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Ocular Involvement as the Primary Presentation of Suspected Systemic Lupus Erythematosus: A Case of Bilateral Papilledema and Macular Edema

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ABSTRACT

Background: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease known for its varied clinical presentations, often affecting multiple organ systems. Ocular involvement is common in SLE, but posterior segment manifestations like papilledema are rare, occurring in only about 1% of SLE patients. This case report details a unique instance of bilateral papilledema and macular edema as the primary presentation of suspected SLE in a young female patient. **Case presentation:** A 24-year-old female presented with a one-week history of sudden-onset blurry vision in both eyes. Ophthalmological examination revealed reduced visual acuity (6/45 in both eyes), bilateral optic disc swelling, and macular edema. Further investigations, including Optical Coherence Tomography (OCT) and Magnetic Resonance Imaging (MRI), confirmed macular edema and optic nerve sheath distention. A positive Antinuclear Antibody (ANA) test suggested an autoimmune etiology. Lumbar puncture results were normal, ruling out idiopathic intracranial hypertension. The patient was diagnosed with bilateral papilledema and macular edema, with suspected underlying SLE. Prompt treatment with high-dose corticosteroids and acetazolamide led to significant clinical improvement. **Conclusion:** This case highlights the rarity of bilateral papilledema and macular edema as initial presenting features of suspected SLE. It emphasizes the importance of thorough ophthalmological examination and relevant investigations in patients with sudden vision loss. Early recognition of such rare presentations is crucial to prevent potentially sight-threatening complications. This case underscores the need for a multidisciplinary approach for accurate diagnosis and management of complex presentations of systemic autoimmune diseases.

1. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by the production of autoantibodies that can target various tissues and organs throughout the body, leading to a diverse range of inflammatory and damaging processes. This complex condition is renowned for its heterogeneous

clinical manifestations, which can vary significantly from patient to patient. The clinical spectrum of SLE can range from relatively mild mucocutaneous involvement, affecting the skin and mucous membranes, to severe, life-threatening damage to major organ systems. This variability in presentation poses significant challenges in diagnosis and

management, as SLE can mimic or overlap with a wide array of other medical conditions. The etiology of SLE is multifaceted and remains an area of ongoing investigation. It is currently understood to involve a complex interplay of several factors, including genetic predisposition, environmental triggers, hormonal influences, and dysregulation of the immune system. Genetic factors play a crucial role in determining an individual's susceptibility to developing SLE, with numerous genes having been implicated in the disease pathogenesis. However, genetic predisposition alone is not sufficient to cause SLE, and environmental factors are believed to play a critical role in triggering disease onset and flares. These environmental triggers can include infections, exposure to ultraviolet radiation, certain medications, and other stressors. Hormonal factors, particularly estrogen, are also thought to contribute to the higher prevalence of SLE in women, especially those of childbearing age. Immune dysregulation is a hallmark of SLE, with abnormalities in both the innate and adaptive immune systems contributing to the production of autoantibodies and the subsequent inflammatory response. SLE predominantly affects women, particularly those of childbearing age, highlighting the potential role of hormonal factors in disease susceptibility and progression. The prevalence of SLE varies across different populations and ethnic groups, suggesting the influence of both genetic and environmental factors in disease distribution. Understanding the complex interplay of these etiological factors is crucial for the development of effective strategies for prevention, diagnosis, and treatment of SLE.¹⁻³

Ocular involvement is a common and clinically significant feature of SLE. Studies have consistently demonstrated a high prevalence of ocular manifestations in SLE patients, with reports indicating that 30% to 50% of individuals with SLE will experience some form of ocular involvement during the course of their disease. These manifestations can affect virtually any structure of the eye and its surrounding tissues, including the eyelids, conjunctiva, cornea, sclera, uveal tract, retina, optic

nerve, and the orbit itself. This broad spectrum of potential involvement underscores the importance of comprehensive ophthalmological evaluation in patients with SLE. The range of ocular manifestations in SLE is extensive, encompassing a wide variety of conditions that vary in severity and potential impact on vision. At the milder end of the spectrum, SLE can cause relatively common conditions such as keratoconjunctivitis sicca, also known as dry eye syndrome. Dry eye syndrome is characterized by ocular discomfort, irritation, and visual disturbances due to inadequate tear production or excessive tear evaporation. While often considered a minor inconvenience, severe dry eye can significantly impact quality of life. In contrast, SLE can also lead to severe, vision-threatening complications. Retinal vasculitis, an inflammation of the blood vessels in the retina, is a serious ocular manifestation of SLE that can cause significant visual loss if not promptly diagnosed and treated. Optic neuropathy, damage to the optic nerve, is another potentially devastating complication that can result in permanent visual impairment. These severe manifestations highlight the importance of early detection and aggressive management of ocular involvement in SLE to preserve visual function.⁴⁻⁶

Posterior segment involvement, affecting the structures at the back of the eye, is a significant aspect of ocular SLE. The optic nerve, which transmits visual information from the eye to the brain, can be affected in various ways in SLE. Optic nerve manifestations in SLE include optic neuritis, an inflammatory condition causing sudden vision loss anterior ischemic optic neuropathy, a condition resulting from insufficient blood supply to the optic nerve and, less commonly, papilledema. Papilledema, defined as swelling of the optic disc secondary to raised intracranial pressure, is an exceptionally rare initial presentation of SLE. The optic disc is the portion of the optic nerve that is visible within the eye. Papilledema is a sign of increased pressure within the skull, which can be caused by a variety of conditions. While papilledema is more frequently associated with conditions that directly cause increased intracranial pressure, such as

intracranial tumors, hydrocephalus (an accumulation of fluid within the brain), central nervous system infections, and idiopathic intracranial hypertension (IIH), its occurrence in the context of autoimmune diseases like SLE has been documented, but only in a limited number of case reports. This rarity underscores the importance of recognizing and reporting such cases to expand the understanding of the diverse manifestations of SLE. Macular edema, characterized by the accumulation of fluid in the macula, is another potential ocular complication of SLE. The macula is the central part of the retina responsible for sharp, central vision, which is essential for tasks such as reading, driving, and recognizing faces. Macular edema can result in blurred vision, distorted vision, and decreased visual acuity. It can occur due to various mechanisms, including inflammation, retinal vasculitis, and increased permeability of the blood vessels in the retina, leading to leakage of fluid into the retinal layers. The presence of both papilledema and macular edema in the same patient with suspected SLE is an unusual and infrequent finding. This co-occurrence warrants detailed investigation and reporting to better understand the underlying pathophysiology and clinical implications.⁷⁻¹⁰ This case report aims to describe a unique and rare presentation of a young female patient who presented with the sudden onset of bilateral papilledema and macular edema as the primary and initial manifestation of suspected Systemic Lupus Erythematosus.

2. Case Presentation

The patient, a 24-year-old female, presented to the ophthalmology clinic with a primary complaint of progressive blurring of vision. This visual disturbance had developed gradually, impacting her ability to discern fine details, such as reading text or recognizing facial features. The insidious nature of the vision loss prompted her to seek medical evaluation. Alongside the primary visual complaint, the patient also reported experiencing mild discomfort localized around the eyes. This discomfort was characterized by a dull ache

and a sensation of pressure. These symptoms were intermittent, and the patient did not identify any specific triggers that exacerbated or alleviated the ocular discomfort. A comprehensive review of the patient's medical history revealed that she had no significant past medical conditions. She denied any history of chronic illnesses, surgeries, or hospitalizations. Similarly, her ocular history was unremarkable, with no prior history of eye diseases, injuries, or corrective lens use. The patient also reported no known allergies to medications, food, or environmental factors. Furthermore, she had not experienced any previous ocular issues or treatments. The patient's family history was notable for its absence of autoimmune or ophthalmological conditions. There was no familial history of systemic autoimmune diseases, such as rheumatoid arthritis or lupus, nor was there any family history of significant eye disorders, such as glaucoma, macular degeneration, or inherited retinal dystrophies. This negative family history for relevant conditions reduced the likelihood of a hereditary component contributing to her presenting symptoms. A thorough general examination was conducted to assess the patient's overall health status. This examination included an evaluation of her pupillary reactions, extraocular movements, and intraocular pressure. The pupillary reactions were assessed for both eyes, and the findings indicated prompt and equal responses to light stimulation. There was no evidence of a relative afferent pupillary defect in either eye, suggesting that the optic nerve function was grossly intact and that there was no significant asymmetry in the afferent visual pathways. Extraocular movements were evaluated to assess the function of the muscles that control eye movements. The patient demonstrated full and unrestricted movement in all gazes, indicating that there were no limitations or abnormalities in the function of the extraocular muscles. This finding suggested that there was no evidence of cranial nerve palsies or other neuromuscular disorders affecting eye movements. Intraocular pressure (IOP) was measured in both eyes using Goldmann applanation tonometry,

the gold standard method for IOP measurement. The IOP in the right eye was recorded as 14 mmHg, while the IOP in the left eye was 15 mmHg. These values are within the normal range for intraocular pressure, which is generally considered to be between 10 mmHg and 21 mmHg. The IOP measurements did not suggest the presence of elevated intraocular pressure, which is a major risk factor for glaucoma. The anterior segment examination involved a detailed assessment of the front structures of the eye using slit-lamp biomicroscopy. This examination revealed no abnormalities of the eyelids, conjunctiva, cornea, anterior chamber, or iris in either eye. The eyelids appeared normal in terms of their position, structure, and function. The conjunctiva, the clear membrane covering the white part of the eye, was clear and without any signs of inflammation, such as redness or swelling. The cornea, the transparent front part of the eye, was also clear, with no opacities, abrasions, or other irregularities. The anterior chamber, the space between the cornea and the iris, was of normal depth and without any evidence of cells or flare, which would indicate inflammation. The iris, the colored part of the eye, had a normal appearance and reacted appropriately to light. The lens, responsible for focusing light onto the retina, was clear in both eyes, without any evidence of cataracts. The posterior segment examination involved a dilated fundus examination, which allowed for a detailed view of the structures at the back of the eye, including the retina, optic nerve, and blood vessels. Following the instillation of mydriatic eye drops to dilate the pupils, a thorough examination of the fundus was performed. The optic discs in both eyes exhibited swelling, with blurred margins that obscured the retinal vessels. The optic disc swelling was a significant finding, as the optic disc is the location where the optic nerve enters the eye, and swelling in this area can indicate a variety of underlying conditions. The cup-to-disc ratio, which is a measurement of the optic cup (the central depression within the optic disc) relative to the optic disc size, was smaller than normal in both eyes, approximately 0.1. A smaller than normal cup-to-disc

ratio is consistent with optic disc swelling. Macular edema was also observed in both eyes, characterized by the presence of exudates in the macular region. The macula is the central part of the retina responsible for central vision, and the presence of edema and exudates in this area can lead to a decrease in visual acuity and distortion of vision. The retinal vessels appeared normal, with no evidence of arteriolar narrowing, venous dilation, or hemorrhages. There were no cotton-wool spots or other signs of retinopathy observed in either eye. The peripheral retina was unremarkable in both eyes, with no evidence of any abnormalities. Neuro-ophthalmological testing was performed to further assess the visual function and integrity of the visual pathways. The Pelli-Robson contrast sensitivity test, which measures the ability to distinguish between subtle differences in contrast, was performed, and the results were 1.35 log units in both eyes. This indicates a mild reduction in contrast sensitivity. Ishihara color vision testing, using 25 plates, was performed to assess color perception. The patient demonstrated normal color perception in both eyes on this test. However, the Farnsworth D-15 test, another color vision test, revealed errors along the blue-yellow axis, indicative of tritanopia. This finding suggests a specific type of color vision deficiency. Humphrey visual field testing, an automated perimetry test, was conducted to assess the extent of the patient's visual field. The results of this testing revealed a superior arcuate defect in the right eye and an enlarged blind spot in the left eye. Visual field defects can indicate damage to various parts of the visual pathway, including the optic nerve. Macular assessment was performed using Optical Coherence Tomography (OCT) imaging. OCT is a non-invasive imaging technique that provides high-resolution cross-sectional images of the retina, allowing for detailed evaluation of the retinal layers. The OCT images revealed the presence of subretinal and intraretinal fluid accumulation in both eyes, confirming the clinical diagnosis of macular edema. The central macular thickness (CMT) was measured using OCT, and it was significantly increased in both

eyes. The CMT was 668 μm in the right eye and 869 μm in the left eye. Normal central macular thickness typically ranges from 250 μm to 300 μm , indicating a substantial increase in retinal thickness due to the accumulation of fluid. Brain and orbit imaging was conducted using Magnetic Resonance Imaging (MRI). The MRI of the head and orbits revealed optic nerve sheath distention on T2-weighted imaging. Optic nerve sheath distention is a finding often associated with increased intracranial pressure or inflammation around the optic nerve. A hyperintense lesion in the macular area was observed on T1-weighted imaging. The significance of this finding required further evaluation and correlation with the clinical findings. The intracranial findings were unremarkable, with no evidence of mass lesions, hydrocephalus, cerebral venous thrombosis, dural arteriovenous fistula, infarction, or hemorrhage. There was also no abnormal enhancement of the optic nerve to suggest optic neuritis or perineuritis. Cerebrospinal fluid (CSF) analysis was performed via lumbar puncture. The opening pressure was 18 cmH₂O, which is within the normal range (10-20 cmH₂O). The CSF composition was normal, with normal protein and glucose levels, and there was no evidence of infection or inflammation. Laboratory workup included a complete blood count, renal function tests, liver function tests, and thyroid function tests. All of these tests yielded results within normal limits. Inflammatory markers, including ESR (erythrocyte sedimentation rate) and CRP (C-reactive protein), were also within normal limits. The Antinuclear Antibody (ANA) test, performed using indirect immunofluorescence, was positive with a titer of 1:100, and the pattern observed was a fine speckled pattern. Further autoantibody testing, including anti-dsDNA and anti-Sm, was initiated, with results pending. Serological tests for common infectious causes of optic disc edema were negative. Based on the comprehensive evaluation, the clinical diagnosis was bilateral papilledema and macular edema, with a strong suspicion of an underlying autoimmune etiology, specifically Systemic Lupus Erythematosus. The constellation of clinical findings,

including the bilateral optic disc swelling, macular edema, visual field defects, and the positive ANA test, raised concern for an autoimmune process affecting the optic nerves and retina. The absence of other identifiable causes of papilledema, such as increased intracranial pressure or mass lesions, further supported the suspicion of an autoimmune etiology. The subsequent investigations and follow-up would be crucial in confirming the diagnosis and guiding appropriate management (Table 1).

The patient's clinical course was characterized by a structured treatment protocol and careful follow-up monitoring to assess the efficacy of the interventions and to manage any potential complications. The treatment strategy was initiated with the primary goal of addressing the bilateral papilledema and macular edema, while also considering the strong suspicion of an underlying autoimmune etiology, specifically Systemic Lupus Erythematosus. The initial treatment phase commenced with the patient's hospital admission to facilitate the administration of high-dose intravenous corticosteroid therapy. This therapeutic approach was deemed necessary due to the severity of the ocular manifestations and the potential for significant visual compromise. High-dose corticosteroids are a cornerstone of treatment for inflammatory conditions affecting the optic nerve and retina, aiming to rapidly reduce inflammation and prevent irreversible damage. The specific corticosteroid administered was methylprednisolone, a potent glucocorticoid known for its anti-inflammatory and immunosuppressive effects. The methylprednisolone was administered intravenously at a dosage of 1000 mg per day. This high-dose regimen was maintained for a duration of three days. Intravenous administration ensures rapid attainment of therapeutic drug levels, maximizing the potential for a prompt and robust clinical response. The rationale for this aggressive initial approach was to quickly suppress the presumed inflammatory process contributing to the optic disc swelling and macular edema, thereby preserving visual function. Following the three-day course of intravenous

methylprednisolone, the patient's treatment regimen transitioned to an oral tapering regimen. This transition aimed to maintain the therapeutic effects achieved with intravenous therapy while gradually reducing the risk of corticosteroid-related side effects associated with prolonged high-dose use. The oral corticosteroid prescribed was prednisone, another commonly used glucocorticoid. The initial dosage of prednisone was 65 mg per day. The tapering schedule would have been determined based on the patient's clinical response and tolerance to the medication. Corticosteroid tapering is a crucial aspect of management, as abrupt cessation of high-dose corticosteroids can lead to adrenal insufficiency and other withdrawal symptoms. In addition to the corticosteroid therapy, the patient was also started on acetazolamide, a carbonic anhydrase inhibitor, at a dosage of 250 mg three times daily. Acetazolamide is frequently used in the management of papilledema, particularly in cases where elevated intracranial pressure is suspected or confirmed, such as in idiopathic intracranial hypertension. While the patient's initial lumbar puncture revealed a normal opening pressure, acetazolamide may have been included in the regimen to address any subtle fluctuations in intracranial pressure that might not have been captured by a single measurement or to provide additional support in reducing optic disc swelling by potentially decreasing fluid production within the eye. Acetazolamide also has a mild diuretic effect, which can contribute to fluid management. During this initial treatment phase, careful monitoring of the patient's clinical status was essential. This included regular assessments of visual acuity, fundus examinations to evaluate the degree of optic disc swelling and macular edema, and monitoring for any potential adverse effects of the medications. Following the initiation of the treatment regimen, the patient demonstrated a positive clinical response. She reported a significant improvement in her visual acuity in both eyes. Specifically, her visual acuity improved to 6/15 in both eyes. This represented a substantial improvement from her initial presentation, indicating

that the treatment was effectively addressing the underlying cause of her visual impairment. Follow-up fundus examinations, conducted to assess the anatomical changes in the optic nerve and retina, revealed a reduction in the optic disc swelling in both eyes. This indicated that the inflammation or edema affecting the optic nerve was beginning to resolve. Furthermore, there was also a noted reduction in the macular exudates, suggesting that the macular edema was also responding to the treatment. The improvement in both optic disc swelling and macular edema correlated with the patient's subjective report of improved visual acuity. These positive outcomes during the initial treatment phase highlighted the effectiveness of the therapeutic interventions in addressing the patient's presenting symptoms and the underlying pathological processes. The combination of high-dose corticosteroids and acetazolamide appeared to be successful in reducing inflammation and edema, leading to improved visual function. Despite the initial positive response to treatment, the patient subsequently developed signs and symptoms suggestive of an adverse reaction. She presented with red, itchy spots on her skin, localized to her face, chest, hands, and back. These dermatological manifestations raised concern for a possible drug eruption or allergic reaction. In addition to the skin findings, the patient also reported experiencing mild shortness of breath. This respiratory symptom further heightened the suspicion of a systemic reaction, potentially involving the respiratory system. The constellation of symptoms, including the skin rash and shortness of breath, was suspected to be an allergic reaction to prednisone, the oral corticosteroid component of her treatment regimen. Corticosteroids, while effective anti-inflammatory agents, are also associated with a range of potential side effects, including allergic reactions. The development of a widespread rash and respiratory symptoms warranted immediate attention and a modification of the treatment plan. In response to the suspected allergic reaction, the oral prednisone was discontinued. The patient's oral corticosteroid was switched to oral

methylprednisolone. The dosage of the oral methylprednisolone was 48 mg per day. This decision to switch to a different corticosteroid, while maintaining the therapeutic class of medication, was based on the possibility that the patient's allergic reaction was specific to prednisone and that she might tolerate methylprednisolone. Methylprednisolone, like prednisone, is a glucocorticoid with anti-inflammatory properties, but it has a slightly different chemical structure, which might reduce the likelihood of cross-reactivity in a patient experiencing an allergic reaction to prednisone. Following the change in medication, the patient's symptoms resolved. The red, itchy spots on her skin disappeared, and the shortness of breath subsided. This resolution of symptoms supported the initial suspicion that the patient had experienced an allergic reaction to prednisone. The successful management of the adverse reaction allowed for the continuation of necessary corticosteroid therapy, albeit with a different agent. This episode underscores the importance of close monitoring for adverse drug reactions in patients receiving corticosteroid therapy. Prompt recognition and appropriate management of such reactions are crucial to ensure patient safety and to allow for the continuation of effective treatment. The patient continued to receive oral methylprednisolone as part of her ongoing management. The dosage and duration of the methylprednisolone therapy would have been guided by the patient's clinical response and the need to prevent recurrence of her ocular symptoms. Long-term management strategies in cases with a suspected autoimmune etiology often involve a gradual tapering of corticosteroids to the lowest effective dose to minimize the risk of chronic side effects. The final follow-up examination revealed a remarkable degree of clinical improvement. The patient's best-corrected visual acuity had returned to 6/6 in both eyes. This represents a complete recovery of visual acuity and a successful resolution of the visual impairment that prompted her initial

presentation. The return of visual acuity to this level indicated that the treatment had effectively addressed the underlying pathology affecting her vision. Fundus photography, a technique used to document the appearance of the retina and optic nerve, showed complete resolution of the optic disc swelling in both eyes. The optic disc margins were sharp, and a normal cup-to-disc ratio was observed. This normalization of the optic disc appearance provided further evidence that the inflammation and edema affecting the optic nerve had resolved. The cup-to-disc ratio, now within normal limits, confirmed the absence of optic disc swelling. Optical Coherence Tomography (OCT) of the macula was performed to assess the status of the macular edema. The OCT images demonstrated significant improvement in the macular edema in both eyes. The central macular thickness (CMT) had decreased substantially. In the right eye, the CMT decreased to 183 μm , and in the left eye, the CMT decreased to 196 μm . These measurements are within the normal range for central macular thickness, indicating the resolution of the subretinal and intraretinal fluid accumulation that characterized the macular edema. The resolution of the macular edema, as confirmed by OCT, correlated with the patient's improved visual acuity. The significant improvement in visual acuity, the complete resolution of optic disc swelling, and the resolution of macular edema all indicated a highly successful treatment outcome. The patient's clinical course, from initial presentation with significant visual impairment to complete recovery, highlighted the effectiveness of the treatment strategy and the importance of close follow-up monitoring. The successful management of this case underscores the need for a thorough diagnostic workup in patients presenting with bilateral papilledema and macular edema, as well as the potential for a favorable visual prognosis with prompt and appropriate treatment, even in cases with suspected underlying systemic autoimmune conditions (Table 2).

Table 1. Summary of patient's clinical findings.

Category	Finding	Description	Right eye (OD)	Left eye (OS)
Demographic data	Age	24 years		
	Gender	Female		
Chief complaint	Progressive blurring of vision	A gradual decrease in clarity, making it difficult to read or recognize faces	Yes	Yes
Associated symptoms	Mild discomfort around the eyes	Dull ache and sensation of pressure, intermittent, no specific triggers	Yes	Yes
Ocular history	Unremarkable	No known allergies or previous ocular issues	N/A	N/A
Medical history	Unremarkable	No significant past medical history	N/A	N/A
Family history	Negative for autoimmune or ophthalmological conditions		N/A	N/A
General examination	Pupillary reactions	Prompt and equal, no relative afferent pupillary defect	Prompt and equal	Prompt and equal
	Extraocular movements	Full and unrestricted in all gazes	Full and unrestricted	Full and unrestricted
	Intraocular pressure (IOP)	Measured using Goldmann applanation tonometry	14 mmHg	15 mmHg
Anterior segment examination	Slit-lamp biomicroscopy	No abnormalities of eyelids, conjunctiva, cornea, anterior chamber, or iris	Unremarkable	Unremarkable
	Lens		Clear	Clear
Posterior segment examination	Dilated fundus examination (Figure 1)			
		Optic disc swelling	Elevated with blurred margins, obscuring retinal vessels	Elevated with blurred margins, obscuring retinal vessels
		Cup-to-disc ratio	Smaller than normal (approximately 0.1)	Smaller than normal (approximately 0.1)
		Macular edema	Exudates present in the macular region	Exudates present in the macular region
		Retinal vessels	Normal, no arteriolar narrowing, venous dilation, or hemorrhages	Normal, no arteriolar narrowing, venous dilation, or hemorrhages
		Other retinal findings	No cotton-wool spots or other signs of retinopathy	No cotton-wool spots or other signs of retinopathy
		Peripheral retina	Unremarkable	Unremarkable
Neuro-ophthalmological testing	Pelli-Robson contrast sensitivity test		1.35 log units	1.35 log units
	Ishihara color vision testing	Using 25 plates	Normal color perception	Normal color perception
	Farnsworth D-15 test		Errors along the blue-yellow axis (tritanopia)	Errors along the blue-yellow axis (tritanopia)
	Humphrey visual field testing (Figure 3)	Automated perimetry	Superior arcuate defect	Enlarged blind spot
Macular assessment	Optical Coherence Tomography (OCT) (Figure 2)	Subretinal and intraretinal fluid accumulation	Yes	Yes
		Central macular thickness (CMT)	668 μm	869 μm
Brain and orbit imaging	Magnetic Resonance Imaging (MRI) (Figure 4)	Optic nerve sheath distention on T2-weighted imaging	Yes	Yes
		Hyperintense lesion in the macular area on T1-weighted imaging	Yes	Yes
		Intracranial findings	No mass lesions, hydrocephalus, cerebral venous thrombosis, dural arteriovenous fistula, infarction, or hemorrhage	No mass lesions, hydrocephalus, cerebral venous thrombosis, dural arteriovenous fistula, infarction, or hemorrhage
		Optic nerve enhancement	No abnormal enhancement to suggest optic neuritis or perineuritis	No abnormal enhancement to suggest optic neuritis or perineuritis
Cerebrospinal fluid (CSF) analysis	Lumbar puncture	Opening pressure	18 cmH ₂ O (normal range: 10-20 cmH ₂ O)	
		CSF composition	Normal protein and glucose levels, no evidence of infection or inflammation	
Laboratory workup	Complete blood count, renal function tests, liver function tests, thyroid function tests		Within normal limits	Within normal limits
	Inflammatory markers (ESR, CRP)		Within normal limits	Within normal limits
	Antinuclear Antibody (ANA) test	Indirect immunofluorescence	Positive with a titer of 1:100, fine speckled pattern	
	Other autoantibodies (anti-dsDNA, anti-Sm)		Results pending	
	Serological tests	For common infectious causes of optic disc edema	Negative	Negative
Clinical diagnosis		Bilateral papilledema and macular edema, with a strong suspicion of underlying autoimmune etiology, specifically Systemic Lupus Erythematosus		

Table 2. Treatment and follow-up.

Treatment phase	Treatment details	Follow-up and outcomes
Initial treatment	- Hospital admission for high-dose intravenous corticosteroid therapy with methylprednisolone 1000 mg per day for three days. - Followed by an oral tapering regimen of prednisone 65 mg per day in combination with acetazolamide 250 mg three times daily.	- Patient reported significant improvement in visual acuity, improving to 6/15 in both eyes. - Follow-up fundus examination revealed a reduction in optic disc swelling and macular exudates.
Adverse reaction management	- Patient developed red, itchy spots on her face, chest, hands, and back, along with mild shortness of breath, suspected to be an allergic reaction to prednisone. - Oral prednisone was switched to oral methylprednisolone 48 mg per day.	- Symptoms resolved after medication change.
Final follow-up	- Continued oral methylprednisolone.	- Best-corrected visual acuity returned to 6/6 in both eyes. - Fundus photography showed complete resolution of the optic disc swelling, with sharp margins and a normal cup-to-disc ratio. - OCT of the macula demonstrated significant improvement in the macular edema, with the central macular thickness decreasing to 183 μ m in the right eye and 196 μ m in the left eye, indicating the resolution of the subretinal and intraretinal fluid.



Figure 1. The fundus photograph showed bilateral optic disc swelling with blurred margins, an asymmetric cup-to-disc ratio, and exudates around the macula.

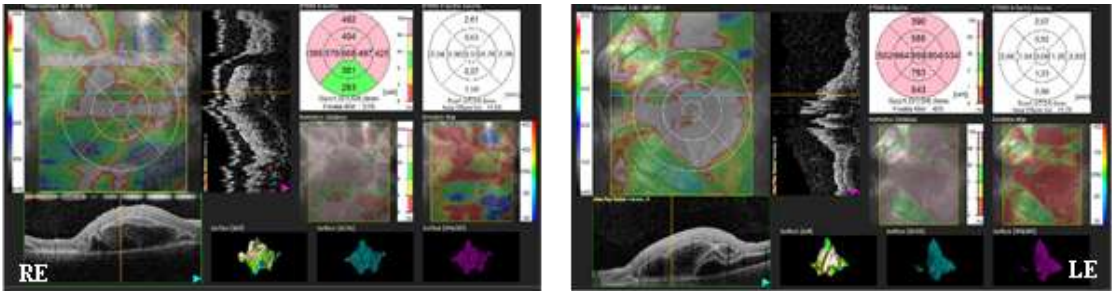


Figure 2. OCT macula showed subretinal and intraretinal fluid in both eyes.

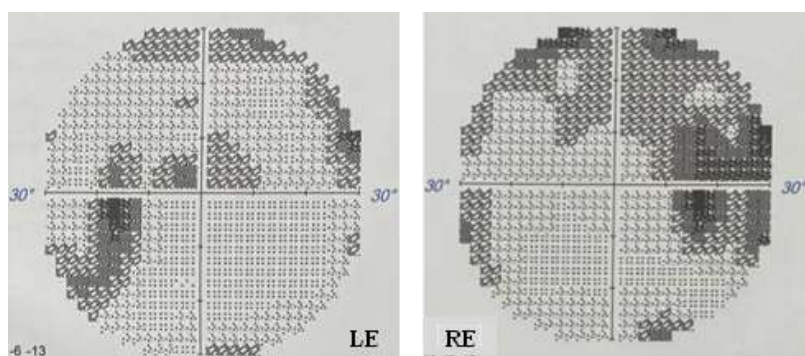


Figure 3. Humphrey Visual Field test showed a superior arcuate defect in the right eye and enlargement of a blind spot in the left eye.

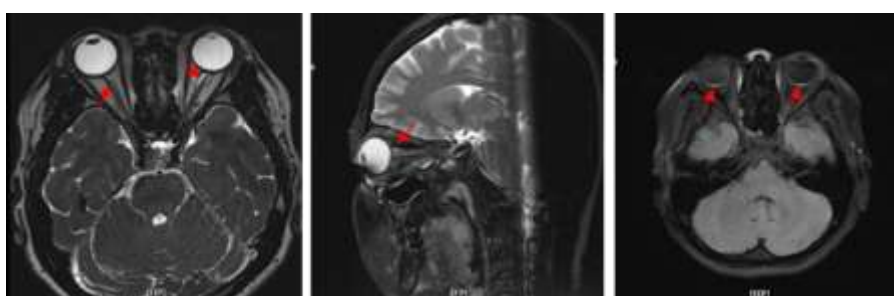


Figure 4. Head MRI T2 Weighted with orbital focus showed optic nerve sheath distention. A head MRI T1 Weighted with orbital focus showed a hyperintense lesion at the macula.

3. Discussion

This case report presents a distinctive instance of a 24-year-old female who initially manifested with bilateral papilledema and macular edema, which ultimately led to the suspicion of underlying Systemic Lupus Erythematosus. While ocular involvement is a recognized complication of SLE, the presentation of papilledema as the primary and initial sign is notably infrequent. This case underscores the critical importance of considering systemic autoimmune diseases in the differential diagnosis of patients presenting with bilateral optic disc swelling, even in the absence of other systemic symptoms at the time of the initial evaluation. The pathogenesis of ocular manifestations in SLE is complex and multifactorial. It involves a combination of factors, including the deposition of immune complexes, autoantibody-mediated inflammation, vasculitis, and microvascular ischemia. These pathological processes can affect various ocular structures, leading to a wide spectrum

of clinical presentations. In the context of papilledema, the underlying mechanism is often related to increased intracranial pressure. Increased intracranial pressure in SLE can arise from several potential causes, including cerebral venous thrombosis, thrombotic obliteration at the base of the arachnoid villi leading to impaired cerebrospinal fluid (CSF) absorption, or direct antibody-mediated injury to the arachnoid villi. In this particular case, the lumbar puncture revealed a normal opening pressure, which initially appeared to mitigate the likelihood of increased intracranial pressure as the primary etiology of the papilledema. However, it is important to acknowledge that intracranial pressure can fluctuate, and a single measurement may not fully capture the dynamic changes that can occur. Furthermore, it is plausible that the papilledema in this case was mediated by alternative mechanisms associated with SLE, such as localized inflammation within the optic nerve sheath itself. Although the MRI did not demonstrate

significant contrast enhancement to definitively support this, the possibility cannot be entirely excluded. The presence of macular edema in conjunction with papilledema in this patient further complicates the clinical picture. Macular edema in SLE can result from retinal vasculitis, which leads to increased vascular permeability and subsequent fluid leakage into the retinal layers. While overt signs of retinal vasculitis were not observed during the fundus examination in this patient, the possibility of subtle inflammatory changes contributing to the macular edema cannot be completely dismissed. It is also conceivable that the macular edema was secondary to the papilledema. Increased hydrostatic pressure at the optic nerve head can potentially lead to secondary fluid accumulation in the macula.¹¹⁻¹⁴

The positive Antinuclear Antibody (ANA) test, characterized by a fine-speckled pattern, was a significant finding that heightened the suspicion for an underlying autoimmune disease. The ANA test is a highly sensitive screening tool for SLE, although it is not entirely specific, as positive results can also be observed in other autoimmune conditions and, in some instances, even in healthy individuals. The titer of 1:100 in this case is considered relatively low but can still hold clinical significance when interpreted in the appropriate clinical context. The fine speckled pattern observed in the ANA test is one of several patterns that can be seen in ANA testing and is commonly associated with SLE and other connective tissue diseases. Further serological testing for more specific SLE-related autoantibodies, such as anti-dsDNA and anti-Sm antibodies, is essential for confirming the diagnosis of SLE, particularly when applying the EULAR/ACR classification criteria. These criteria stipulate the presence of a positive ANA titer ($\geq 1:80$) as an entry criterion, followed by the accumulation of points based on various clinical and immunological domains. In this case, the patient exhibited several clinical features suggestive of SLE. These included the ocular involvement, specifically papilledema and macular edema, visual field defects, the development of mucocutaneous abnormalities

during the treatment course, and the positive ANA result. The initial management of this patient involved a course of high-dose intravenous corticosteroid therapy. Methylprednisolone was administered intravenously at a dosage of 1000 mg per day for three days. This was followed by a tapering regimen of oral prednisone at a dosage of 65 mg per day, in combination with acetazolamide 250 mg three times daily. High-dose corticosteroids are the mainstay of treatment for severe ocular manifestations of SLE, including optic neuritis and retinal vasculitis. They are also utilized in the management of papilledema associated with inflammatory conditions. Acetazolamide, a carbonic anhydrase inhibitor, is commonly used in the treatment of papilledema, particularly in cases of idiopathic intracranial hypertension, as it functions to reduce CSF production. In this instance, even though the lumbar puncture opening pressure was within the normal range, acetazolamide was included in the treatment regimen. This may have been done to address any subtle or transient increases in intracranial pressure that were not detected by the single lumbar puncture measurement, or to address any component of optic nerve sheath fluid accumulation that may have been contributing to the papilledema. During the initial phase of treatment, the patient exhibited a positive response, with a reported improvement in visual acuity to 6/15 in both eyes. Follow-up fundus examination corroborated this clinical improvement, revealing a reduction in optic disc swelling and macular exudates. However, the patient subsequently developed a constellation of symptoms, including red, itchy spots on her face, chest, hands, and back, accompanied by mild shortness of breath. These symptoms were concerning for a potential allergic reaction to prednisone. In response to these adverse effects, the oral prednisone was switched to oral methylprednisolone at a dosage of 48 mg per day. The patient experienced resolution of the symptoms following this medication change. The final follow-up examination demonstrated further clinical improvement. The patient's best-corrected visual

acuity had returned to 6/6 in both eyes. Fundus photography confirmed the complete resolution of the optic disc swelling, with sharp margins and a normal cup-to-disc ratio. Optical Coherence Tomography (OCT) of the macula also demonstrated significant improvement in the macular edema. The central macular thickness decreased to 183 μm in the right eye and 196 μm in the left eye, indicative of the resolution of the subretinal and intraretinal fluid.¹⁵⁻²⁰

4. Conclusion

This case report elucidates a rare presentation of suspected systemic lupus erythematosus in a 24-year-old female who initially presented with bilateral papilledema and macular edema. The uniqueness of this case lies in the uncommon occurrence of papilledema as the primary and initial manifestation of SLE, as posterior segment manifestations like papilledema are rare, occurring in only about 1% of SLE patients. This case underscores the importance of considering systemic autoimmune diseases in the differential diagnosis of patients presenting with bilateral optic disc swelling, even in the absence of other systemic symptoms at the time of the initial evaluation. A positive ANA test further heightened the suspicion of an underlying autoimmune etiology, necessitating further investigations to confirm the diagnosis of SLE. The patient's clinical course was notable for a positive response to high-dose corticosteroid therapy and acetazolamide, with significant improvement in visual acuity and resolution of optic disc swelling and macular edema. This case also highlights the challenges in diagnosing SLE due to its heterogeneous clinical manifestations and the importance of a multidisciplinary approach for accurate diagnosis and management of complex presentations of systemic autoimmune diseases. Early recognition of such rare presentations is crucial to prevent potentially sight-threatening complications and underscores the need for a thorough diagnostic workup in patients presenting with bilateral papilledema and macular edema.

5. References

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