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CASE REPORT OF GLANZMANN THROMBASTHENIA IN PUBERTY MENORRHAGHIA

KEY WORDS: Glanzman thrombasthenia, glycoprotein deficiency, puberty, menorrhaghia, uncommon

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ABSTRACT

Glanzmann Thrombasthenia is an uncommon autosomal recessive bleeding syndrome affecting the megakaryocyte lineage and characterized by no platelet aggregation. Platelets have qualitative and quantitative deficiencies of platelet of fibrinogen receptor. The bleeding manifestations are typically purpura, peliosis, epistaxis, gum bleeding, and menorrhagia, gastrointestinal bleeding. Peripheral blood smear by light microscopy shows normal thrombocyte count, size, and granularity and abnormal platelet assays .It is an uncommon coagulopathy in which the platelets has defective or low levels of glycoprotein IIb/IIIa (GpIIb/IIIa), that is a receptor for fibrinogen. With proper treatment modality, Glanzmann's Thrombasthenia features a good prognosis. Blood transfusions and platelet transfusions are commonly being used nowadays.

CASE REPORT :-

A 21 year old women who is a nulligravida, married for 3 years, in a consanguineous marriage, she had complaints of lower abdominal pain and nausea, since 10 days prior to the day when she attended gynaecology OPD. She has regular menstrual cycles and has history of heavy menstrual cycles since menarche, changing 7-8 pads per day since she attained menarche. Urine bioassay was negative. Vitals were stable. She had history of epistaxis and bleeding gums frequently since childhood. Patient also had a past surgical history of Septoplasty done for deviated nasal septum 5 years ago. On abdominal examination, there was a mass felt in the midline, and size couldn't be assessed. Mass was firm in consistency, not mobile. On vaginal examination, a left sided adnexal mass could be felt around 12 X 15cms along side forniceal tenderness and normal sized anteverted uterus was felt, other fornices were free and non tender. A heterogenous lump 13 X 20 X 8 cms, haemorrghagic in nature arising from the left adnexa was pictured on ultrasonography. Haemoglobin was 8gms, platelets were 3.8lakhs/cumm and coagulation profile was within normal limits. An outsized multicystic lesion of 11x14x16 cms within the left adnexa with subacute haemorrhage was seen in CT scan. CA- 125 was raised 66 IU/L. Alternative tumour markers were normal. Clot retraction was absent, which was noted after 24 hours, where blood is allowed to clot in a test tube and there was no further clot retraction after 24 hours, which is an Implicative of GT.

Glycoprotein IIb/IIIa flow cytometry shows 93.22%. She was diagnosed as a case of Glanzmann Thrombasthenia and in view of high risk for surgery, she was managed conservatively. She has recieved Tranexamic acid 500 mg per oral thrice daily and platelet transfusion to regulate the current bleeding episode and double doses of factor VII (Proconvertin) 2mg were administered intravenously with six units of platelet transfusion as a part of conservative management. After one week, at the time of discharge the dimensions of the mass has reduced clinically to around 14 weeks size. Counselling was given regarding the utilization of combined oral contraceptive pills to stop future episodes of heavy menstrual bleeding. Since she features a requirement for repeated blood transfusions, and was immunized with Hepatitis B vaccine in order to wardoff transfusion associated hepatitis.

DISCUSSION:-

GT is a most uncommon autosomal recessive disorder seen in consanguineous marriages. Heterozygotes are not symptomatic and have normal platelet function tests. It is most commonly seen in French Gypsies, Iraqi Jews, Jordanian Arabs, and South Indians. The disorder has been classified into three varieties initially based on amount of fibrinogen and extent of clot retraction, before the concept of GPIIb/IIIa complex was understood. These categorizations have been adapted to the more recent molecular understanding of the disease.

Type I and Type II are the outcome from alteration of GPIIb or GPIIIa that cause absence of (i.e., less than 5% of normal) or reduced expression (i.e., 10-20% of normal), respectively, of the IIb/ IIIa complex on the platelet surface and also, atypical gene mutations may lead to defective complex formation that is expressed at adequate or inadequate quantity resulting in the Type III, or variant form.

Advanced modalities of treatment are being used. Like factor VII a (Proconvertin), which is additionally utilized in severe episodes of bleeding to regulate it. The recommended dose is 90mcg/kg. In patients who have already or prone to develop allo-immunization, the simplest treatment of choice is plasmapheresis.

SUMMARY :-

We came to a conclusion that Glanzmann's Thrombasthenia is one among the causes of puberty menorrghagia, though not commonly encountered. Tranexamic acid and factor VII (Proconvertin) is the only treatment modality for conservative management apart from transfusions.

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