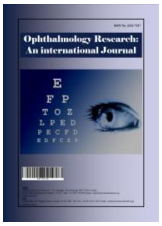

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A Rare Case of Anti Phospholipid Syndrome Associated Intermediate and Posterior Uveitis

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Authors' contributions

This work was carried out in collaboration among both authors. Both authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Autoimmune disorders are often associated with Uveitis. Anti-Phospholipid syndrome(APS) is characterized by increased hypercoagulability and divergent ocular features.
Case Presentation: A 45-year-old male patient presented with complaints of headache for 2 days. The patient gives a history of fever and loss of consciousness 20 days back and was treated symptomatically. On examination Visual acuity was 6/6 in the right eye and 6/12 in the left eye. On slit-lamp examination, the anterior segment was normal, vitreous cells were 2+ suggestive of vitritis in the left eye. Dilated fundus examination showed exudates in both eyes with macular parafoveal edema in the left eye. Laboratory investigations showed Antiphospholipid Antibody Immunoglobulin M (IgM) and Beta 2 glycoprotein IgM positive and raised ESR, PTT, and triglycerides. MRI brain showed acute embolic infarcts in multiple areas.
Management: Topical Steroids and cycloplegic for uveitis and systemic steroids and anticoagulants for systemic manifestations were given and the patient's vision improved to 6/6 and macular edema reduced in the left eye during the follow-up.
Conclusion: APS is a life and vision-threatening multisystem disorder, needs monitoring for INR and Anti phospholipid antibodies. A rare manifestation of uveitis is noted and prompt treatment with topical, oral steroids resolves the uveitis. But to prevent recurrences and further thrombotic events long-term immunosuppression, anticoagulation treatment is required.

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Keywords: APS-antiphospholipid syndrome; APL antibodies- antiphospholipid antibodies; IgM-immunoglobulin M.

1. INTRODUCTION

Antiphospholipid syndrome is defined as the presence of antiphospholipid antibodies, arterial or venous thrombosis, recurrent spontaneous abortions, and thrombocytopenia. The antiphospholipid syndrome patients have a risk of a thrombotic event of 0.5 to 30%. The syndrome can occur within the context of several diseases, mainly autoimmune [1].

Auto-immune disorders are often associated with Uveitis, which may vary in severity. Most common are HLA B27 associated disorders.

Anti Phospholipid Syndrome(APS) is characterized by increased hypercoagulability, vasculitis, and divergent ocular features.

Anti-phospholipid antibodies(Abs) are Lupus Anticoagulant Abs, anti-apolipoprotein Abs, and anti-cardiolipin Abs.

APS is 5 times more common in females than males.

Production of Abs is triggered by environmental factors, such as infections occurring in genetically susceptible individuals.

The history of this disorder dates from the 1950s when an in vitro anticoagulant phenomenon without deficiency of any clotting factor was first described in patients with systemic lupus erythematosus (SLE).

2. CASE PRESENTATION

A 45-year-old male patient presented with chief complaints of blurring of vision in both eyes from 2 days, which was insidious in onset and was associated with headache.

The patient had a past history of fever and generalized weakness for 10 days duration, later developed altered sensorium and skin lesions, suggestive of vasculitic lesions 15 days back for which he was admitted to Intensive Care Unit.He Underwent detailed evaluation, showed widal positive suggestive of typhoid fever and was treated. And connective tissue work up was done, showed strongly positive to Beta 2 glycoprotein Ab IgM and positive to Anticardiolipin Ab IgM.CSF analysis was done and was normal.MRI brain showed multifocal acute embolic infarcts.

Rheumatologist's opinion was taken and was diagnosed as Connective tissue disorder with Vasculitis (Primary antiphospholipid syndrome).

Then the patient was treated with systemic steroids, antibiotics, and anticoagulation.

No other known comorbidities and no significant family history was noted.

On ocular examination, we found the following features (Table 1).

Right eye optical coherence tomography (OCT) showing normal macular thickness.

Left eye OCT showing increased thickness para centrally and superiorly to the macula (CMT-327microns).

3. MANAGEMENT

3.1 Laboratory Blood Investigations – Showed

Increased Total leucocyte count(10,100 cells/cumm), CRP(47.7mg/dl), ESR(62mm/hr), and Procalcitonin levels (1.19ng/ml)suggestive of infection or inflammation.

Prolonged Prothrombin Time indicates the hypercoagulable state.

Lipid profile showed increased Triglyceride (318 mg/dl) and VLDL levels.

CSF analysis was done and found to be normal.

Widal test was done due to prolonged fever and turned out to be positive.

In Metabolic panel – showed strongly positive to Beta 2 glycoprotein Ab IgM and positive to Anticardiolipin Ab IgM as shown in Table 2.

3.2 On Radiological Investigation

MRI Brain (plain and contrast) – Multifocal diffusion restrictions with T2 and FLAIR

hyperintensities suggestive of Acute Embolic infracts.

Doppler of Carotid and vertebral artery showed- focal plaque in the posterior wall of the right carotid bulb.

By blood and radiological investigations it was confirmed to be Connective tissue disorder with Vasculitis.

3.3 Treatment and Outcome

Systemic Treatment: The patient was started on oral steroids and anticoagulants.

Tablet Prednisolone 4mg twice daily for 5 days followed by once daily for 5 days.

Anticoagulants Tablet Atorvastatin 75mg once daily was given.

Ocular treatment: The patient was started on topical steroids, cycloplegics.

Prednisolone eye drops 6 times/day for 1 week tapered accordingly, Cycloplegic Homatropine 2% eye drops twice daily for 1 week was given.

4. RESULT

Inflammation reduced significantly within 1week, follow up visit patient's visual acuity improved to 6/6, no cells in the anterior vitreous face (resolved vitritis). OCT showed resolved macular edema.

5. DISCUSSION

APS is defined as the presence of antiphospholipid Abs, arterial or venous thrombosis and thrombocytopenia and recurrent abortions, but all patients do not develop all symptoms.

APS is of two types - primary and secondary [2].

- Primary APS occurs in the absence of any other disease.
- Secondary APS occurs with other autoimmune diseases.

The risk of a thrombotic event is 0.5% to 30%.

90% of primary APS patients develop ocular involvement, 30% of them are asymptomatic.

The systemic features of the syndrome are characterized by large variability depending on the affected organs. In this context, neurological and behavioral disturbances, dermatological features as livedo reticularis and renal, ocular, liver, or valvular heart manifestations have been reported in APS patients [3,4].

Ocular involvement in APS includes varied manifestations from the anterior and posterior segment or the presence of neuro-ophthalmologic features [5].

In our case, we have seen bilateral intermediate and posterior uveitis with left eye macular edema and decreased vision as his presenting feature. He never had a previous history of uveitis. He had neurological symptoms and a history of fever 20 days back.

Laboratory investigations revealed Increased TLC, CRP, ESR, and Pro-calcitonin levels suggestive of infection or inflammation. Prolonged Prothrombin Time indicates the hypercoagulable state. In Metabolic panel, showed strongly positive to Beta 2 glycoprotein Ab IgM and positive to Anti cardiolipin Ab IgM.

MRI Brain showed multifocal diffusion restrictions with T2 and FLAIR hyperintensities suggestive of Acute Embolic infracts. Doppler of Carotid and vertebral artery showed- focal plaque in the posterior wall of the right carotid bulb.

The patient responded well to ocular treatment with topical steroids and cycloplegics and systemic treatment with oral anticoagulants oral steroids. steroids were tapered slowly and continued for the long term but anticoagulants had to be continued for even more time to prevent further thrombotic events and recurrences.

A Study done by Allam RS et.al. reported a case of a 25-year-old woman who was a confirmed a case of primary APS, presented with right eye chronic anterior uveitis with complicated cataract and appositional angle-closure glaucoma managed surgically by phacoemulsification–trabeculectomy. Was started on systemic steroids and hydroxychloroquine. After 2 years presented with an attack of posterior uveitis and started on immunomodulators [6].

Another study done by Takkar B et.al., reported a case of a 50-year-old female, presented with acute redness and low vision in the right eye and

was diagnosed with necrotizing scleritis with keratoiritis associated with APLS. She had recurrent untreated redness in this eye for the past 5 years. Was treated with oral steroids and immunomodulators and anticoagulants [7].

Table 1. Ocular examination

	RIGHT EYE	LEFT EYE
VISION – UCVA – BCVA	6/12 6/6	6/12 6/6
EYELIDS AND ADNEXA	Normal	Normal
EYEBALL POSITION	Normal	Normal
EXTRAOCULAR MOVEMENTS	Normal	Normal
CORNEA	Clear	Clear
PUPIL	3mm, round, regular, reacting to light	3mm, round, regular, reacting to light
ANTERIOR CHAMBER	Normal depth and quiet	Normal depth and quiet
LENS	Clear	Clear
VITREOUS CELLS	2+	2+
IOP BY GOLDMANN APPLANATION TONOMETRY	12 mm Hg	12 mm Hg
DILATED FUNDUS EXAMINATION	Hard exudates present , away from macula	Hard exudates present para-centrally and superiorly to macula
OCT - Macula	Central macular thickness(CMT)-284 microns	Central macular thickness(CMT)- 327 microns

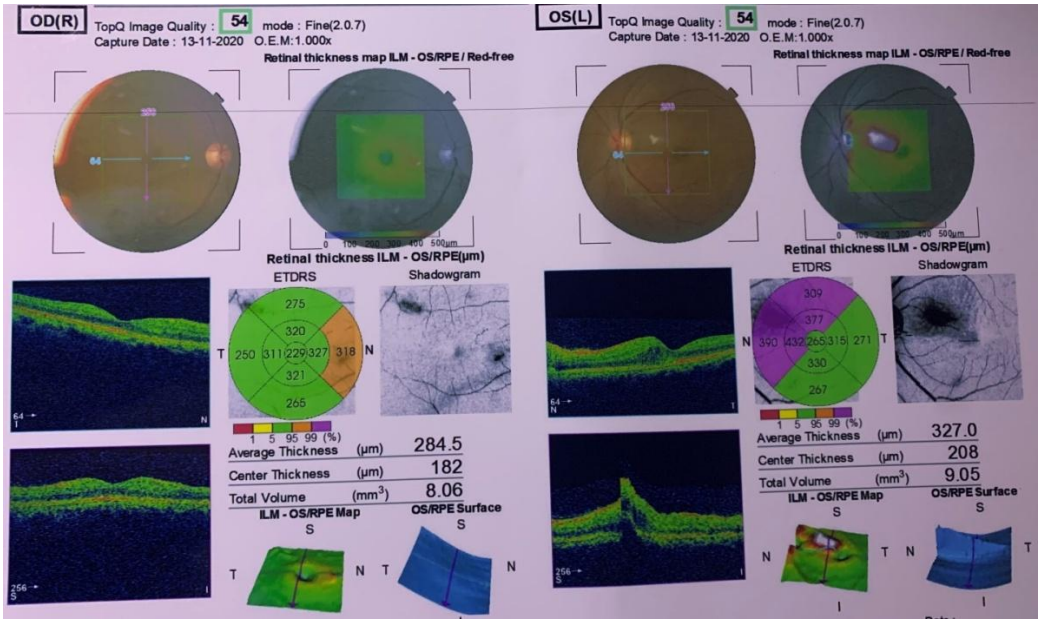

Fig. 1. OCT macula of both eyes

Table 2. Antiphospholipid laboratory profile of the patient

ANA	Negative
ENA	Negative
ACL IgG	2.50 U/ml
ACL IgM	12.00 U/ml
LAC	39.00s
Anti-β2GP IgG	0.8 U/ml
Anti-β2GP IgM	19.0 U/ml

ANA, antinuclear antibody; ENA, extractable nuclear antigens;
ACL IgG, anticardiolipin immunoglobulin G (normal: <10 U/ml);
ACL IgM, anticardiolipin immunoglobulin M (normal: <10 U/ml);
LAC, lupus anticoagulant (normal: 42 s).
Antiβ2GP IgG, anti-β2 glycoprotein immunoglobulin G (normal: <7 U/ml);
Antiβ2GP IgM, anti-β2 glycoprotein immunoglobulin M (normal: < 7U/ml);

6. CONCLUSION

APS is a life-threatening and vision-threatening multi-systematic disorder. A rare manifestation of uveitis is noted and prompt treatment with topical, oral steroids resolves the uveitis. But to prevent recurrences and further thrombotic events long-term immunosuppression, anticoagulation treatment is required along with regular monitoring of INR and Anti Phospholipid Antibodies. Essentially considered in patients with unexplained vascular occlusion as in this patient.

CONSENT

We report a rare case of antiphospholipid syndrome associated uveitis in a male patient.Infromed consent was taken from the patient and done according to declaration of Helsinki.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

What we knew:

1. Antiphospholipid syndrome is 5 times more common in females than males.

2. Anti Phospholipid Syndrome(APS) is characterized by increased hypercoagulability, vasculitis and divergent ocular features.

3. Anti-phospholipid antibodies(Abs) are Lupus Anticoagulant Abs, anti-apolipoprotein Abs, and anti-cardiolipin Abs.

4. APS can be associated with rare ocular manifestations including uveitis, scleritis.

What is new in our case report:

1. This is a very rare presentation of Anti Phospholipid Syndrome(APS) associated ocular features in a male patient.

2. From the review of the literature search, we could find very few cases of APS associated with uveitis in females.

3. In this patient, we noted Intermediate and posterior uveitis with macular edema associated with APS.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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