intraocular pressures measured by applanation tonometry were 16 mmHg in the right eye and 19 mmHg in the left eye. There was the mild flare, but no iris nodules or cells in the anterior chamber. The vitreous humor showed no cells in the right eye, but red blood cells were detected in the left eye. Funduscopic examination showed perivascular sheathing along with scattered preretinal hemorrhages and retinal venous tortuosity in both eyes. Besides moderate VH, superior temporal retinal vein occlusion and a fibrovascular membrane were observed in the left eye. Capillary leakage, micro aneurysm and new vessel formation were also confirmed by angiography (Figure 1). On physical examination, blood pressure was 130/70 mmHg. The lungs were clear and resonant, and there was no hepatomegaly. Neurological examination showed no abnormalities. Laboratory studies revealed a hematocrit reading of 35.7% and a hemoglobin value of 12.6 g per 100 mL. The white blood cell count was 6500 per mm^3, with a

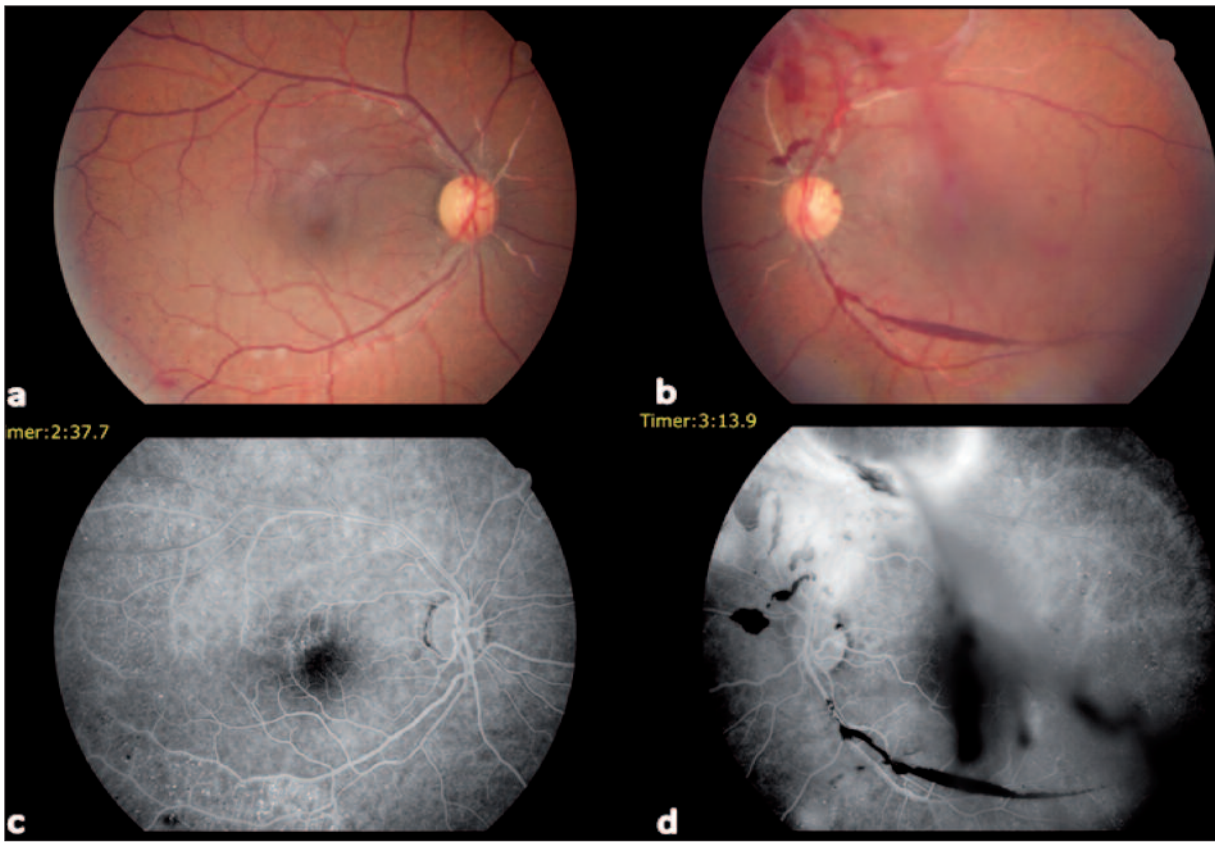

FIGURE 1: On examination of the fundus, perivascular sheathing was observed, along with scattered preretinal hemorrhages, retinal venous tortoise, similar to acute retinopathy findings' in both eyes, vitreous hemorrhage, superior temporal retinal vein occlusion and a fibrovascular membrane in the left eye. Capillary leakage, microaneurysms and new vessel formation were also confirmed by angiography.
(See color figure at http://www.turkiyeklinikleri.com/journal/oftalmoloji-dergisi/1300-0365/)

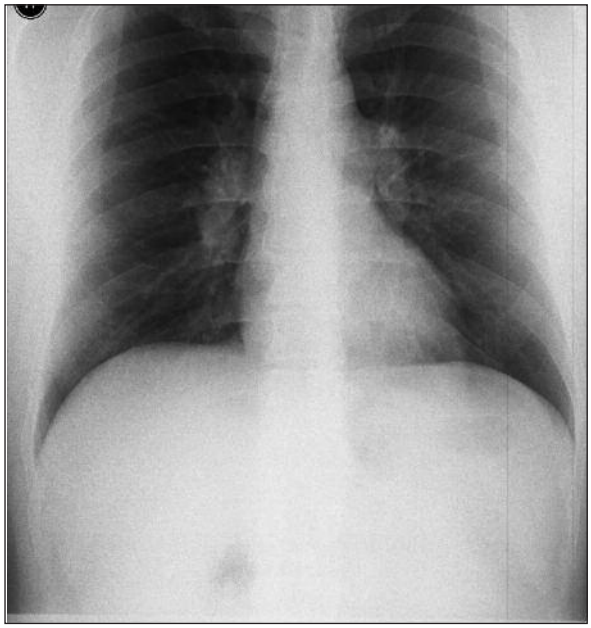

FIGURE 2: Chest X-ray demonstrated bilateral hilary lymphadenopathy.

normal differential cell count. The platelets were $19x10^4$ per mm^3. A chest Xray demonstrated bilateral hilary lymphadenopathy (Figure 2). Respiratory function tests, angiotensin-converting enzyme (ACE) and hepatic enzyme (aspartate transaminase and alanine transaminase) levels were within normal limits. Right paratracheal, aortic pulmonary window, subcarinal, and bilateral hilary lymph nodes were determined as hyperplastic by positron emission tomography, the largest being 10x5 mm (Figure 3). Samples from bronchoscopic lavage and biopsy were determined as normal. Mediastinal lymph node biopsy was taken from the bronchoalveolar space via a transthoracic approach (mediastinal biopsy), and sarcoidosis was histologically diagnosed (Figure 4). Informed consent was obtained from the patient for all procedures.

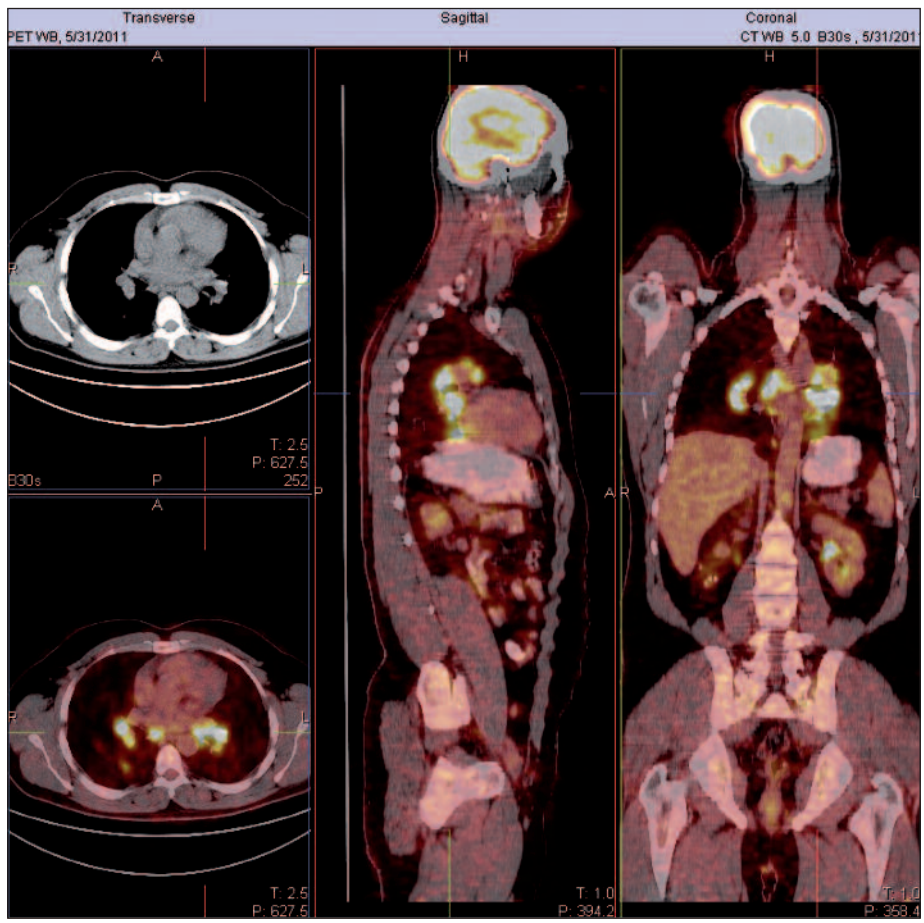

FIGURE 3: Right paratracheal, aortic pulmonary window, subcarinal, and bilateral hilary lymph nodes were determined as hyperplastic by positron emission tomography.
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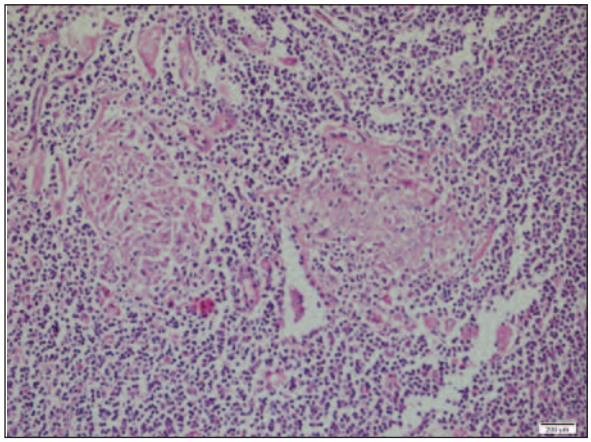

FIGURE 4: Small sarcoid granulomas were detected in lymphoid tissue (Hematoxylin eosin staining, magnified 200 times; HE, x200).

The patient received prednisone 60 mg per day orally for the treatment of systemic sarcoidosis. Cycloplegic eye drops twice daily, and prednisolone acetate (1%) drops four times daily were applied for flare in the anterior chamber of the left eye. After one month, the patient underwent vitrectomy surgery owing to increased VH (Figure 5). Our patient was followed up for six months.

DISCUSSION

Sarcoidosis is a multisystem disorder and can involve multiple organs. Ocular involvement in sarcoidosis may occur at any time during the condition. In about 80% of the cases, ocular manifestations appear before or within one year of the onset of systemic signs of sarcoidosis.[10]

Posterior segment lesions are reported in about 14-28% of patients with ocular sarcoidosis and include vitreitis, chorioretinitis, periphlebitis, vascular occlusion, retinal neovascularization, and optic nerve head granuloma.[11,12] Sarcoidosis optic neuropathy is rare but can cause permanent blindness.[6] Peripheral retinal neovascularization is generally seen in patients with a defined vasoocclusive disorder, such as a branch retinal vein occlusion, and may even simulate a sea fan, similar to that seen in sicklecell disease. There are several reports that retinal neovascularization developed due to sarcoidosis, but only one case with persistent VH has been declared in the literature.[13-15] Sanders and Shilling have described an "acute retinopathy of sarcoidosis" with extensive perivascular sheathing, vascular occlusion, and intraretinal hemorrhages.[16] In the present case, there were comprehensive perivascular sheathing, intraretinal hemorrhages, and vascular occlusion consistent with the 'acute retinopathy' defined by Sanders and Shilling. Abnormalities on chest radiographs are found in 90% of patients with pulmonary sarcoidosis, including bilateral hilary and mediastinal lymphadenopathy. Serum ACE levels elevated in 65% of sarcoidosis patients; however, normal levels do not exclude the diagnosis of sarcoidosis.[17,18] A tissue biopsy is

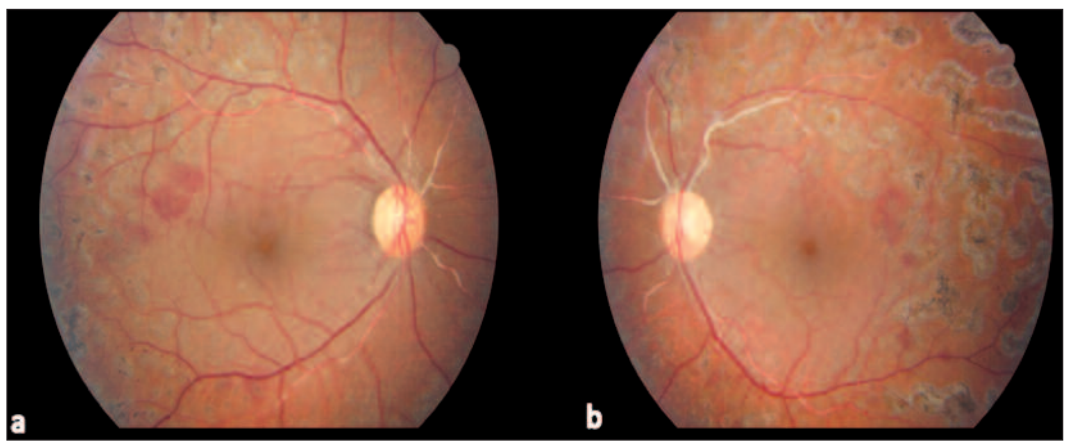

FIGURE 5: Vitrectomy and membranectomy surgery was performed following one month because of persisted vitreous hemorrhage in the left eye.
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needed for a definitive diagnosis of sarcoidosis and can usually be performed from an appropriate site, except for intraocular tissue due to the high risk of vision loss.[19] The lung is the preferred biopsy site when it is involved. In our case, the result of endobronchial lung biopsy was negative, but bronchoalveolar space biopsies were positive.

In conclusion, although it is not common, atypical ocular manifestations can evolve in the early stages of sarcoidosis. It should be considered in the etiology and should be ruled out in patients with acute retinopathy or branch retinal vein occlusion, or VH. Improved outcomes can be achieved with recognition of the underlying disease and appropriate therapy.

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