INTRODUCTION
Hemorrhagic and thrombotic disorders are largely mediated by congenital or acquired abnormalities of blood coagulation, platelet number, or platelet function. Abnormal bleeding can result from excessive expression of plasminogen activators or from deficiencies of fibrinolysis inhibitors. Most inherited bleeding disorders result from genetic defects that reduce hemostatic protein expression, secretion, or function. Inherited conditions that cause bleeding by increasing gene expression are uncommon and include Quebec platelet disorder (QPD), an autosomal dominant disorder with high and possibly complete penetrance. QPD is associated with a unique gain-of-function abnormality in fibrinolysis due to increased platelet stores of urokinase plasminogen activator (uPA) without systemic fibrinolysis or increased uPA in plasma, urine, or CD34 hematopoietic progenitors. QPD increases risks for a number of bleeding symptoms, including delayed-onset bleeding after hemostatic challenges that respond only to fibrinolytic inhibitor therapy. Diagnostic tests for QPD include assays for increased platelet uPA and alpha granule protein degradation from intraplatelet plasmin generation, platelet function test is assessed with the help of platelet aggregation assay by exposing platelets to various stimulants like ADP, arachidonic acid, collagen, epinephrine, thrombin, and ristocetin. In QBD, platelets show very abnormal aggregation with epinephrine because there is defect in alpha granules proteolysis of proteins and a deficiency of alpha granule multimerin, a protein that binds factor V within granule and leads to a decreased content of factor V and several other proteins like fibrinogen, vWF etc.

CASE REPORT
A 33 years old woman presented with history of epistaxis and gum bleeding since childhood and menorrhagia and bleeding per vaginum after puberty, also had history of excessive blood loss after birth of child. Her coagulation profile was normal but platelet function testing by platelet aggregation assay showed abnormal aggregation of platelet with epinephrine. This type of response is seen in “Quebec platelet disorder” which is a rare autosomal dominant disorder of platelet function characterized by increased bleeding after any injury or trauma.

Key words: Quebec platelet disorder, Platelet function test, Bleeding time, Abnormal aggregation, Epinephrine.
raised strong suspicion of tuberculosis as effusion was
exudative lymphocytic, which was augmented with
positive tuberculin skin test (Mantoux test). She was
started on anti-tuberculosis treatment.

Further workup was done due to her symptoms of
bleeding. Her complete blood count showed hemoglobin
of 5.2 g/dl with mean corpuscular volume of 56 FL. The
white cell and reticulocyte count were normal. Peripheral
smear showed microcytic hypochromic anemia. Her
platelet count was normal. Coagulation profile showed
normal PT and APTT which bleeding time (BT) was
more than 9 minutes. Platelet function testing was done
by platelet agregometry which showed normal response
to other factors like collagen, ADP, thrombin, but an
abnormal aggregation response to epinephrine. All
other factor assays like factor VIII, vWF, factor XIII were
normal. This type of platelet response is seen in Quebec
platelet disorder. So she was diagnosed to have Quebec
platelet disorder and was started on fibrinolysis inhibitors
(tranexamic acid) after which her gum bleeding stopped
but she needed blood transfusion for severe anemia.

**DISCUSSION**

Quebec Platelet Disorder (QPD) is an autosomal
dominant bleeding disorder which is more prevalent in
province of Quebec in Canada. The disorder is
characterized by large amounts of the fibrinolytic
enzyme urokinase-type plasminogen activator (u-PA) in
platelets. Consequently, stored platelet plasminogen is
converted to plasmin, which is thought to play a role in
degrading a number of proteins stored in platelet α-
granules. These proteins include platelet factor V, Von
Willebrand factor, fibrinogen, thrombospondin-1, and
osteonectin. There is also a quantitative deficiency in the
platelet protein multimerin 1 (MMRN1). Furthermore, upon
QPD platelet activation, u-PA can be released into
forming clots and accelerate clot lysis, resulting in
delayed-onset bleeding (12-24 hours after injury).
Individuals with QPD are at risk for experiencing a
number of bleeding symptoms, including joint bleeds,
hematuria, and large bruising. This patient likewise also
experienced bleeding episodes especially from gums,
and menorrhagia. The genetic cause of QPD has not yet
been determined. Whether the abnormal bleeding in this
arisor results from the degradation of platelet
hemostatic factors, premature lysis of thrombi due to
u-PA release from platelets, or both, is unresolved. However,
theses of transgenic mice with platelet-specific over-
expression of u-PA suggest that premature clot lysis due
to localized release of u-PA from platelets is the
predominant cause of bleeding.

In bleeding patients with the factor V Quebec platelet
disorder, platelet transfusions are generally without
effect; fibrinolytic inhibitors are reported to be effective.
Tranexamic acid andaminocaproic acid are lysine
analloges that bind to the kringle domains of plasminogen
and disrupt interactions between plasminogen (and
plasmin) and lysine residues within fibrin. Uptill now
tranexamic acid has benifitted in resolving bleeding
episodes in our patients. A meta-analysis of clinical trials
indicated that lysine analogues significantly reduce
perioperative blood loss in patients undergoing coronary
t artery bypass grafting with cardiopulmonary bypass,
without increasing the incidence of myocardial infarction.
This lady, admitted with history of bleeding gums,
epistaxis and menorrhagia and history of multiple blood
transfusions, was diagnosed to have “Quebec platelet
disorder” on the basis of history, examination and
laboratory findings of increased bleeding time and on
platelet function testing by platelet aggregation assay
showing abnormal aggregation to epinephrine. She was
managed with giving lysine analogues like Tranexamic
acid which act as fibrinolytic inhibitors. She responded
well to treatment and her bleeding symptoms resolved.

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