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ANEMIC RETINOPATHY IN EVANS SYNDROME A CASE REPORT

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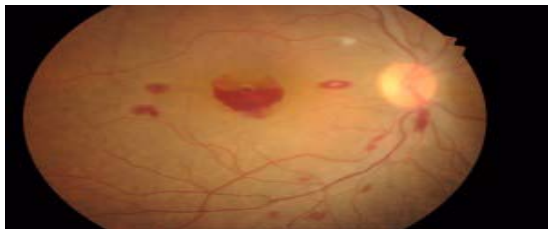
Abstract :
Evans syndrome is a rare disorder characterised by the presence of simultaneous or sequential direct Coombs-positive auto immune haemolytic anemia in conjuction with immune mediated thrombocytopenia with no underlying etiology

Keyword :
Evans syndrome, Anemic Retinopathy, Roth's spot

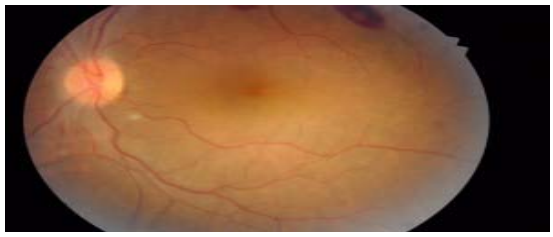
Evans syndrome is an autoimmune disease in which an individual's antibodies attack their own red blood cells and platelets. Both of these events may occur simultaneously or one may follow the other. Its overall pathology resembles a combination of autoimmune haemolytic anemia and immune thrombocytopenic purpura

CASE REPORT :

A 45 year old female was referred to RIOGOH from GH as a case of Evans syndrome for ophthalmologist opinion. Apparently normal patient, developed fever 1 month back associated with dyspnoea on exertion and easy fatiguability. She also developed progressive loss of vision in RE since past 20 days and yellowish discoloration of eyes since past 10 days. Patient had no similar complaints in the past and no past history of diabetes mellitus, hypertension, bronchial asthma & ischemic heart disease. She consumes mixed diet and her menstrual cycles were regular with normal flow. On general examination, patient was moderately built and poorly nourished. Pallor and icterus was present. Her pulse rate was 100/mt and BP was 100/60 mm Hg. Her cardiovascular system, respiratory system & central nervous system examination was within normal limits. Abdomen examination showed splenomegaly palpable 3mm below costal margin. Epigastric tenderness was also present. On ocular examination, vision was OD 6/60 not improving with pinhole & OS 6/18 with pinhole improving to 6/12.Anterior segment examination in both the eyes was normal. On fundus examination, RE media was clear, with normal disc and vessels. Multiple Roth's spots were present in the posterior pole. Sub hyaloid haemorrhage was seen at the macula. LE media was clear, with normal disc and vessels. Few Roth's spots were seen in supero temporal quadrant. IOP was normal in both the eyes.



RE FUNDUS



LE FUNDUS

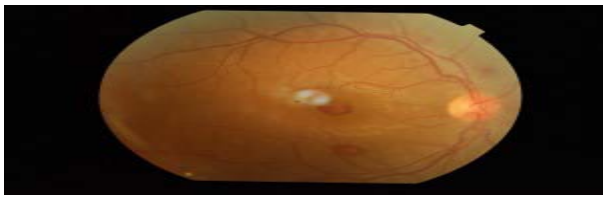
Investigations

Hb(g/dl)	7.1
Total Count	12100
Differential count	P63 L34 E3
RBC	1.1
Platelet Count	42000
Reticulocyte Count	1%

Liver Function Test

Total Bilirubin(mg/dl)	3.5
Direct Bilirubin	0.7
SGOT	36
SGPT	26
ALP	62
Serum Albumin	3.4
LDH	350 u/l

Peripheral Smear – Microcytic hypochromic anemia with thrombocytopenia
Direct Coomb's Test – Positive
USG Abdomen – Splenomegaly
Upper GI scopy - Normal
FINAL DIAGNOSIS-EVANS SYNDROME(AUTO IMMUNE HEMOLYTIC ANEMIA WITH THROMBOCYTOPENIA)
Patient was started on treatment with IV Methyl Prednisolone 750 mg OD ,IV Antibiotics, blood & platelet transfusion. Patient was followed up on outpatient basis and on review after 2 months, her repeat Hb was 9.0 mg/dl, platelet count was 1.8 lakhs/mm3 Vision improved to RE-6/18 NIP,LE- 6/9 NIP. Fundus examination revealed resorption of hemorrhages and Roths spots



RE FUNDUS

LE FUNDUS

DISCUSSION

Evans syndrome :

It was initially described by Evans et al in 1951.Spontaneous remissions and exacerbations are common and a few patients have neutropenia. Exact pathophysiology of Evans syndrome is unknown. Non cross reacting auto antibodies targeting different antigenic determinants on RBCs and platelets are assumed to cause isolated episodes of haemolytic anemia and thrombocytopenia respectively. The cytopenias may be related to T-cell abnormalities, decrease in helper T-cells and increase in suppressor T-cells. Wang et al demonstrated decreased serum levels of immunoglobulin G, immunoglobulin M and immunoglobulin A. Savasan et al observed that more than half of the patients with Evans syndrome have autoimmune lymphoproliferative syndrome. Pathophysiology is described as mutations of Fas gene that result in defective lymphocyte apoptosis. Etiology of Evans syndrome remains unknown. Manifestations of Evans syndrome are due to thrombocytopenia, anemia & occasionally neutropenia. Symptoms are purpura, petechiae and ecchymoses due to thrombocytopenia and pallor, fatigue, light headedness due to anemia. Jaundice may indicate hemolysis. Complications are haemorrhage with severe thrombocytopenia and severe infections with neutropenia Diagnosis can be done by laboratory studies which include Comlplete Blood Count, Reticulocyte count, Coombs test (direct antiglobulin test),tests for anti erythrocyte, antineutrophil and antiplatelet antibodies, Lupus antibody and ANA tests, measurement of serum immunoglobulins, flow cytometry of blood samples and gene mutation studies. Differential diagnosis include IgA deficiency, AIDS, Autoimmune lympho proliferative syndrome, Paroxysmal nocturnal hemoglobinuria, Thrombotic thrombocytopenic purpura, Hypersplenism

Management

Medical therapy is the main stay of management. Respiratory and cardiovascular functions should be stabilised, transfusion of blood products and pharmacological therapy IV Methyl Prednisolone is the most commonly used first-line agent IV immunoglobulin can be considered in resistant cases Other pharmacological therapies tried –Danazol ,Cyclosporine, Azathioprine, Cyclophosphamide, Vincristine, Rituximab, Alemtuzumab Additional therapies considered in refractory cases-Splenectomy and autologous stem cell transplantation Prognosis includes periods of remission and exacerbation. Recurrences of thrombocytopenia is seen in 60 % of patients and AIHA recurred in 31% of cases Prevalence of anemic Retinopathy increases with severity of anemia or thrombocytopenia. Retinal abnormalities in anemia without accompanying thrombocytopenia are less common unless the anemia is profound. Pathogenesis include dialatation of retinal vessels in response to retinal hypoxia, resulting in increase in transmural pressure leading on to vascular leakage causing hemorrhages and retinal edema. Infarction of NFL of retina –cotton wool spots. Roth's spot-white centered haemorrhage, represent focal ischemia, inflammatory infiltrate, fibrin & platelets. FFA reveals an increased retinal transit time

CONCLUSION

Anemic retinopathy has been associated with diseases of red blood cell elements or as a secondary manifestation of other systemic diseases. Treatment is correction of anemia, correction of underlying cause of anemia ,ophthalmic observation of anemic retinopathy In this case, the patient had resolution of sub hyaloid haemorrhage ,Roth's spots and vision also improved with IV steroids, blood & platelet transfusion This case highlights the need for fundus examination in all patients of anemia with defective vision and it proves the fact that ophthalmic manifestations do not require any specific treatment other than systemic management

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