World Journal of Medical Sciences 10 (1): 41-42, 2014 ISSN 1817-3055 © IDOSI Publications, 2014 DOI: 10.5829/idosi.wjms.2014.10.1.8284

Coombs Negative Hemolytic Anaemia in Wilsons Disease

E. Halleys Kumar and A. Radhakrishnan

Sree Balaji Medical College, Chrompet, Chennai, India

Abstract: Wilson's disease is a rare inherited disorder of copper metabolism. Liver and brain are the main organs affected. Hemolytic anemia is a rare clinical manifestation of Wilson's Disease. We present a case of Wilson's Disease who presented with hemolytic anemia.

Key words: Wilson's Disease • Hemolytic Anemia

INTRODUCTION

Wilson's disease is an autosomal recessive disorder of copper metabolism characterised by excessive amount of copper in liver, brain, eye and other body tissues. The main clinical symptoms are hepatic (42%) and neurological (34%) [1]. Hemolytic anemia is rare [2] We present this case for its rarity.

Case Report: 37yr, female patient a known case of Wilsons disease came to the out patient department with complaints of ecchymotic patches over right hand and purpuric spots over both lowerlimbs for two weeks. There was no history of trauma,gum bleeding, fever, weight loss, loss of appetite. Patient did not have any other medical illness. General examination showed anemia.

Her blood pressure was 120/80 mm hg, pulse rate was 82/min.

There were non palpable pupuric spots seen in both upper limb and lower limb echymosis, kayser flescher ring was present in both the eyes.

Patient had dysarthria. There was no cardiac and renal involvement.

Abdominal examination showed moderate splenomegaly, mild hepatomegaly.

Investigations: Hemoglobin was 6.8gm/dl, total leucocytes were 3550, platelet count was 52000 The differential count was N 69% L26% E4% M1%. Peripheral smear showed normocytic normochromic anemia, with pancytopenia. Serum bilirubin was 4mg/dl; direct bilirubin 0.4 and indirect bilirubin 3.9mg/dl. Serum albumin 3.8gm/dl, alkaline phosphatase 192 IU/l, reticulocyte count was 9%.





Urine for hemoglobin was absent. HBsAg and ANTI HCV were negative.

Coomb's test performed twice showed negative results.

Ultrasound abdomen showed moderate splenomegaly.

Bone Marrow Aspiration: Out of 500 cells studied 2 megakaryocytes, 156 myeloid series cells, 14 erythroid series cells are noted, indicating normoerythroid hyperplasia with lesser myeloid megakaryocytic cells.

D-pencillamine was stopped to confirm drug induced hemolytic anaemia but there was no improvement.

During followup patient had one episode of hemolysis and two units of blood transfusion was given.

Corresponding Author: Halleys Kumar, Sree Balaji Medical College, Chrompet, Chennai, India.

DISCUSSION

Wilson's disease is a rare inherited disease presenting between 5-35 years of Age [3] The disease does not manifest clinically before 4-5 years of age as it takes time for copper to accumulate in liver. Hepatic manifestations are common early and neurological symptoms are common in adolescents [4]. Hemolysis in Wilson's disease (10-15%) is uncommon and is due to deficiency of ceruloplasmin, a copper transport protein which results in excessive inorganic copper in blood circulation much of it which accumulates in red blood cells. The exact mechanism of hemolysis is not known. The diagnosis of Wilson's disease is based on KF rings, low serum ceruloplasmin levels and elevated basal urinary copper excretion. The deficiency of copper causes anemia and congenital inability to excrete copper which may lead to Wilson's disease [5]. Hemolytic anemia often remits and may occasionally recur. Copper toxicity has been characterized in patients with Wilson's disease, a genetic disorder that causes an abnormal accumulation of Cu in body tissue.

Manifestations of Wilson's disease include brain damage and progressive demylination, psychiatric disturbances-depression, suicidal tendencies and aggressive behavior-hemolytic anemia, cirrhosis of the liver, motor dysfunction and corneal opacities [6].

CONCLUSION

Hemolytic anemia is rare in Wilson's disease and-is considered a cause of anemia Hemolytic anemia may also be a presenting episode in Wilson's disease. This is a case of Coombs negative hemolytic anaemia in Wilsons disease patient not precipitated by D-pencillamine which is a rare presentation Hemolytic anemia with liver failure should make a doctor suspect Wilson's disease as a cause.

REFERENCES

- Grudeva-Popova, J.G., M.I. Spasova, K.G. Chepieva and Z.H. Zaprianov, 2000. Acute haemolytic anemia as an initial clinical manifestation of Wilson's disease. Folia Med. (Plovdiv), 42(2): 42-46.
- McIntyre, H., H.M. Clink, A.J. Levi, Comings and J.N. Sherlocks, 1967. Hemolytic anemia in Wilson's disease. N Eng. J. Med., 276: 439-444. doi:10.1056/NEJM 196702232760804.
- Roberts, E.A. and M.L. Schilsky, 2003. A practical guidelines on Wilson's Disease. Hepatology, 37(6): 1475-1492. doi:10.1053/jhep.20003.50252.
- Kalra, V., D. Khurana and R. Mittal, 2000.Wilson's disease-Early onset & lessens from a pediatric cohort in India. Indian Pediatri, 37: 595-601.
- American-Eurasian J. Agric. and Environ. Sci., 10(3): 434-439, 2011 ISSN 1818-6769© IDOSI Publications, 2011.
- World Journal of Agricultural Sciences, 9(1): 10-16, 2013, ISSN 1817-3047© IDOSI Publications, 2013; DOI: 10.5829/idosi.wjas.2013.9.1.1133.