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Case Report: Acquired von Willebrand Disease Causing Puberty Menorrhagia Secondary to Hypothyroidism

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ABSTRACT

Eighteen-year-old patient presented with complaint of menorrhagia since menarche, along with severe anaemia. Investigations showed low vWF factor levels for her "B" Rh "+"ve- non-'O' blood group, without any family history of bleeding disorder or thyroid disfunction, further probing in thyroid function revealed severe hypothyroidism, though clinical examination did not show any signs suggestive of hypothyroidism. Oral Thyroxin for correction of hypothyroidism helped to stop menorrhagia effectively, showing consistency in diagnosis of Acquired von Willebrand Factor deficiency which in turn resulted in Factor VIII dysfunction secondary to hypothyroidism.

Key words: Menorrhagia, Acquired von Willebrand Syndrome, Hypothyroidism

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INTRODUCTION

Commonly puberty menorrhagia is thought as AUB causing anaemia, mostly treated by Tranexamic Acid/ hormonal therapy for long terms causing their own side effects without much of relief. It is not uncommon, as observed in clinical practice, subclinical or frank hypothyroid women presenting with symptoms of easy bruising and menorrhagia [1]. that resolves with thyroid replacement therapy within 3 to 6 months. Low vWF along with Factor VIII dysfunction, as a first to be noted investigation, creates confusion for treating patient. Obtaining a baseline TSH is a good starting point in such scenarios. Similar case was studied and reported in "Medical Case Reports", in 2012, where hypothyroidism was diagnosed much late. Patient as on present day is under control with oral thyroxin with regular cycles, married and having two kids [2].

CASE REPORT

18 years old unmarried girl visited the Gynae outpatient, with the chief complaints of irregular and prolonged menstrual cycles with duration of 8-10 days of heavy bleeding at interval of bleeding free period of 25 to 30 days since menarche which was attended at the age of 12 years with severe anaemia. Initial trying with Tranexamic acid would help to reduce amount of blood loss, but then duration of bleeding would prolong. Bleeding would stop only with large doses of Oestrogen followed and supported by Progesterone and then had additional complaints due to side effects of high-dose hormones. Family history did not indicate any family member suffering from bleeding disorder.

On general examination, positive signs showed severe pallor with puffiness of face and generalised oedema without any other feature of hypothyroidism, lymphadenopathy, or splenomegaly.

Investigations revealed the following

- Haemoglobin-5 gm%, CBC and Platelets within normal range.
- PBS: Microcytic Hypochromic.
- BT: 2.11' CT: Bleeding Time: 5.15'
- Blood group: 'B' Rh Positive.
- vWF Ag: 31.11% (Reference range for 'O' Blood Group: 52-154% and for Non-'O' blood group: 60-200%).
- PT: 16 seconds with control of 12 seconds.
- INR - 1.16.
- APTT- 38 seconds with control being 31 seconds.
- TSH: 20.08 uIU/ml (normal range: 0.25-5 uIU/ml).
- TPO: 76.14 U/mL: Positive (negative < 5.61U/mL).

Diagnosis: Low Type I von Willebrand disease with hypothyroidism.

Final diagnosis: Acquired von Willebrand disease secondary to hypothyroidism.

Treatment

- PCV Transfusion two units 3 days apart.

- I.V. Iron Sucrose 5 units.
- Inj Tranexamic acid followed by Oral 8 hrly scheduled treatment for menorrhagia which didn't help much.
- Tab Thyroxin Initially 50µg, stepped up to 100 µg daily.
- Menorrhagia subsided slowly over the period of 15 days after starting Thyroxin.
- There was no need to give Low Purity Factor VIII.
- vWF Ag done after two months, within normal range 108.02% (Ref range 60-200%.

DISCUSSION

Puberty menorrhagia is commonly considered as a case of abnormal uterine bleeding (AUB) due to hypothalamus -pituitary- ovarian axis dysfunction in early ages after menarche, treated commonly with Tranexamic acid / combination oral pills / progesterone only supports [3].

Von Willebrand Factor (vWF) gene gives instructions for making von Willebrand factor, essential for clotting blood through stabilization of Factor VIII to prevent blood loss after injury or in females, during menstruation. vWD is genetically inherited autosomal Dominant or recessive disorder. It may occur first time in the affected person without any other cases in the family, known as a de novo mutation [4].

Biochemically vWF is a large multimeric glycoprotein produced by Weibel-Palde bodies in endothelial cells, megakaryocytes: α- granules of platelets and from subendothelial connective tissues. In coagulation process to stop bleeding vWF is necessary for proper platelet adhesion, it also protects Factor VIII from getting degraded.

Commonly VWD presents as an inherited problems, Type I and Type 2A the most common, Type 2B and Type 2M less common, are autosomal dominant manner. VWD type 2N, type 3 is inherited in autosomal recessive. Acquired von Willebrand syndrome (AvWS) is non inherited bleeding disorder with similar laboratory findings as in congenital von Willebrand Disease.

Level and activity of vWF decides the diagnosis of AvWS based on assays measuring the level and activity of (vWF) which tend to be low though factor VIII levels may be normal with its normal coagulant activity. Pathogenesis: early degradation of factor VIII couldn't be stopped due to low vWF levels resulting in bleeding disorders, pts usually presents with mucocutaneous bleeding episodes which may be managed with local therapy and/or antifibrinolytic agents, desmopressin can be used in patients who are not responding to antifibrinolytic drugs. vWF/ factor VIII concentrates have been used in few patients. Administration of thyroid hormones can reverse this abnormality. Association between vWD and hypothyroidism is being cited recently, complete correction of the clotting abnormality after thyroxine supplementation has been cited. Deficient protein synthesis seen in hypothyroidism with reduction in clotting factor levels is the suggested patho-physiological mechanism [5].

The possibility of hypothyroidism should be considered in patients diagnosed as von Willebrand's disease. Even if hypothyroidism on prima face is diagnosed as a primary cause of menorrhagia, suggestion is to perform vWF levels as well, to differentiate between Congenital / Acquired von Willebrand Disease [6].

SUMMARY

Menorrhagia considered as a most common symptom of ovarian steroidogenesis disorder, either primary or secondary to hypothalamus – pituitary dysfunction, most of the time treated without keeping other causes in view, turns out sometimes ineffective, leading to chronic loss of blood, iron deficiency and chronic ill-health. This may also result in absentees from school and curricular disturbances in pubertal age group. Implementation of hormonal therapies as initial management of menorrhagia delays the diagnosis of basic hormonal disturbances. Investigations done for congenital bleeding disorder revealed the coagulation factors vWF deficiency along with raised TSH values- diagnosed as Acquired von Willebrand Syndrome secondary to hypothyroidism. Correcting Thyroxin deficiency with oral thyroxin normalised the vWF levels and helped treating menorrhagia.

DECLARATION

The case report submitted is an original case diagnosed as Acquired von Willebrand Disease secondary to hypothyroidism. Similar case was reported in 2012, a case of menorrhagia since menarche, initially diagnosed as congenital vWD causing menorrhagia not responding to treatment, though hypothyroidism was detected late. Treatment with oral Thyroxine resolved menorrhagia, woman got married and having two kids.

REFERENCES

1. Dalton RG, Savidge GF, Matthews KB, et al. Hypothyroidism as a cause of acquired von wdllebrand's disease. Lancet 1987; 329:1007-10099.
2. Manfredi E, Zaane BV, Gerdes VE, et al. Hypothyroidism and acquired von Willebrand's syndrome: A systematic review. Haemophilia 2008; 14:423-33.
3. Kujovich JL. von Willebrand's disease and menorrhagia: Prevalence, diagnosis, and management. Am J Hematol 2005; 79:220-228.
4. Kumar S, Kishore R, Gupta V, et al. Prevalence and spectrum of von Willebrand disease in Eastern Uttar Pradesh. Indian J Pathol Microbiol 2010; 53:486.
5. Kumar S, Kishore R, Gupta V, et al. Prevalence and spectrum of von Willebrand disease in Eastern Uttar Pradesh. Indian J Pathol Microbiol 2010; 53:486.
6. Shankar M, Lee CA, Sabin CA, et al. von Willebrand disease in women with menorrhagia: a systematic review. Int J Obstetr Gynaecol 2004; 111:734-740.