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HYPERTENSIVE OPTIC NEUROPATHY AS A PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

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Abstract

***Introduction**: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease affecting multiple organ systems, including the eyes. Hypertensive optic neuropathy is a rare but serious manifestation of SLE that may precede SLE diagnosis. This case highlights the early ocular involvement in a young patient with undiagnosed SLE.*

***Case Report**: A 21-year-old female presented to our ophthalmology clinic with blurry vision in both eyes, along with elevated blood pressure at 165/126 mmHg. Ophthalmic examination revealed visual acuity of 20/200 on both eyes, bilateral optic disc swelling, macular edema, flame-shaped hemorrhages, and cotton-wool spots. These findings were consistent with hypertensive optic neuropathy. The antihypertensive drugs were initiated and planned for intravitreal bevacizumab administration. The patient was referred to internal medicine. Laboratory tests and clinical findings indicated secondary hypertension, anemia, leukopenia, and thrombocytopenia. Immuno-serological testing confirmed a diagnosis of SLE. The patient was initiated on systemic immunosuppressive therapy. Eight weeks after the first visit, she showed significant improvement, with the resolution of macular edema and optic disc swelling. The patient's visual acuity improved to 20/50 in the right eye (RE) and 20/20 in the left eye (LE).*

***Discussion**: Hypertension in SLE is multifactorial, involving endothelial dysfunction, kidney injury, immune activation, and autoantibodies. Hypertensive retinopathy progresses through three phases: vasoconstrictive, sclerotic, and exudative, characterized by arterial narrowing, structural vascular changes, and blood-retina barrier disruption, leading to macular edema and ischemia. Diagnosis is based on fundoscopic examination and Optical Coherence Tomography (OCT), while management focuses on blood pressure control, anti-VEGF therapy, and close monitoring to prevent further complications.*

***Conclusion**: Early detection and multidisciplinary management are crucial in preventing irreversible visual loss and systemic complications. Regular ophthalmic follow-up and blood pressure monitoring are essential in SLE management.*

Keywords: hypertensive optic neuropathy, hypertensive retinopathy, macular edema, lupus nephritis, systemic lupus erythematosus, anti-VEGF **Cite This Article:** ZAINI, Lia Meuthia; MULYA, Putri Nabillah. HYPERTENSIVE OPTIC NEUROPATHY AS A PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS. **International Journal of Retina**, [S.l.], v. 8, n. 2, sep. 2025. ISSN 2614-8536. Available at: <https://www.ijretina.com/index.php/ijretina/article/view/323>. Date accessed: 30 sep. 2025. doi: https://doi.org/10.35479/ijretina.2025.vol008.iss002.323.

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INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease driven by antibodies that can impact nearly every organ in the body. Its clinical manifestations are diverse, involving the skin, kidneys, musculoskeletal, and nervous systems. Ocular involvement can affect the external eye, the anterior segment, the retina, the choroid, or present as a neuro-ophthalmological manifestation. These ocular manifestations are not included in the diagnostic criteria established by the 2019 European League Against Rheumatism/American College of Rheumatology (EULAR/ACR).[1] However, severe complications are relatively common, and treatment becomes significantly more challenging or less effective once ocular involvement progresses to advanced stages. Therefore, periodic ophthalmological reviews are recommended in all patients with lupus.[2]

Among ocular manifestations, retinal changes are particularly common and are often used as clinical indicators of disease activity.[3] Retinopathy in SLE can be caused either as a direct consequence of SLE-related inflammation or as a secondary complication, such as lupus nephritis-induced hypertension. The prevalence of retinopathy ranges from 3% in well-controlled patients to 29% in those with more active systemic disease.[4] Here, we report a case of hypertensive optic neuropathy presented to our ophthalmology clinic before the diagnosis of SLE. This highlights the importance of early detection and management of hypertensive complications in SLE, as they can significantly impact visual prognosis.

CASE REPORT

A 21-year-old Acehnese female presented to our ophthalmology clinic with complaints of bilateral blurry vision since the previous day. There were no other ophthalmic symptoms, such as scotoma, red eye, pain, diplopia, proptosis, or dryness. Additionally, the patient reported persistent headaches, myalgia, hair loss, and rashes on her ears for the past three weeks. She had a history of wearing spectacles with a prescription of -0.5 diopters in both eyes. Neither the patient nor her family had a history of autoimmune disease. There was no history of recent medication use. On initial examination, her blood pressure was elevated at 165/126 mmHg, with a heart rate of 89 beats per minute and a respiratory rate of 22 breaths per minute. The remainder of her systemic physical examination was unremarkable. Ophthalmological evaluation revealed a visual acuity of 20/200 in the RE and the LE. Intraocular pressure was within normal limits. A dilated fundus examination (Figure 1) revealed bilateral optic disc swelling, with a cup-to-disc ratio of 0.3 in both eyes. The macula appeared elevated, with no foveal reflex surrounded by exudates with star configuration. Optical coherence tomography (OCT) (Figure 2) showed increased central macular thickness. Numerous flame-shaped hemorrhages and cotton-wool spots were observed. A diagnosis of hypertensive optic neuropathy was made, and hypertensive medication was prescribed. Intravitreal bevacizumab was recommended for both eyes to treat macular edema.

Based on her symptoms and physical findings, the patient was referred to the internal medicine department for further evaluation of the underlying cause of hypertension. Laboratory tests, as shown in Table 1, revealed hemoglobin: 10.2 g/dL, white blood cells: 3,390/mm^3,

platelet count: 94,000/mm^3, elevated D-dimer at 2,140 ng/mL, low HDL at 37 mg/dL, and elevated LDL at 124 mg/dL. Kidney function tests were normal, and the renal ultrasound was unremarkable. She was treated with nifedipine 30mg, bisoprolol 5mg, and rivaroxaban 10mg once daily. While undergoing regular follow-up at the ophthalmology and internal medicine clinics, the patient developed anemia three weeks later, with a hemoglobin level of 8 mg/dL and was hospitalized for further management. She received a packed red cell transfusion and was started on intravenous furosemide 40 mg and methylprednisolone 125 mg, followed by oral methylprednisolone 8 mg three times daily, spironolactone 25 mg twice daily, and terazosin 2 mg once daily. Further workup was done, including an antinuclear antibodies (ANA) profile test and urinalysis. Immuno-serological tests showed high anti-double-stranded DNA levels (512.9 IU/mL) and a positive antinuclear antibody profile, including double-stranded DNA, nucleosomes, histones, and ribosomal protein. Complement and antiphospholipid antibody tests were not performed. Urinalysis showed positive leukocytes, protein, and granular and hyaline casts. These findings confirmed the diagnosis of systemic lupus erythematosus with lupus nephritis. The patient was treated with methylprednisolone, hydroxychloroquine, cyclosporine, nifedipine, calcium carbonate, clopidogrel, furosemide, terazosin, and valsartan.

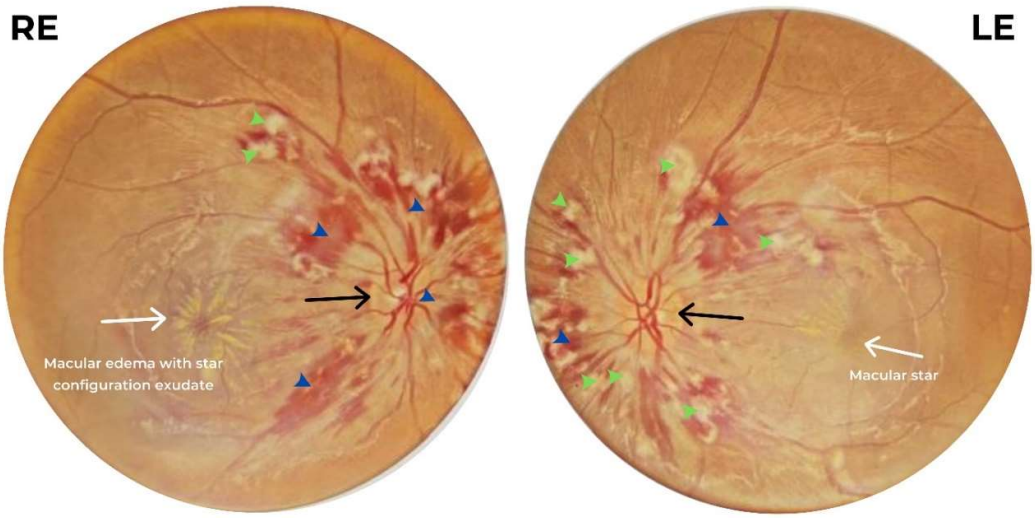

Figure 1. *Fundus photograph of the patient at the initial presentation. Optic disc edema (black arrow). Macular edema with 'star' exudate configuration (white arrow). Retinal hemorrhages (blue arrow). Cotton-wool spots (green arrow)*

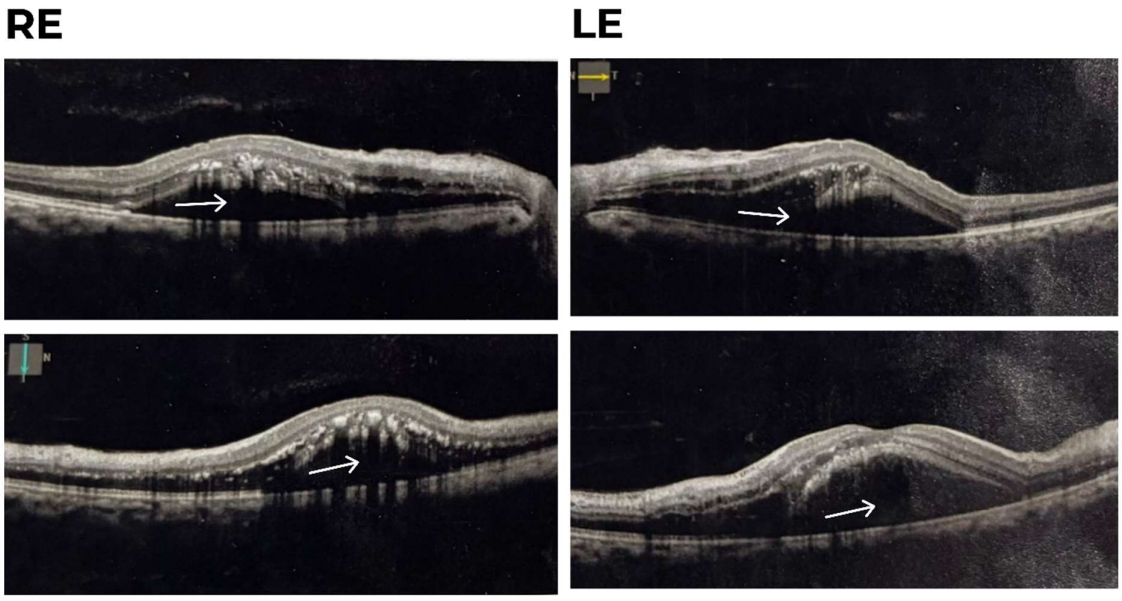

Figure 2. *Optical Coherence Tomography of the patient at the initial presentation. Increased central macular thickness associated with subretinal fluid and exudation (white arrow).*

Table 1. *Laboratory Findings*

Laboratory	Results	Reference Range
Hematology		
Hemoglobin	10,2 g/dL	12,0-15,0 g/dL
Hematochrits	29 %	37-47 %
Leukocytes	3,39 x 10^3/mm^3	4,5-10,5 x 10^3/mm^3
Thrombocytes	94 x 10^3/mm^3	150-450 x 10^3/mm^3
Ureum	25 mg/dL	13-43 mg/dL
Creatinine	0,86 mg/dL	0,51-0,95 mg/dL
Erythrocytes Sedimentation Rate	12 mm/hour	<20 mm/hour
C-Reactive Protein	negative	negative
Rheumatoid Factor	negative	negative
D-dimer	2140 ng/mL	<500 ng/mL
Urinalysis		
Leukocytes	25-50	0-2
Erythrocytes	25-50	0-2
Epithelial Cells	10-25	0-1
Protein	positive (+2)	negative
Keton	positive	negative
Granular Cast	positive	
Hyaline	positive	
Bacteria	positive	
Immunoserology		
Anti-double-stranded DNA	512,9 IU/mL	<100 IU/mL
Nucleosome (NUC)	+++	
Histones (HI)	+	
Ribosomal Protein (RIB)	+++	
Control (Ko)	+++	

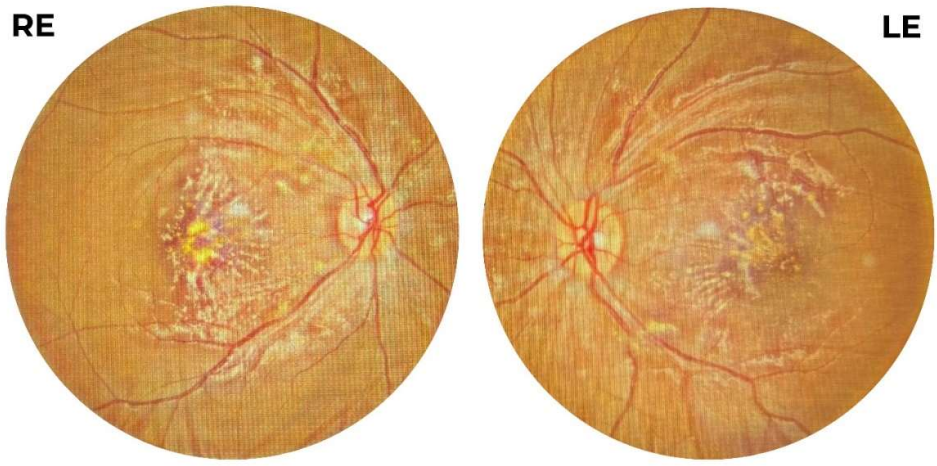

Figure 3. *Eight-week follow-up after the first visit. The papilledema and retinal hemorrhages had resolved, leaving a star-shaped exudate.*

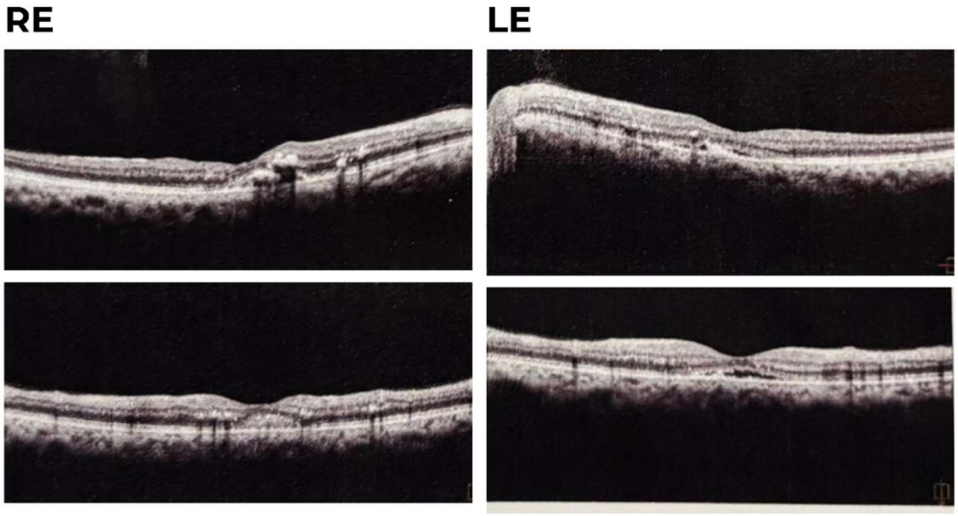

Figure 4. *Optical Coherence Tomography of the patient after the administration of intravitreal bevacizumab. Reduction in central macular thickness.*

At the eight-week follow-up after the first visit, the patient showed significant improvement. The papilledema had resolved, leaving star-shaped exudates, as shown in Figure 3. The macular edema had also resolved, as confirmed by OCT, which demonstrated a reduction in central macular thickness, as shown in Figure 4. Fundus examination revealed a marked reduction in optic disc swelling, with the resolution of flame-shaped hemorrhages and cotton-wool spots. The macular contour appeared more defined, and the foveal reflex was restored. The patient also reported subjective improvement in visual acuity. Visual acuity after therapy showed improvement to 20/50 in the right eye and 20/20 in the left eye. Blood pressure was better controlled with antihypertensive therapy, and systemic lupus erythematosus treatment was continued under the supervision of the internal medicine department. Regular follow-up was scheduled to monitor disease progression and prevent recurrence.

DISCUSSION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that predominantly affects women. Ophthalmic involvement occurs in approximately one-third of SLE patients, either as a direct consequence of SLE-related inflammation or as a secondary complication such as lupus nephritis-induced hypertension.[4] Lupus nephritis is one of the most frequent complications of SLE, occurring in approximately two-thirds of patients and plays a significant role in determining the disease's overall prognosis.[5] Retinopathy, another complication, serves as an indicator of severe disease, often accompanied by renal impairment and poor outcomes. Thus, early detection of subclinical retinopathy is crucial, as it is closely linked to nephropathy in SLE patients.[6] In this case, the ophthalmic findings were consistent with hypertensive optic neuropathy, which are rare but serious manifestations of hypertensive crisis in SLE patients. Studies have reported that individuals with SLE tend to have higher blood pressure, with prevalence rates varying by population. Notably, young women with SLE have a strikingly high prevalence of hypertension, with some studies reporting rates as high as 40%, compared to only 11% in age-matched controls.[7,8] Hypertension in SLE is driven by multiple factors, including endothelial dysfunction, kidney damage, immune activation, and autoantibodies like anti-dsDNA. As the disease advances, vascular inflammation and immune cell infiltration contribute to rising blood pressure, with neutrophil activity and chromatin release exacerbating blood vessel damage and worsening hypertension.[9] Malignant hypertension can lead to three primary types of ocular damage: choroidopathy, retinopathy, and optic neuropathy.[10] Secondary arterial hypertension in SLE, often due to autoimmune kidney inflammation, disrupts retinal microcirculation by causing arterial wall stiffening and impaired blood flow regulation.[11] This patient's grade 4 hypertensive retinopathy with macular edema resulted from acute and chronic blood pressure elevation.

The stages of hypertensive retinopathy include vasoconstriction, sclerosis, and exudation as the disease advances. The vasoconstrictive phase is the initial response to elevated blood pressure, leading to vasospasm and narrowing of retinal arterioles as part of an autoregulatory mechanism to optimize blood flow. Clinically, this phase is characterized by generalized or focal narrowing of the retinal arteries and a reduced artery-to-vein ratio. As hypertension persists, structural changes occur, including intimal thickening, medial wall hyperplasia, and hyaline degeneration, which define the sclerotic phase. This stage is marked by arteriovenous nicking- where thickened arterioles compress the underlying venules- and an accentuated vessel wall light reflex, described as copper or silver wiring. The exudative phase occurs in severe systemic hypertension and is characterized by blood-retina barrier breakdown, leading to flame-shaped and dot-blot hemorrhages, hard exudates, and cotton-wool spots, which indicate microvascular infarction of the retinal nerve fiber layer. In extreme cases, malignant hypertension can cause increased intracranial pressure, leading to ischemic optic neuropathy and optic disc swelling.[12] The pathophysiology of macular edema in hypertension involves a complex interplay of vascular dysfunction, increased capillary

permeability, and inflammatory mediators. Hypertension disrupts the blood-retina barrier through endothelial cell dysfunction, leading to plasma protein and fluid leakage into the retinal layers. Elevated systemic blood pressure increases capillary hydrostatic pressure, resulting in further vascular leakage. Additionally, oxidative stress and inflammation in hypertension promote the release of vascular endothelial growth factors (VEGF), exacerbating vascular permeability. Retinal ischemia and hypoxia further contribute to macular edema by triggering compensatory neovascularization, which increases fluid accumulation. These mechanisms underscore the necessity of early blood pressure control and the use of targeted ocular therapies such as anti-VEGF agents to manage hypertensive macular edema.[13]

The diagnosis of hypertensive retinopathy is primarily based on a fundoscopic examination, which reveals characteristic retinal changes in hypertensive patients. Hypertension is diagnosed when systolic blood pressure (SBP) exceeds 140 mmHg and/or diastolic blood pressure (DBP) is over 90 mmHg.[14] Identifying and managing hypertension in SLE patients is crucial, as uncontrolled blood pressure can lead to severe ocular and systemic complications. The long-term prognosis of hypertensive retinopathy in patients with SLE depends on timely intervention and effective management of systemic hypertension. Recurrence risk is heightened in cases of poor blood pressure control and SLE flares. Long-term monitoring strategies include regular ophthalmic examinations with fundus photography and OCT to detect early signs of recurrence, optimizing antihypertensive therapy, controlling systemic inflammation with immunosuppressive agents, and educating patients about symptoms that may indicate worsening ocular involvement.[15]

CONCLUSION

Hypertensive optic neuropathy can be an initial manifestation of systemic lupus erythematosus. This case highlights the importance of considering autoimmune diseases in young patients presenting with severe hypertensive retinopathy, particularly in the absence of a prior history of hypertension. Timely detection and a collaborative approach are essential to prevent permanent vision loss and avoid systemic complications. Further research is needed to establish standardized guidelines for the ophthalmologic screening of SLE patients, particularly those at high risk for hypertensive complications.

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