Systemic Lupus Erythematosus with Antiphospholipid Syndrome Presenting as Seizure Disorder and Retinopathy

[1]Sharma AK, [1] Sitaula S, [1]Thapa M, [1]Gautam P, [1]Shrestha GS, [2] Gajurel BP, [3] Sigdel MR

[1] Department of Ophthalmology, B.P. Koirala Lions Centre for Ophthalmic Studies,

[2] Department of Neurology, and [3] Department of Nephrology,

Tribhuvan University Teaching Hospital, Institute of Medicine Maharajgunj, Kathmandu, Nepal

Correspondence: Dr. Ananda Kumar Sharma, MD

Email: dr.anandasharma@gmail.com

Introduction

Clinical manifestation of systemic lupus erythematosus which is an autoimmune disease has been well documented in medical and ophthalmic literatures.[1,2,3,4] About one-third of patients suffering from systemic lupus erythematosus have ocular manifestations. Keratoconjunctivitis sicca being the most common ophthalmic manifestation.[2] Retinal vasculitis and optic neuritis neuropathy are vision threatening conditions that promptly require relevant investigations, diagnosis and appropriate treatment by the team of specialists. In the past, we have reported the ocular involvement in 47.3% of the patients, the commonest manifestation being dry eye (39.5%) followed by lupus retinopathy (21%) and drug induced ocular complications (21%).[3]

A case of suspected systemic lupus erythematosus in a 14 years young girl who complained red eye, periocular swelling and blurring of vision in both eyes was studied. She had history of seizure disorder, anemia, and multi systemic complaints .The past medical records had revealed chronic subdural hemorrhage and anemia.

Initially, the diagnosis of systemic lupus erythematosus was not made because the laboratory tests were negative. Subsequently, on ophthalmic examination, she had bilateral optic disc edema, retinal and subhyaloid hemorrhage with macular star, besides subconjuctival haemorrhage and periorbital edema. The diagnosis of systemic lupus erythematosus with antiphospholipid syndrome was made which was confirmed on repeated laboratory investigations for systemic lupus erythematosus. She had the medical treatment including systemic corticosteroid. Her conditions was also improved along with the significant visual recovery.

This case report demonstrates that antiphospholipid syndrome (APS) with seizure disorder and retinopathy warrants careful investigation in the line of SLE. So a detailed and timely ophthalmic evaluation along a relevant laboratory investigations are necessary to prevent vision loss.

Case Report

A 14 years young girl presented to the department of medicine of Tribhuvan University, Teaching Hospital (TUTH) with chief complaints of two episodes of generalized tonic clonic seizure followed by abnormal sensation of the body associated with staring look, twitching of the mouth, irrelevant talk, abnormal posturses and weakness of the body 15 days ago. She had history of seizure disorder, anemia and pain abdomen. The past medical records had revealed chronic subdural hemorrhage and anemia. She received blood transfusion and neurosurgical intervention done elsewhere years ago. She had regular menstruation with last menstrual period (LMP) 25 days back. She had no history of hypertension, diabetes, tuberculosis and other medical illness. Currently, had been receiving anti-seizure and hormonal drugs for last three years.

On physical examination, vitals were within normal limits. But, She has generalized weakness and unable to perform normal essential activities independently.

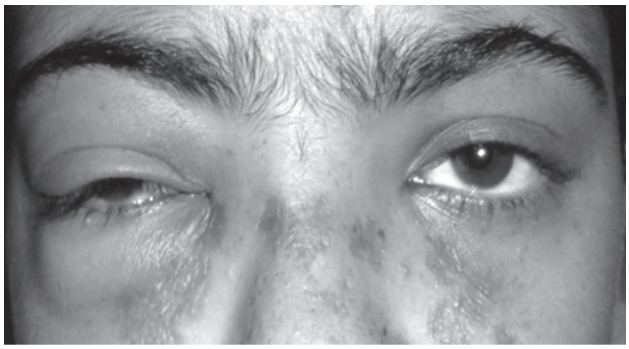

Figure 1 RE periorbital edema with subconjunctival haemorrhage and hyperpigmentation of skin

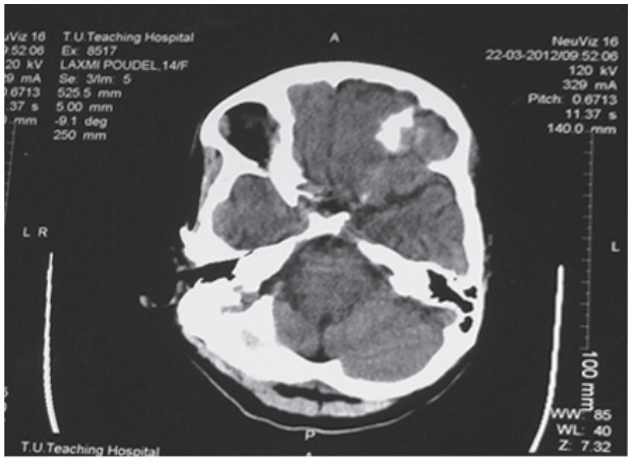

Figure 2 CT scan head with resolving subdural haemorrhage (SDH)

On systemic examination, she had normal vesicular breath; S1 & S2 was audible with systolic murmur, mild tenderness and mild distension of abdomen.

In a view of chronic illness, subdural hemorrhage (SDH), anaemia, clinical findings, detail laboratory investigations, ultrasonography and imaging (Figure 2, Table 1), she had received anti seizure treatment along with supportive treatment. When, she developed red eye, periocular swelling and blurring of vision in both eyes after one week of hospital admission, she was referred for ophthalmic examination in B. P. Koirala Lions Center for Ophthalmic Studies. On examination, her visual acuity in right eye was 3/60 and left eye was 4/60 respectively. Ocular motility and eye alignment were normal. Anterior segment examination revealed right subconjunctival hemorrhage, both pupils were mid-dilated and showed sluggish reactions. Fundus examination of both eyes showed the features of early disc edema with vascular changes, retinal and subhyaloid hemorrhage and macular star.

Table 1 Detail laboratory Investigation, Ultrasonography and Imaging

Investigations		Finding
Hematologic investigation	Hb%	9.6gm/dl
	WBC	11000/cumm
	DLC	
	Neutrophil	53%,
	Lymphocytes	36%,
	Eosinophils	3%
	Monocytes	8%
	PCV	38.4%
	RBC	5 million
	Platelets	1,10,000/cmm
Biochemical investigation	Urea	5.3 mmol/L
	Creatinine	166 Umol/dl
	Random Blood Sugar	7.7mmol/L
	Na+	134.7 mEq/L
	K+	3.99 mEq/L
	Albumin	37
	Calcium	1.8 mmol/L
	Phosphorous	4.62 mmol/L
	Amylase	50
	PT	28
	INR	2.23
	APTT	96
	Fibrinogen	600mg/dl
	Factor VIII activity	84%
	Factor IX activity	67%
	Anti ds DNA	7.0 IU/ml
	RPR (VDRL) test	Non Reactive
Urine analysis	crenated RBC	Plenty
CT of Brain	calvarian defect in left frontal region with adjacent soft tissue thickening of the scalp and pneumocephalous	
Ultrasonography of abdomen	small fluid collection around bilateral adnexa and pelvis.	
Echocardiography	no RWMA, LVEF 51%, dilated (LV, RA, & RV), moderate MR, AR, and moderate to severe TR, no I/C clot, vegetation or PE	

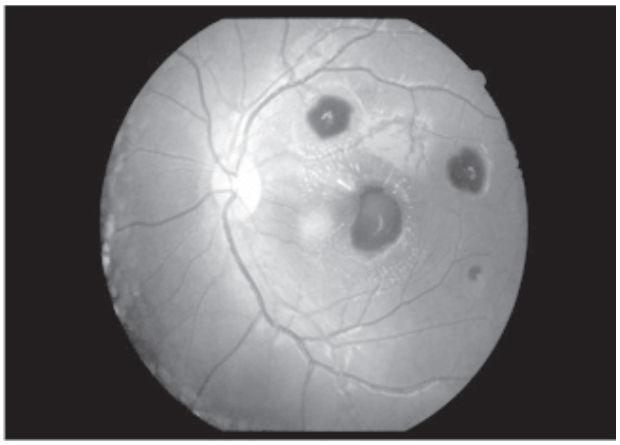

Figure 3 RE fundus picture with multiple retinal haemorrhage and macular edema

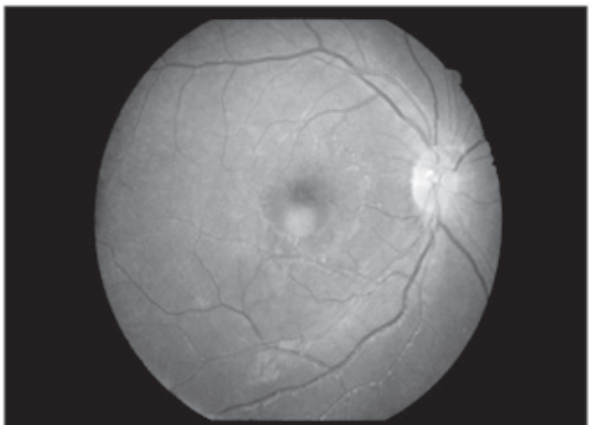

Figure 4 (a) LE fundus picture with resolved retinal haemorrhage and resolving macular edema

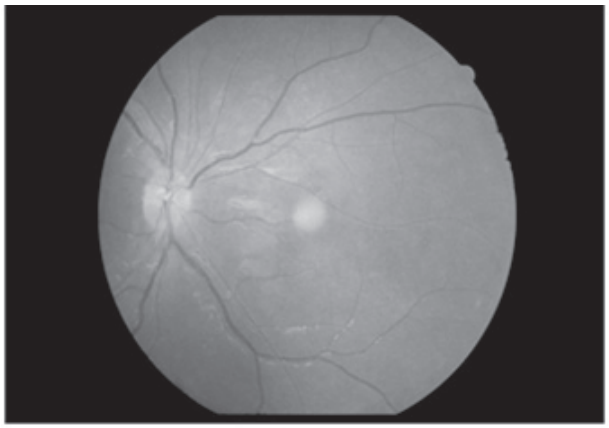

Figure 4 (b) RE fundus picture with resolved retinal haemorrhage after medical treatment

In a view of chronic illness, multi systemic involvement, rashes on the face, anemia and bleeding disorder including ophthalmic findings, systemic lupus erythematosus was suspected and relevant laboratory investigations were repeated. ANA was found positive with primary dilution of 1:40, Primary intensity on IF 2+, ANA pattern of speckled and end point titer of 1:160. Visually Evoked Potential (VEP) showed delayed P100 latencies in both eyes with normal amplitude. With the medical treatment along with systemic corticosteroids, her general condition improved and she was able to walk indecently. Besides these, visual acuity improved to 6/12 in right eye and 6/18 in left eye with the resolution of retinal haemorrhage and macular edema.

The final diagnosis was made as systemic lupus erythematosus with antiphospholipid syndrome and seizure secondary to intracranial haemorrhage. Ophthalmic findings such as subconjuctival haemorrhage in the right eye, bilateral retinal haemorrhage, and disc edema with macular star and optic neuropathy supported the diagnosis. Such kind of ophthalmic manifestations were not observed in previous published report of our centre, hence we would like to report this case.

She was discharged from the hospital with final treatment of oral drugs: Tab.Phenytoin 300mg once daily, Tab. Clobazam 10mg twice a day, Tab. Prednisolone 40 mg once after meal, Tab. Azathioprine 50 mg once daily, Tab. Calcium 500mg once after meal and Tab. Ranitidine 150mg twice a day before meal. We have noticed significant improvement in her general conditions and improved visual functions with medical treatment on her follow up visit.

Discussion

Multisystem involvement of Systemic lupus erythematosis (SLE) has been well documented in literatures. These manifestations may occur from general symptoms such as fever, malaise, arthralgias, myalgias, headache, loss of appetite and weight loss[5] to life threatening conditions.[6] This case report demonstrates that antiphospholipid syndrome (APS) with seizure disorder and retinopathy warrants careful investigation in the line of SLE. Diagnosis of SLE was confirmed by repeated laboratory investigations.

APS is manifested by venous or arterial thromboembolic events and various neurological manifestations.[7] these manifestations may arise secondary to ischemic processes involving brain tissue or direct action of antiphospholipid antibodies to neuronal tissue.[8] APS-associated antibodies can also directly affect neural tissues and the blood– brain barrier, causing various neurologic manifestations. The syndrome may be an isolated autoimmune phenomenon or manifestations of systemic disorders related to SLE.[9] In our study, APS manifested as a seizure disorder and

anemia that was the main reason for hospitalization and blood transfusion. She had been receiving anti-seizure and hormonal drugs for PV bleeding from last three years. Our finding closely agrees with the literatures that seizure disorder is generally manifested in APS in patients with SLE.[10,11]

Various ocular tissues have been found to be involved in SLE.[1] In our study, a case developed subconjunctival hemorrhage in right eye and bilateral disc edema with vascular changes, retinal and subhyaloid hemorrhage, and macular star. The ocular finding suggests vaso-occlusive nature of APS in SLE like in the other literatures. [12,13,14]

In this study, presenting visual acuity was significantly reduced to the level of less than 4/60 in both eyes. Clinical finding suggests bilateral optic nerve and retinal involvements. Our finding aligns with published literatures that optic neuritis and ischemic optic neuropathy was reported in patients with SLE with presenting visual acuity worse than 6/60 in most of the patients[15] and bilateral optic neuropathy is a well-known ocular manifestation occurring in patients with APS associated with SLE causing blindness[16]. Optic neuropathy is less frequently described in APS patients without SLE and tends to be unilateral in these cases.[17,18,19] In the literatures, the other causes of severe vision loss were central retinal artery/vein occlusions, vitreous hemorrhage, retinal ischemia, and neovascularization in SLE.[20,21,22] Such type of posterior segment manifestations including retinal involvement is remarkably high in patients with positive antiphospholipid titers in SLE.[22] In our study, antiphospholipid antibody titer was also positive.

Visual recovery was attained to 6/12 in right eye and 6/18 in left eye with resolution of retinal haemorrhage and macular edema [Figure 4 (a) & 4(b)]. The unpredictable and poor visual recovery has been documented in such patients with optic nerve involvement. Furthermore, complete visual recovery will not be always possible due to secondary changes in the optic nerve and the macula. [23]

Conclusion

Systemic Lupus Erythematosus with Antiphospholipid Syndrome can sometimes manifest as a retinopathy and optic neuropathy that might be neglected. Besides detailed systemic evaluation, relevant and repeated laboratory investigations, a detailed ophthalmic examination is mandatory to prevent visual loss.

Conflict of interest: None declared

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