CASE REPORT

Enigma of Menorrhagia Since Menarche

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Received 14 July 2007; received in revised form 03 August 2007; accepted 26 November 2007; published online 10 March 2008

Abstract

Blood loss in the normal menstrual cycle is self-limited due to the action of platelets and fibrin. Individuals with thrombocytopenia or coagulation deficiency may have excessive menstrual bleeding. Coagulation disorders are the most common inherited cause of menorrhagia among young girls. The incidence of coagulopathy in puberty menorrhagia may be found in 12 to 33%. We present here, a rare case of a 24 year old girl having menorrhagia since menarche secondary to coagulopathy in liver disease and portal hypertension. A brief review of the literature is given along with references. Coagulopathy secondary to liver dysfunction presents with raised thrombin time, d-dimer and fibrinogen degradation products, hypo or dysfibrinogenemia with platelet function defects. Hypo or dysfibrinogenemia with platelet function defects, raised prothrombin time and aPTT have been described in the presence of normal liver enzymes. The contributory factors for defective hemostasis in portal hypertension are attributed to decreased synthesis of procoagulant and anticoagulant proteins, impaired clearance of activated coagulation factors, nutritional deficiency (vitamin K and folate), synthesis of functionally abnormal fibrinogen with splenomegaly due to sequestrational thrombocytopenia and qualitative platelet defects. In portal hypertension more than 80% of the platelet pool is sequestered in the spleen; platelet count rarely falls below 30 000/mm^3 and spontaneous bleeding is uncommon. Bleeding can be fatal in young girls at menarche if not previously diagnosed. Treatment is rewarding and prognosis good provided platelet transfusion is available.

Keywords: puberty menorrhagia, platelet dysfunction, coagulation disorders, liver disease

Özet

Menarştan İtibaren Menoraji Bilmecesi

Normal âdet siklusunda kan kaybı, trombosit ve fibrin sayesinde kendi kendini sınırlayan bir olaydır. Trombositopenisi olan ya da koagülasyon eksikliği olanlarda aşırı âdet kanaması görülebilir. Koagülasyon bozuklukları genç kızlarda gözlenen menorajinin en sık kalıtımsal sebebidir. Koagülopati insidansı pubertal menorajide %12-33 arasındadır. Bu sunumda, 24 yaşında karaciğer rahatsızlığı ve portal hipertansiyon nedeniyle oluşan sekonder koagülopatisi nedeniyle menarştan itibaren menoraji şikâyeti olan nadir bir hastadan bahsedilmektedir. Bu konudaki literatür özeti de yapılmıştır. Karaciğer hastalığına sekonder koagülopati artmış trombin zamanı, D-dimer ve fibrinojen yıkım ürünleri, hipo-disfibrinojenemi ve trombosit fonksiyon defektleri ile olmaktadır. Hipo-disfibrinojenemi ve trombosit fonksiyon defektleri, artmış protrombin ve APTT süresi, normal karaciğer enzimleri varlığında da gözlenebilir. Defektif hemostaza yol açan faktör portal hipertansiyonda prokoagülan ve antikoagülan proteinlerin azalmış sentezi, aktive olmuş koagülasyon faktörlerinin yıkımının azalması, beslenmedeki eksiklik (K vitamini, folat), fonksiyonel olarak anormal fibrinojen sentezi, splenomegali ile azalan trombosit sayısı ve kalitesi olabilir. Pulmoner hipertansiyonda trombosit havuzunun %80'den fazlası dalakta sekestre olmakta ve trombosit sayısı 30 000/mm^3 altına düşmektedir. Ancak spontan kanama nadirdir. Kanama önceden bilinmezse menarş ölümcül olabilir. Tedavi yüz güldürücüdür ve trombosit transfüzyonu yapılabilirse prognoz genellikle iyidir.

Anahtar sözcükler: puberte menorajisi, trombosit disfonksiyonları, koagülasyon hastalıkları, karaciğer hastalığı

Introduction

Blood loss in the normal menstrual cycle is self-limited due to the action of platelets and fibrin. Individuals with thrombocytopenia or coagulation deficiency may have excessive menstrual bleeding. Coagulation disorders are the most common inherited cause of menorrhagia among young girls. The incidence of coagulopathy in puberty menorrhagia may be found in 12 to 33% of the cases. We present here a rare case of a 24 year old girl having menorrhagia since menarche, secondary to coagulopathy in liver disease with portal hypertension. A brief review of the literature is given along with references.

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Introduction

Blood loss in the normal menstrual cycle is self-limited due to the action of platelets and fibrin. Individuals with thrombocytopenia or coagulation deficiency may have excessive menstrual bleeding. Coagulation disorders are the most common inherited cause of menorrhagia among young girls. The incidence of coagulopathy in puberty menorrhagia may be found in 12 to 33%. We present here, a rare case of a 24 year old girl having menorrhagia since menarche secondary to coagulopathy in liver disease and portal hypertension. A brief review of literature is given along with references.

Case Report

A 24 year old, unmarried girl presented in the gynecology outpatient department on November 28th, 2006 with complaints of heavy, frequent periods since menarche and a history of gum bleeding, epistaxis, hematuria, melena and easy bruisability in 2 to 3 episodes in the last one year. She attained menarche at 16 years. Her cycles lasted 8-9 days every 10-15-20 days, heavy flow with clots and congestive dysmenorrhoea.

On examination she was severely anemic. Her pulse was 110/min, blood pressure 100 mmHg systolic with an ejection systolic murmur on cardiovascular examination. Secondary sexual characters were well developed, breast and thyroid showed no abnormality. Per abdominal examination revealed spleenomegaly 6 cms below costal margin. On per rectal examination uterus was normal in shape and size with no pelvic mass. A clinical diagnosis of polymenorrhagia since menarche with severe anemia probably due to coagulation disorder was made. The patient received two units of packed cells. The results of her tests are given in Table 1. The patient responded to hemostatics, hematinics and was started on high dose oral contraceptive pills (50 µg ethinyl estradiol).

The coagulation profile suggested mild disseminated intravascular coagulation with thrombocytopenia, normal bleeding and clotting time but poor clot-retraction time. Immunological work-up was normal. Tests for platelet function revealed abnormal platelet function studies (secondary to liver disease) (Figure 1). The presence of normal vW factor and Glyc IIb-IIIa on her platelets ruled out von Willebrand disease, Bernard Soulier syndrome and Glanzmann's thromboesthenia.

The patient's bone marrow biopsy was compatible with hyperspleenism. Her liver function test showed mildly elevated OT/PT with reversed albumin:globulin ratio, and normal bilirubin level. Ultrasound showed spleenomegaly of 16 cm with the portal vein dilated to 14 mm, patent spleenoportal axis with biphasic flow velocity of 11 cm/sec, suggestive of early portal hypertension. Upper GI endoscopy revealed a single short column of Grade I esophageal varices with an impression of early portal hypertension. Uterus and bilateral adnexa were normal. Viral markers were negative.

A final diagnosis of acquired coagulopathy secondary to liver dysfunction and portal hypertension was made. Patient is still on oral contraceptive pills and hematinics. Her bleeding is under control and she is awaiting a liver biopsy.

Discussion

Coagulation disorders are the most common inherited cause of menorrhagia among young girls (1).

Kadir et al. (2) reported menorrhagia as the most common presenting feature in females with platelet function defects. Coagulopathy secondary to liver dysfunction presents with raised thrombin time, d-dimer and fibrinogen degradation products, hypo or dysfibrinogenemia with platelet function defects, raised prothrombin time, d-dimer and fibrinogen degradation products. Hypo or dysfibrinogenemia with platelet function defects, raised prothrombin time and aPTT have been described in the presence of normal liver enzymes. There is decreased aggregation with arachidonic acid, adenosinodiphosphate and adrenaline. Sallah et al (3). studied the contributory factors for defective hemostasis in portal hypertension and attributed it to decreased synthesis of procoagulant and anticoagulant proteins, impaired clearance of activated coagulation factors, nutritional deficiency (vitamin K and folate) synthesis of functionally abnormal fibrinogen with splenomegaly due to sequestrational thrombocytopenia and qualitative platelet defects.

In portal hypertension more than 80% of the platelet pool is sequestered in spleen, platelet count rarely falls below 30 000/mm^3 and spontaneous bleeding is uncommon (4,5). Bleeding can be fatal in young girls at menarche if not previously diagnosed. Treatment is rewarding and prognosis good provided platelet transfusion is available.

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