CASE REPORT

Evans Syndrome

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ABSTRACT
In the present case report we describe a rare case of a 21 years old girl patient presented to OPD with sudden acute painless loss of vision in both eyes (left>right) two weeks back. She has been diagnosed as Evans Syndrome 10 years ago. This case report highlights the step wise approach to diagnose a rare ocular manisfestations of Evan syndrome and its management leading to favourable results.
Keywords: Evans syndrome; Retinal Haemorrhages; thrombocytopenia purpura; Pre macular haemorrhages; Auto immune hemolytic anemia.

INTRODUCTION

Evans syndrome is an auto-immune disorder in which antibodies attack RBC (which deliver oxygen to blood tissues), Platelets (which help blood clot) and or Neutrophil (a type of WBC). The disorder resembles a combination of AIHA(autoimmune hemolytic anemia) + ITP(Idiopathic thrombocytopenia) and/or Neutropenia. They may occur simultaneously or may follow the other. The type of AIHA that presents in Evans syndrome is warm AIHA,in which IgG antibodies react with red blood cell(RBC) surface antigens at body temperature,as opposed to cold AIHA. In ITP,the immune system is directed against GPIIb/IIIa on the platelets.

Evans syndrome was first described in 1951 by Dr.Robert Evans and associates.

Recently ,a proportion has been laid out to classify the condition as primary(idiopathic) or secondary (associated with underlying disorder). Secondary Evans syndrome has been associated with diseases such as Systemic lupus erythematosus(SLE),Common variable immunodeficiency(CVID) and Autoimmune lymphoproliferative syndrome (ALPS) in Non Hodgkin lymphoma,Chronic lymphocytic leukemia and Viral infections such as HIV, Hepatitis C and CMV. .

There is no known genetic cause of Evans syndrome and occurrence in families are rare. It is considered a chronic disease with periods of remission and exacerbation. Thrombocytopenia receives in about 60% of patients, AIHA recurs in about 30%.

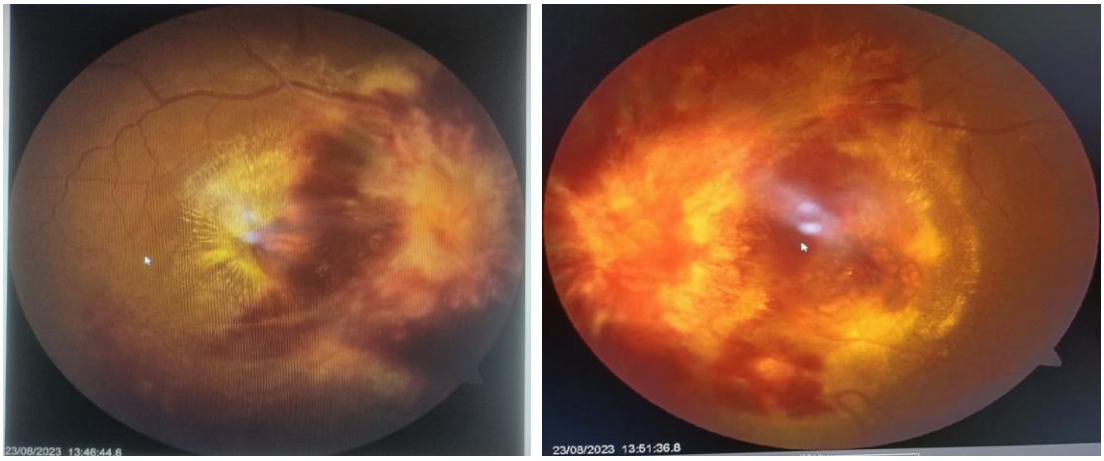
CASE PRESENTATION

A 21 years old girl with Evans syndrome presented with a sudden acute painless loss of vision in both eyes (left>right) during a period of remission from the disease for the past two weeks. She has been diagnosed as Evans syndrome 10 years ago, when she first presented with menorrhagia, gum bleeding, petechiae, weakness and fever. She reported no history of trauma or prior ocular condition.

METHODS

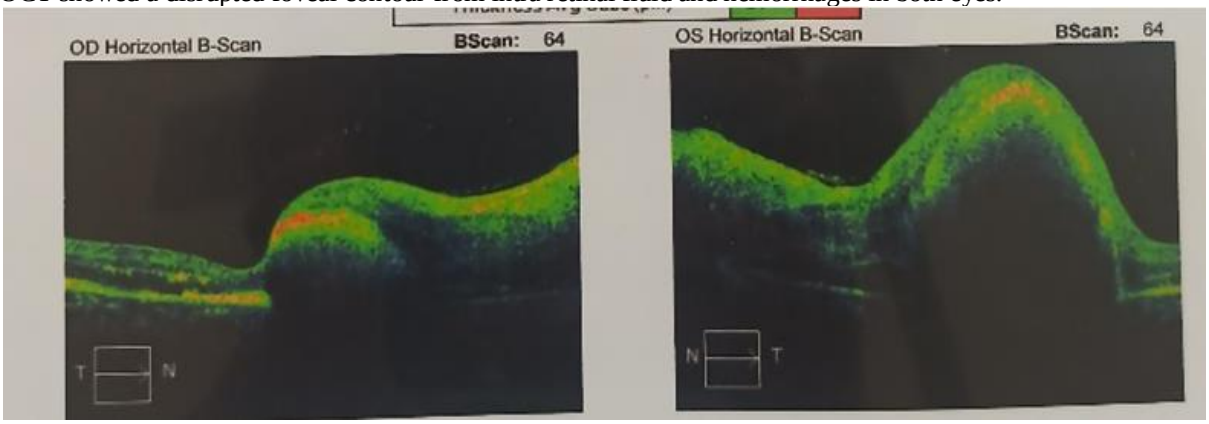
On examination the visual acuity was 6/60 and HM+ respectively. Anterior segment examination and SLE was normal.

A fundus examination revealed Disc edema,flame shaped haemorrhages around the disc,pre retinal haemorrhages in all quadrant,pre macular haemorrhages on L/E with multiple hard exudates around the macula resulting in Macular star. Tortousity of vessels was seen in both eyes.



OD **OS**

OCT showed a disrupted foveal contour from intra retinal fluid and hemorrhages in both eyes.



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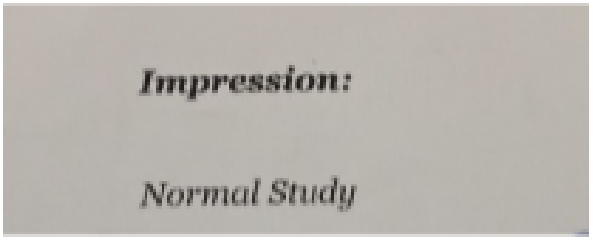

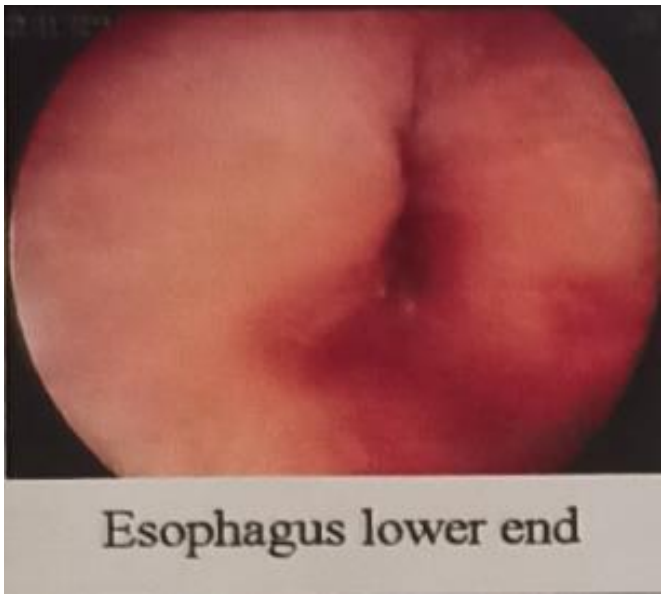

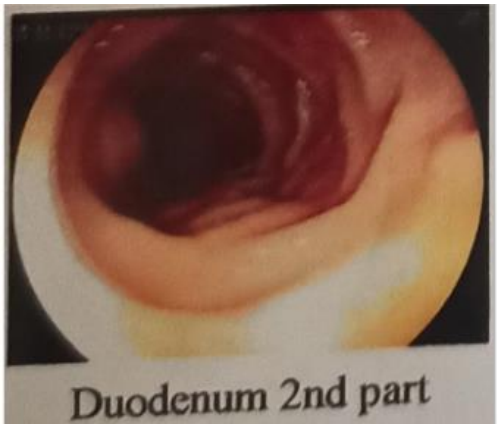

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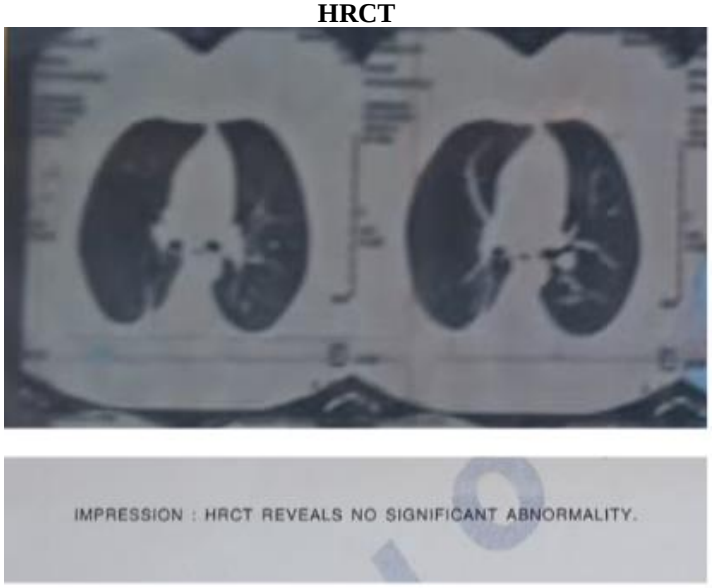

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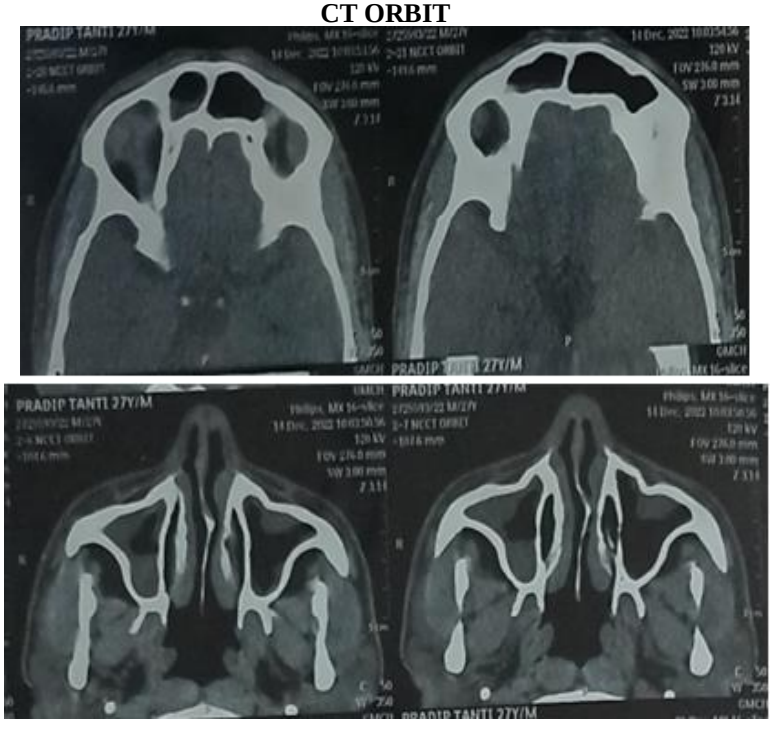


IMPRESSION: Suggestive of Subarchnoid Haemorrhage

DIFFERENTIAL DIAGNOSIS

It will includes PDR, CRVO, EALES DISEASE,DIFFUSE BILATERAL RETINAL HAEMORRHAGES, SICKLE CELL RETINOPATHY AND CMV RETINITIS.

RESULTS

Patient presented with Anemia (hb 6.9g/dl), Thrombocytopenia(10000/ul),jaundice and petechiae. Patient indirect bilirubin was raised, Coombs test was positive.LDH (310/ul) and reticulocytes count were raised. Viral markers were non reactive. ANA profile was negative.TSH,free T3 and free T4 were within normal limit. Bone marrow biopsy shows megakaryocytes. PBS shows normocytic normochromic anemia. Vit B12 level was within normal limit.

Based on Coombs test positive,Hemolytic Anemia and Thrombocytopenia. Patient was diagnosed as EVANS SYNDROME.

DISCUSSION

Evan syndrome is often managed by Platelet transfusion,Blood transfusion, Corticosteroid and other immunosuppressive agents.

TREATMENT

Ocular Treatment :

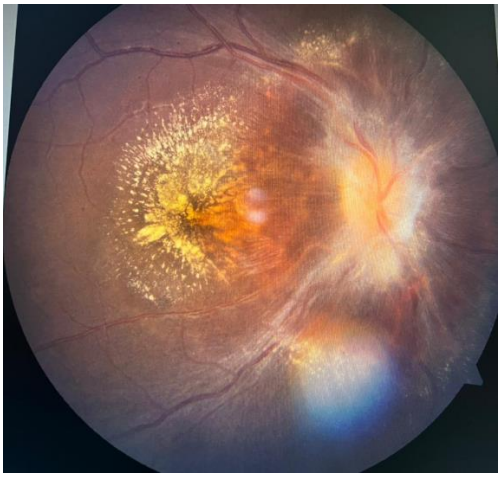
E/D Bromfenac,E/D CMC, Cap Anti oxidant.

Medicine Department :

The patient received platelet transfusion,blood transfusion,corticosteroid and chemotherapy(RITUXIMAB)of 3 cycles weekly .The patient got relieved and discharged.

FOLLOW UP

Fundus shows resolving retinal haemorrhages



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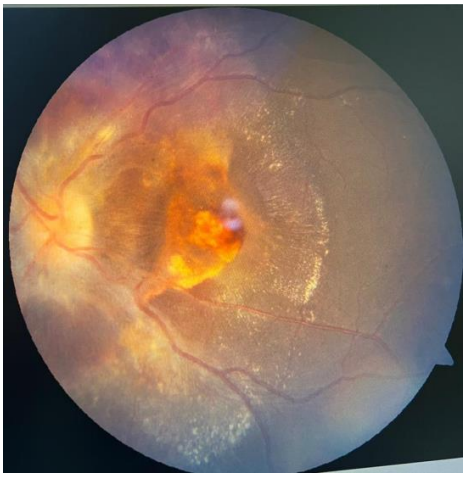

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CONCLUSION

This is a case describing a rare ocular complications of Evans syndrome leading to severe loss of vision due to vascular occlusion and preretinal and pre macular haemorrhages.

CONFLICT OF INTEREST

Nil

FINANCIAL INTEREST

Nil

REFERENCES

1. AA MAZHARUDDIN et al 2022, Ophthalmic Manisfestationas First Presenting Sign of Evans Syndrome, J Vitreoretin Dis.2022 Nov-Dec; 6(6):479-484.doi:10.1177/24741264211062931.PMCID : PMC9954779/PMID:37009543
2. Verma G, etal. BMJ Case Rep 2019;12 :e229636.doi :10.1136/bcr-2019-229636 ; premacular haemorrhage in a child with evans syndrome ;
3. Gl Guillaume et al .Retin cases Brief Rep.2023 Jul 1;17(4):359-361.doi:10.1079/ICB.0000000000001185, subhyaloid haemorrhages in evans syndrome.