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Case Report

Combined factor VIII and IX deficiency: role in puberty menorrhagia: a case report

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ABSTRACT

The prevalence of menorrhagia in adolescent populations with bleeding disorders varies between 14% to 48%. Hereditary bleeding disorders are an important cause of puberty menorrhagia and the consequences can be life threatening. We report a case of 12 years old girl, known case of Factor VIII and IX deficiency who presented to our centre with menorrhagia at menarche. This case is reported as combined factor deficiency is a rarity. Early diagnosis, consultation with haematologist and appropriate treatment are essential.

Keywords: Menorrhagia, Hereditary bleeding disorders, Puberty menorrhagia, Factor VIII and IX deficiency

INTRODUCTION

Puberty menorrhagia is defined as bleeding of more than 80 ml/cycle or for more than 7 days. Most common cause of puberty menorrhagia is immature hypothalamo-pituitary-ovarian axis. Other causes include hereditary bleeding disorders, hypothyroidism and anovulatory cycles due to polycystic ovarian disease. Haemophilia, though not very common, is an important cause for puberty menorrhagia and can be life threatening. It usually presents with excess bleeding at menarche with mid-cycle abdominal pain. History of repeated excess bleeding from gums or wounds along with family history of similar complaints may be corroborative.[1,2]

CASE REPORT

Miss SSK, a 12 year old girl presented with history of attainment of menarche 1 year ago. Her cycles were normal for 3 months and then she suffered from continuous bleeding p/v on and off with use of 6-7 pads per day. Bleeding was not associated with pain in abdomen or any other complaints. She was a diagnosed case of Factor VIII and IX deficiency since last 1 year during evaluation of menorrhagia and epistaxis. There was no history suggestive of thyroid disorder or tuberculosis. There was no significant family history.

On examination, the patient was stable with significant pallor. There was no evidence of petechiae or bruises. Thyroid, breast and systemic examinations were unremarkable. Abdomen was soft and non-tender. On local examination bleeding was seen. Per rectal examination revealed small uterus.

On investigations, haemoglobin was 7 g%, platelet count was normal. Significant abnormality was seen in coagulation profile with a normal prothombin time (PT 12 seconds) and abnormal activated partial thromboplastin time (APTT 78.40 vs. 28.0 control) and International Normalised Ratio (INR) of 1.49. Thyroid profile was within normal limits. Ultrasonography revealed normal uterus and ovaries. Her Factor VIII was 40% and Factor IX level was 54%. Haematologist opinion was sought for her. She was transfused 3 units of cryoprecipitate and 2 units of fresh frozen plasma and was started on tranexamic acid thrice daily. Oral iron and folic acid was given for correction of anaemia. She was

also advised Factor VIII and vWF transfusion but it was not done due to monetary constraints. Subsequently, she was started on oral contraceptive pills.

DISCUSSION

Bleeding disorders should be suspected in patients with puberty menorrhagia with history of repeated gum bleeding, delayed wound healing, easy bruising, epistaxis, bleeding from tooth extraction, repeated blood transfusions and family history.[3] Commonly seen disorders are haemophilia A and B (deficiency of Factor VIII and IX respectively), von willebrand disease, Factor V deficiency, Glanzmann's thrombasthenia and idiopathic thrombocytopenia. Though combined deficiencies as seen in our case are known to occur, they are rare and have severe consequences. Combined Factor VIII and IX deficiency is a rare X linked disorders with incidence of 1 in 1,000,000 cases. Treatment modalities used to control bleeding are tranexamic acid, progesterone alone or oral contraceptive pills, though often bleeding may not respond to these drugs alone. Factor VIII deficiency can be treated by factor VIII replacement therapy and spontaneous bleeding is controlled if the patient's factor VIII level is raised to 30-50% of normal.[4] Desmopressin provides an alternative means of increasing the plasma factor VIII level in milder haemophiliacs. The principles of replacement therapy are similar in case of factor IX deficiency.[4]

Hereditary bleeding disorders should be suspected in all cases of puberty menorrhagia. Detailed personal and family history may guide to the diagnosis and appropriate treatment may prevent life threatening consequences.

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REFERENCES

1. Bevan JA. Bleeding disorders: A common cause of menorrhagia in adolescents. J Pediatr. 2001;138(6):856-61
2. Mikhail S, Varadarajan R, Kouides P. The prevalence of hemostasis in adolescents with menorrhagia referred to a haemophilia treatment centre. Hemophilia. 2007;13:627-32.
3. Kenneth Kaushansky. Inherited deficiencies of coagulation factor II, V, VIII, X, XI, XIII, combined deficiency of factor V and VIII and of vitamin K dependent factors. Kenneth Kaushansky, Ernest Beutler, Marshall A Lichtman. Williams hematology. 8th edition. China, 2010. The McGraw-Hill companies, Inc.
4. Fogarty PF, Kessler CM. Hemophilia A and B. In Kitchens C, Kessler C, Konkle B, editors. Consultative hemostasis and Thrombosis; 3rd ed.Elsevier. 2013:50-53.

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