Research Paper Medical Science

A Rare Case of Evans Syndrome With Hashimotos Thyroiditis

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ABSTRACT

The simultaneous or sequential development of autoimmune hemolytic anemia and immune thrombocytopenia and/or immune neutropenia in the absence of any underlying cause was described as Evans syndrome(ES).Since its first description , ES was considered idiopathic , and mainly as a diagnosis of exclusion.Few case reports showed a common association of ES with other diseases, such as , SLE, lymphoproliferative disorders,which warranted the classification of ES into primary and secondary.However , the association of Evans syndrome with Hashimoto's thyroiditis was found to be an extremely rare and only 5 such cases were published till date to the best of our knowledge.

KEYWORDS evans syndrome, hashimtos thyroiditis, autoimmune haemolytic anemia,coombs, immune thrombocytopenia.

SUMMARY

A 23yr old who was a known case of HASHIMOTOS THYROIDITIS was evaluated and found to have low HB , elevated bilirubin levels , splenomegaly. Her peripheral smear showed evidence of immature RBC and RBC destruction. She also tested positive for COOMBS and TPO antibodies.Taking patients clinical picture and laboratory findings into consideration we diagnosed the case as autoimmune haemolytic anemia with immune thrombocytopenia and associated Hashimoto's thyroiditis.

CASE REPORT

HISTORY

A 23 yr old female who is a known case of HASHIMOTO'S thyroiditis on treatment presented to us with complaints of low grade fever (not associated with chills) and easy fatiguability since 10 days. In the past 3 years she had frequent attacks of jaundice , fatigue , generalised edema and an episode of red cloured urine.She has h/o since 4 months.

She got diagnosed for Hashimoto's thyroiditis with hypothyroidism and is on treatment since one and half year.(has discontinued since 1 month).She is not using any other drugs except Eltroxin She has no significant family history of any thyroid or blood disorders.

PHYSICAL EXAMINATION

She weighed 37 kg and measured 151 cm height (body mass index 18.1 kg/m^2).She has pallor and icterus.**b/l proptosis present. VITALS** body temperature-98.6°f, pulse rate-100/minute. blood pressure -100/70 mmHg, respiration rate- 20 breaths/minute, and

CVS examination revealed normal jvp, a loud first heart sound and systolic haemic murmur present

Per abdomen – soft, non tender , **spleen palpable** –10 cms below the lt costal margin,firm , non tender. Mild hepatomegaly(2cms below rt costal margin) was present.soft , non tender.

Other systems normal

Investigation	Report	Normal value
HB	**2.5gm/dl**	Men.13.5-18.0gms/dl. Women.11.5-16.4gms/dl
WBC	6650/cumm (N-43%,l-54%, E01%,M-02%)	4000-11000/cumm
PCV	**8.1%**	Men.40-50% women-36-46%
Platelet	1.38lakh/cumm, **previously 1.42 lakh/cumm(2years back)**	1.5-4.0lakhs
ESR	90mm/1sthr	0-15mm/1sthr
Sr.sodium Sr.pottasium	135mmol/l 4.2mmol/l	135-155mmol/l
RBS	100mg/dl	<140mg/dl

Bilirubin direct **indirect** AST ALT ALP	6.9mg/dl 1.2 **5.7** 14 29 107	 Upto 1mg/dl Upto 0.25mg/dl < 37 IU < 40 IU
MPICT	weakly reactive for pf and pv	
LDH RETIC count	210 7%	140-280 U/L 0.5%-2.5%
Coombs Direct Indirect	 Positive **Positive**	
ANA and DSDNA	negative	
TPO anti bodies	positive (>1300 IU/ml)	
TSH	2.79 uIU/ml	- 4.2 uIU/ml

Peripheral smear showed anisopoikilocytosis , polychromatocytes and nucleated RBC .

Ultrasound showed findings suggestive of splenomegaly ,mild hepatomegaly, multiple cysts in both ovaries.

FNAC thyroid showed diffuse lymphocytic and plasma cell infiltration with formation of lymphoid follicles. Atrophy of thyroid parenchyma is evident.

In summary she has low HB , elevated bilirubin levels , splenomegaly. Her peripheral smear showed evidence of immature RBC and RBC destruction. She also tested positive for COOMBS and TPO antibodies.Taking patients clinical picture and laboratory findings into consideration we diagnosed the

case as autoimmune haemolytic anemia with immune thrombocytopenia and associated Hashimoto's thyroiditis.

Treatment given

In view of severe anemia we planned for blood transfusion but due to severe incompatibility to all blood types (due to presence of auto antibodies), we first started her on steroids (iv prednisone) Then we started her on PRBC transfusions on day 3 of steroids At the time of discharge she was improved clinically and her Hb was 9.4gm/dl. We sent the patient home with oral prednisolone 1mg/kg , thyroxine ,iron suppliments , vitamin suppliments and asked for regular followups for further dose adjustments.

On her recent visit one wk back,She has no complaints, clinically improved.Hb was 11.8gm/dlPlatelets – 2.8 lakh/cumm LFT – with in normal limitsPresently she is on maintanence dose of prednisolone 10mg/kg

DISCUSSION

AUTOIMMUNE HAEMOLYTIC ANEMIA is an aquired disorder in which an IgG antibody is formed that binds to red blood cell membrane due to unknown alterations in immune regulatory mechanisms. The Fc portion of this antibody is recognised by macrophages in spleen and other portions of reticulo endothelial system. Due to this interaction the RBC membrane is removed and spherocytes are formed. These spherocytic cells have decreased deformability and hence trapped due to the their inabiliy to squeeze through splenic sinusoids. Direct complement lysis is rare but presence of c3b on RBC surface allows kupffer cells to participate in haemolysis. The destruction of RBC in spleen and liver designates that this is an extravascular haemolysis. Approx one half of AIHA are idiopathic and rest are seen in association with SLE,CLL etc… Drug induced haemolysis must be ruled out. (penicillin,ceftriaxone, pipericillin,fluderabine –anti neoplastic.)

Clinical picture-Anemia is often severe and life threatining Fatigue, dyspnea, angina pectoris, heart failure are common presentations. On examination, jaundice and splenomegaly are usually present.

Investigations

A very low HB,Reticulocytes and spherocytes on peripheral smear .In case of severe haemolysis the stressed bone marrow may also release nucleated RBC as seen in this case.Indirect bilirubin is increased

Approx 10% of patients with AIHA have coincident immune thrombocytopenia.(evans syndrome) .The antiglobulin test(coombs) forms the basis for the diagnosis.Direct coombs indicate presence of antibody on the RBC surface.Indirect coombs indicate presence of free antibody in the patients serum.In 10% of cases coombs test may be negative .Since the patients serum usually contains the autoantibody it may be difficult to obtain a compatible cross match for transfusions.

Other investigations include Anti erythrocytic antibodies,Anti neutrophil antibodies, Antiplatelet anticodies ,Measurement of serum immunoglobulins , Flow cytometry , Gene mutation studies

TREATMENT-

1st line of treatment is prednisone 1-2mg/kg orally with divided doses.Decision regarding blood transfusion is controversial in initial stages because the complete compatibility cant be achieved. If prednisone is ineffective or relapse occurs after tappering , splenectomy should be considered. In patients with rapid haemolysis , therapeutic plasmapheresis should be done to remove autoantibodies. Patients refractory to prednisolone and splenectomy can be treated with rituximab,danazol,cyclophosphamide azathioprine, mycophenolate mofetil, cyclosporine, may also be used.High dose iv immunoglobulins may be effective in controlling haemolysis temporarily.Treating with stem cell transplant is being studied. Long term prognosis is good in patients if there is no other underlying autoimmune or lymphoprolioferative disorders.

IMMUNE THROMBOCYTOPENIA

There is a diversity of autoimmune mechanisms in idiopathic thrombocytopenic purpura (ITP), such as, antiplatelet antibodies and B-cell, and T-cell tolerance. Platelet antibodies are only detected in approximately 60% of the patients and failure to detect may be due to limited test specificity or undetected antigens.

CONCLUSION

There is an increased susceptibility for people with one autoimmune disease to develop another. The increased relative risk of acquiring a second autoimmune disease may be due to a genetic susceptibility that affects both diseases, the alteration of the body's homeostasis by one disease that creates a susceptibility to another .

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