Abstract: Anti phospholipid antibody syndrome, as an acquired pro thrombotic disorder is increasingly being recognised as an important cause of systemic venous and arterial thrombosis. The defining feature of this condition is the presence of raised levels of antibodies to negatively charged phospholipids in the serum. Antibodies may be found in patients who have SLE, syphilis and also rarely in other conditions. We here report a case of primary anti phospholipid syndrome and its ophthalmic manifestations.

Keyword: Anti phospholipid antibody syndrome, prothrombotic disorder, venous and arterial thrombosis

Anti phospholipid syndrome (Hughes syndrome) is characterised by arterial and venous thrombosis, thrombocytopenia and foetal loss occurring in the presence of anti phospholipid antibodies such as lupus anticoagulant, anti cardiolipin antibodies and others. It is now recognised as a primary distinct clinical entity, as most patients with the syndrome have no signs suggestive of SLE. Here we are reporting a case of primary anti phospholipid syndrome complicated by retinal neovascularisation.

CASE REPORT:
A 21 year old unmarried female came to our hospital with complaints of sudden onset defective vision RE for the past two months duration. She did not have any other ocular complaints like redness, pain or photophobia. She had been extensively investigated for the present condition elsewhere also. She had severe headache on presentation. She did not have any other history suggestive of thrombotic episodes. She had history of vague joint pains one year back for which she was on medications.

Her visual acuity OD was CFCF and OS was 6/6 by Snellens visual acuity. Her anterior segment examination was unremarkable BE and there were no cells in the anterior chamber or AVF. Pupils were brisk and acting BE. Fundus examination of her OD revealed a clear media with disc oedema and hyperaemia, multiple splinter and flame shaped haemorrhages at the posterior pole and mid periphery with sheathing of vessels. Macula was oedematous. OS showed disc hyperaemia and sheathing of vessels in mid periphery. We decided to do a fluorescein angiography in this patient suspecting vasculitis probably SLE.

Investigations that were done outside showed anti phospholipid antibody positive and ANA negative. Our initial differential diagnosis were SLE with occlusive vasculitis, IRVAN syndrome or renal hypertension. Her blood pressure on presentation was 200/110. Physician opinion was sought for immediately and the patient started on antihypertensives like tab atenolol 50 mg od and tab amlodipine 2.5 mg bd and she was concurrently investigated for the cause of elevated blood pressure and a rheumatologist opinion was also sought for suspecting vasculitis. A diagnosis of primary anti phospholipid antibody syndrome was made and patient was started on tab aspirin 75mg OD and tab warfarin 2mg OD. We started steroids in view of her vasculitis (tab prednisolone 60 mg OD)

FFA revealed OD multiple leaking dilated tortuous vessels over the disc with early leakage increasing in intensity and size in late phases suggestive of neovascularisation of the disc. Early leakage and capillary non perfusion zones in the peripheries was also noted. Extensive blocked fluorescence due to haemorrhages and multiple capillary non perfusion zones noted in the posterior pole.

OS showed staining of the disc and no leakage.

A OCT macula showed a CFT OF 370 microns in her RE and 158 microns in LE. Her blood pressure came under control with systemic medications. She had 3+ proteinuria and nephrologist suggested an elective renal biopsy and renal artery Doppler was planned. Other system evaluation were all within normal limits.

A RARE CASE REPORT OF PRIMARY ANTIPHOSPHOLIPID ANTIBODY SYNDROME

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A diagnosis of BE VASCULITIS was made and since neovascularisation and capillary non perfusion zones were present, pan retinal photocoagulation was performed in three sittings. Inspite of our expedient measures, the patient went on to develop vitreous hemorrhage. The inability to achieve adequate anticoagulation, in addition to persistent vasculitis, may have contributed to an unavoidable prolongation of her retinal ischemia and subsequent development of neovascularisation, despite attempts to counteract this ischemia with laser and steroids.

**DISCUSSION:**
Antiphospholipid antibodies are a group of autoantibodies which are detected by precipitation or complement fixation tests used in the detection of syphilis, lupus anticoagulant test, solid phase radioimmunoassay for anti cardiolipin antibodies. In vitro, antiphospholipid antibodies are anticoagulant but in vivo their effect is paradoxically to promote a thrombotic tendency. This is because of the actions of phospholipid components on platelet membranes and vascular endothelium as well as on thrombolytic factors such as antithrombin 3, prostacyclin and protein c. The lupus anticoagulant and the BFP-STS are antiphospholipid antibodies with similar specificity to anticardiolipin antibodies and collectively they are all referred to as antiphospholipid antibodies. Vaso occlusive retinopathy is a feature of thrombotic tendency in these patients, however the milder form is haemorrhages and cotton wool spots. In patients who have SLE, thrombosis in brain, retina is due to direct interference with the coagulation system mediated by antiphospholipid antibodies rather than by circulating immune complexes. Retinal artery and vein occlusions associated with antiphospholipid antibodies have also been reported in Sneddons syndrome (livedo reticularis, variable cerebrovascular disease and labile hypertension and lupus like disease). Three out of five patients reported by Levine et al presenting to an ophthalmologist with a variety of cerebral and retinal ischemic events did not have SLE and did have lupus anticoagulant. Three patients described by kleiner et al had lupus anticoagulant without SLE or any other disease. All three had severe visual loss secondary to vascular obstructions, rubeosis and vitreous hemorrhage.

**CONCLUSION:**
Antiphospholipid antibody syndrome is of great interest primarily because of reports of poor visual prognosis and secondly because of its association with thrombosis in the brain and elsewhere. Treatment with anticoagulation and immunosuppression and by control of associated hypertension may improve the prognosis in cases where stroke and recurrent abortions are prominent features. Ischemic retinopathy in APS can be due to either thrombosis alone or combination of thrombosis and vasculitis. Adequate anticoagulation and immunosuppression should be commenced immediately on diagnosis of this condition. It may be valuable to look for clotting screen abnormalities in younger patients particularly if there is a history of thrombosis at other sites or abortions. In these cases a search for specific antiphospholipid antibodies is justified.

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