Research Paper Medical Science

A Rare Case of Pure Red Cell Aplasia

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KEYWORDS

A 50 Years old female patient presented to VSGH with breathlessness.Patient was relatively asymptomatic before 4 months. Then she developed breathlessness.weakness and easy fatiguability on exertion which gradually increased over the period of 4 months and at the time of presentation, she was breathless even at rest.Patient also gave history that she had similar complaint 3 years back which was resolved after blood transfusion and she was asymptomatic during this period.Her history was negative for cough,fever,cold,PND,chest pain,palpitation,abdominal distention and pedal edema.

On general examination, patient was found to be severly anemic. Conjuctiva, tongue, lips, nail bed showed severe pallor. Nails were showing koilonychia.Patient was breathless with respiratory rate 20/min. And on auscultation, there was grade -2 systolic murmur near apex.

So clinical diagnosis diagnosis of anemia made and patient was inquired about history of blood loss in detail.But, patient denied blood loss in any form.

Patient's blood was sent for primary investigations

Which showed
Hb:-1.68 gm%
Creatinine:- 0.81mg%
Total WBC count:- 5350/cumm
Bilirubin:- 0.6mg%
Absolute Platelet Count:- 3.94lacs/cumm.

RBCs were predominantly normocytic normochromic with presence of eliptocyes, leptocyes, and tear drop cells.

Total WBC count and platelets were adequate on smear.

Differential WBCs showed very mild lymphocytosis.

Retic count was 0.5%

Stool R/M was negative for occult blood as well as worms for 3 consecutive days.

Urine R/M was normal.

Considering the diagnosis of symptomatic anemia under investigation for symptomatic improvement , it is decided to give 2 units of packed cell volume.Before starting PCV patients Blood sample are taken for further investigation

Which showed
Vitamin B12 :- .1200 pg/ml
Iron:- 179mcgdl
TIBC:-463mcg/dl
Ferritin:-382ng/ml

So, iron deficiency, B12 deficiency, worm infestation, etc. were ruled out.

Patient's blood is also sent for G6PD and LDH which were normal.

After 2 units of packed cell volume patient's investigations are repeated which were as follows

Hb-4.67 gm%

Total WBC count:-10000/cumm

Absolute platelet count:-1.03lacs/cumm

After this patient was sent for ultrasonography, which was normal except for mild splenomegaly with long axis 12.6cm.

Even after these much of investigations, cause of anemia remained elusive.So, help from expert hematologist sought.

On her advice bone marrow aspiration,bone marrow biopsy and blood for ANA sent for further Work up.

BONE MARROW ASPIRATION REPORT:-

Site:-PSIS

Aspiration:- Particulate marrow mildly diluted by peripheral blood

Cellularity:- Normocellular

Erythropoesis:-Cells of erythroid precursors are markedly reduced. Only few basophilic ertthroblasts are seen.

Leukopoesis:- Cells of myeloid series are normal in number and maturation.

Megakaryocytes:- Megakaryocytes are normal in number and morphology.

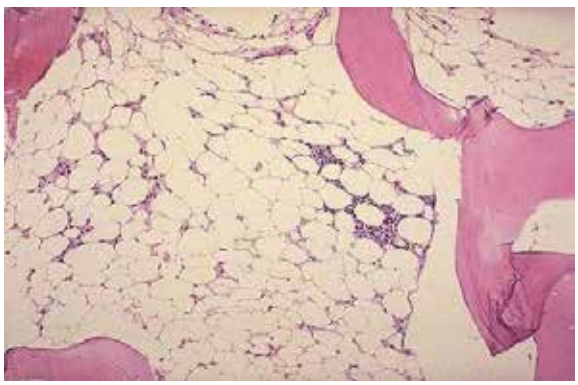
Plasma cells:- Mildly increased mature plasma cells(<50%)

Other cells:- No other abnormal cells or parasites are seen.

COMMENT:- Markedly reduced erythropoesis.

(??? Pure Red Cell Aplasia)

- Bone marrow biopsy:- Bone marrow examination revealed complete loss of erythroid lineage
- Some scattered, morphologically unremarkable erythroblasts were seen especially with IHC
- Other hematopoietic lineages are morphologically normal and show intact maturation.
- So,final diagnosis of Pure red cell aplasia made.and further investigations were done to rule out causes for secondary PRCA.



ANA report
Indirect immune fluorescence by using human epithelial cells(HEp-2) and primate liver.

Fluorescence pattern=+++ +ve for speckled pattern.

But ANA blot for Sm,nRNP,SS-A,SS-B was negative.

Ig M against Parvo B19 virus was negative.

HBsAg,HIV by ELISA,Anti HBC were negative.

Ultimately idiopathic PRCA is established as a cause of severe anaemia.

Discussion:
Pure red cell aplasia (PRCA) or erythroblastopenia refers to a type of anemia affecting the precursors to red blood cells but not to white blood cells. In PRCA, the bone marrow ceases to produce red blood cells. The condition has been first described by Kaznelson in 1922.

Major causes are:
- Autoimmunity
- Thymoma
- Viral infections such as herpes parvovirus B19(Fifth disease) Lymphoproliferative
- Association of pure red cell aplasia with T large granular lymphocyte leukemia is well recognized, especially in China.
- Idiopathic. Many cases of PRCA are considered idiopathic in that there is no discernible cause detected.
- Drugs such as mycophenolic acid or erythropoietin.
- Congenital. The term "hereditary pure red cell aplasia" has been used to refer to Diamond-Blackfan anemia.

Treatment
- PRCA is considered an autoimmune disease as it will respond to immunosuppressant treatment such as cyclosporine in many patients though this approach is not without risk.
- It has also been shown to respond to treatments with Rituxan and Tacrolimus.

REFERENCES

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