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"Can Save Vision in Haemorrhagic Crisis": A Case of Evans Syndrome, Presenting as Vitreous Haemorrhage

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Abstract: ***Evans syndrome is a rare condition characterized by simultaneous or sequential development of autoimmune hemolytic anemia and immune thrombocytopenia (and/or immune neutropenia) A 30 year old lady present with diminision of vision of both eyes to light perseption and perseption of rays. It was associated with pervaginal bleeding, red colour urine, recurrent apathus ulcer and abdominal pain. On investigation presence of antierythrocyte antibodies detected by direct antiglobulin test in combination with haemolytic anemia with severe thrombocytopenia in presence of normal leucocyte count was present with detection of AIHA along with ITP confirming the diagnosis of Evan syndrome. On distant direct fundoscopy dull glow was present. On indirect ophthalmoscopy dense vitreous hemorrhagic seen covering the retina. After multiple transfusion and iv immunoglobulins visual acuity improves to CF 1/2 m. Further B/L vitrectomy done lead to improvement of visual acuity to 6/60. Detailed ophthalmological evaluation is of paramount importance in any bleeding disorder. Prompt treatment can lead to early visual rehabilitation***

Keywords: Evans syndrome, Autoimmune haemolytic anemia, vitreous haemorrhage, Pars plana vitrectomy

1. Introduction

Evans syndrome, which was first described in 1951, is an autoimmune disorder characterized by the simultaneous or sequential development of autoimmune haemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP) and /or immune neutropenia in the absence of an underlying cause. [(1) (2)] Although Evans syndrome has been since it's first description considered as an idiopathic condition and thus mainly diagnosis of exclusion, Evans syndrome may be associated with or show other diseases or conditions such as systemic lupus erythematosus, [(3)] lymphoproliferative disorders[(4)(5)] or primary immunodeficiencies. [(6)]

The diagnosis of Evans syndrome is made by the presence of anti erythrocyte antibodies detected by direct antiglobulin test. Based on temperature, at which autoantibodies will react with red blood cells, AIHA is classified as warm and cold antibody types. Warm antibody AIHA is more common. [(7)]

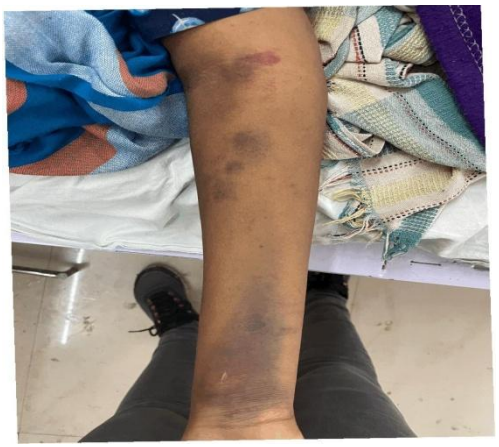
2. Case Presentation

A 30 year old female, resident of Sangli, Hindu by religion, homemaker by occupation, married since 10 years, presented with chief complaints of diminution of vision of both eyes since 2 days which was gradual in onset and rapidly progressive in nature and painless with associated complaints of pervaginal bleeding, red colour urine, recurrent aphthous ulcer and abdominal pain since 7 days. No h/o fever, joint pain No h/o trauma No h/o genital ulcer No h/o weight loss No h/o intake of any drug No h/o ocular surgery.

Physical Examination
Built: Cachexic
Afebrile
Pulse: 86 /min
BP: 100/70 mmHg
Pallor ++
No icterus, clubbing, cyanosis, edema or lymphadenopathy seen

Systemic Examination

- Generalized petechiae, bruise distributed over extremities, buttocks and entire trunk
- Recurrent oral ulcer
- Splenomegaly
- Frank pervaginal bleed with soft uterus
- Urine was red colour tinged



Generalized petechiae

Ophthalmic Examination

Right eye Left eye

Lid	Normal	Normal
Conjunctiva	Subconjunctival haemorrhage	Subconjunctival haemorrhage
Cornea	clear	clear
Anterior chamber	Normal depth	Normal depth
Iris	Normal colour pattern	Normal colour pattern
Pupil	Ill sustained reaction	Ill sustained reaction
Lens	clear	clear
EOM	Full in all directions	Full in all directions
IOP	Digitally normal	Digitally normal
Vision	Perception of light and rays present	Perception of light and rays present

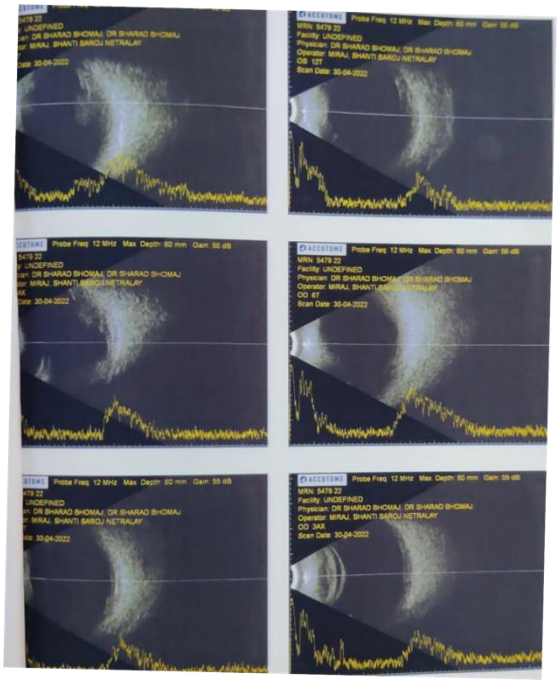
Subconjunctival haemorrhage

Posterior segment examination

Direct ophthalmoscopy - dull glow seen in both eyes

Indirect ophthalmoscopy - vitreous haemorrhage seen covering most of the retina in both eyes

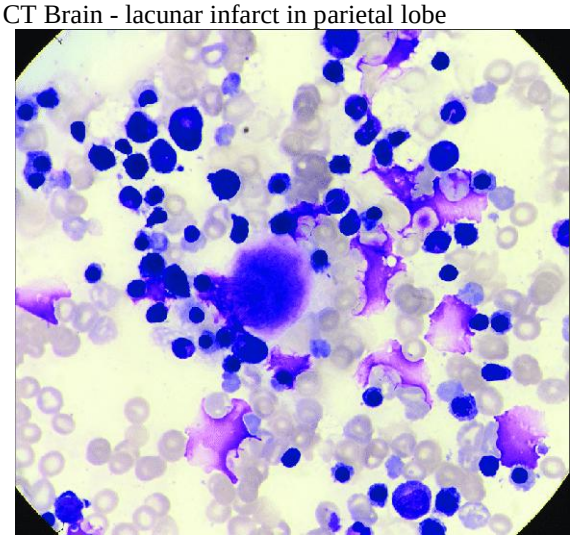
On B scan - e/o vitreous haemorrhage seen in BE with no e/o retinal or vitreous detachments.



B scan showing vitreous haemorrhage

Investigations

- Lab reports were showing severe anaemia (Hb 5 gm%), thrombocytopenia (Platelet 30, 000/cu. mm, with TLC 7000/cu. mm
- Positive for antierythroid antibodies
- Bone marrow examination showed hypercellularity with erythroid hyperplasia and crowding of megakaryocytes
- Coombs test positive AIHA
- CT abdomen - severe splenomegaly
- CT Brain - lacunar infarct in parietal lobe



Bone marrow smear

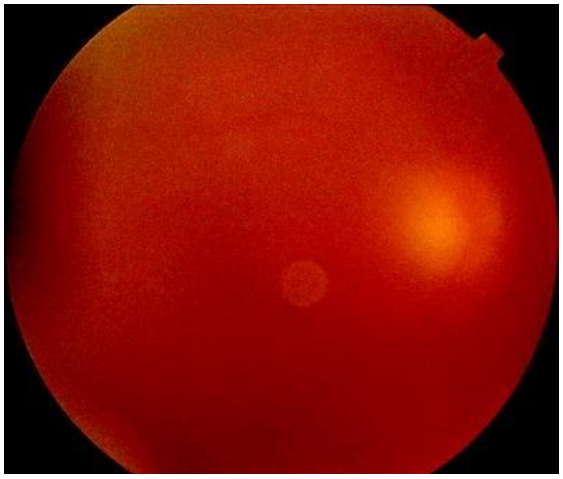
Final diagnosis

Based on clinical, coombs and fundoscopy findings, the diagnosis of Vitreous haemorrhage with Evans syndrome was confirmed.

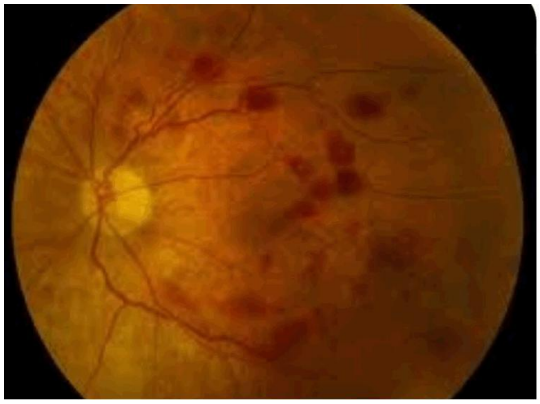
Management

- Since the patient is having severe anemia with thrombocytopenia multiple transfusions of PCV and FFP given with other symptomatic treatment.
- Further iv steroids and iv immunoglobulin were given

- The vision started to resolve to counting fingers 1 metre.
- Petechiae started resolving, bleeding ceased. Clinically, the patient improved.
- Bilateral Pars plana vitrectomy was planned and visual acuity improved to 3/60 further.
- After 2 months, visual acuity improved to 6/24as fundus changes regress for hemorrhagic retinopathy.



Before treatment



After treatment

3. Discussion

The typical course of Evans syndrome is characterized by an heterogenous chronic disease with clinical variability at onset, spontaneous remissions and exacerbations. First an exhaustive clinical history must be taken to determine the risk factors for developing Evans syndrome, such as infections, malignancies, autoimmune diseases, recent vaccinations, drugs, or family history of autoimmune diseases, [8]complemented with thorough physical examination focussed on the signs of anemia and thrombocytopenia. It is important to mention that primary Evans syndrome is a diagnosis of exclusion and secondary ES must be searched for to determine baseline disease because treatment and responses considerably differ between primary and secondary ES.

Anemia and bleeding tendencies can lead to intraretinal bleeding and sometimes massive vitreous haemorrhage in patients with haematologic diseases. In the present case, the intraretinal and vitreous bleeding was considered to be caused by a bleeding tendency resulting from Evans syndrome.

The management of Evans syndrome is mostly empirical and extrapolated from guidelines from both isolated AIHA and isolated ITP. Corticosteroids remain the first line therapy. Second line treatments include immunosuppressants and splenectomy. Ocular manifestations like vitreous haemorrhage in this case may need pars plana vitrectomy.

4. Conclusion

- A consensus has been reached to Evans syndrome leading to subretinal haemorrhage and haemorrhagic retinopathy.
- The condition responds excellently to transfusion treatment and vitrectomy
- Ophthalmological manifestation may be the initial clinch to diagnosis of haematological disorder.
- Prompt treatment can lead to early visual rehabilitation.
- Detailed ophthalmological evaluation is of paramount importance in any bleeding disorder.

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