CASE REPORTS

Chronic Disseminated Intravascular Coagulation: A Case Report

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Abstract:
In health there is a balance between the coagulation and anti-coagulation systems, but in disseminated intravascular coagulation (DIC) the coagulation mechanism is activated inappropriately and in a diffuse way. This may lead to thrombosis, but more often haemorrhage occurs when the clotting factors are exhausted. DIC may present as acute, subacute, and rarely chronic form. Here we present a case of chronic DIC following pelvic inflammatory disease (PID) as a consequence of repeated menstruation regulation (MR). We treated her with fresh frozen plasma, fresh blood, doxycycline with significant clinical improvement.

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Introduction

Disseminated intravascular coagulation (DIC) is a syndrome characterized by inappropriate and excessive activation of haemostatic system. DIC is usually initiated by exposure of blood to tissue factor, presents on the cell surface that surround blood vessels. Brain and placenta are especially rich in tissue factor[1]. DIC results from trauma, obstetric accident, diffuse vascular injury, increased endothelial permeability or circulating cancer cells resulting pathologic activation of the extrinsic and/ or intrinsic pathway of coagulation or impairment of clot inhibiting influences[2,3]. In subacute or chronic DIC, there is slow activation of haemostatic system, with spontaneous bruising rather than major clinical bleeding episodes. Rarely, a chronic compensated form of DIC can continue for many years, usually associated with intrauterine infection, internal malignancy or vascular malformations. The principles of management of DIC are control of haemorrhage, elimination of precipitating factors and specific coagulation factor replacement therapy. Here we are going to present such a rare case of chronic DIC.

Case report

A 35-years-old woman was admitted in Medicine department of Bangabandhu Sheikh Mujib Medical University (BSMMU) on 11th March 2004 with the complaints of echymoses of variable sizes and color for 5 months. She complained of epistaxis, gum bleeding, haematuria, melaena and menorrhagia. She gave history of prolonged bleeding after minor trauma and delayed wound healing for the said duration. She stated about repeated oral ulcers without any fever, arthralgia/ arthritis, bone pain, photosensitivity or malar rash. She had no history of such previous bleeding episode either in her or in her family members. She was mother of two healthy children. She gave a history of MR seven months back, but she remained amenorrhic for more than two months. Her urinary HCG test was positive at that time, and sonogrphic examination revealed retained product of conception and Dilatation and curettage (D&C) was done. She had no significant drug history except oral contraceptive pill. On general examination, she was anxious, moderately anemic, with multiple non-tender, non-palpable echymoses on the extensor surface of both limbs, of variable sizes and colours, which didn't blanch on pressure.

Her peripheral blood film was suggestive of **anemia of chronic disorder** (Haemoglobulin 7.8gm/dl) with **high ESR** (98mm in 1st hour) and normal platelet count (155,000/uL). Sonogaphic examination of pelvic organ revealed that uterus was mildly enlarged. **Clotting time** was prolonged i.e. 18 minutes, with normal bleeding time. Her **prothrombin time** was 48 second (control: 13 sec), activated partial thromboplastin time (**APTT**) was 128 seconds (normal 30-40 seconds), **thrombin time** was 22 seconds (control: 16 sec) and **factor VIII activity** was 25% (normal range 60-150%). Her **ANA** was positive in low titer (two fold rise) and **anti-dsDNA** and **anti-Sm** test were negative. Initially she was suspected of having vasculitic disorders and treated with steroid, but there was no clinical improvement. Her plasma **fibrinogen** was 2.37g/l (normal 1.5-4.0gm/l) but fibrin degradation

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product (**FDP**) and **D-dimer** (normal ‹0.2mg/dl) were 5 µgm /l and 0.5 µgm/ ml respectively, i.e., both were increased. Per vaginal examination revealed that uterus was bulky with fluid collection in the pouch of Douglas and diagnosed a case of chronic DIC due to pelvic inflammatory disease (PID) as a consequence of retained products of conception. She was treated with cap. Doxycycline (100 mg) b.d. for one month as well as with five units of fresh frozen plasma and one unit of whole fresh blood with significant clinical improvement. After a few days, her coagulation profile was repeated and was within normal limit (**PT** -14 seconds, **APTT** -38 seconds, **TT** -12 seconds, plasma **fibrinogen** 250 mg/dl, **FDP**-<05 µgm /ml and **D-dimer**-<0.5 µgm /ml).

Discussion:

Disseminated intravascular coagulation is an acute, subacute or chronic thrombohaemorrhagic disorder occurring as a complication in a variety of diseases. It is characterized by activation of the coagulation sequence that leads to the formation of microthrombi throughout the microcirculation of the body and accelerated fibrinolysis[4].

DIC usually presents as an acute, often catastrophic, acquired haemorrhagic tendency. Rarely it can also manifest as a low grade disorder with predominantly thrombotic manifestations[5]. Chronic DIC may occur due to intrauterine fetal death, giant haemangiomas (Kasabach Merritt syndrome) or adenocarcinoma. Intrauterine infection causes endotoxins to be released into the maternal circulation and damage of the blood vessels releases thromboplastins, which causes chronic DIC. Other etiologic factors of chronic DIC are aneurysms, vasculitis, leiomyomas, hydatidiform mole etc[6]. In chronic DIC, intravascular coagulation and fibrinolysis don't proceed fast enough to outstrip the rate of synthesis of clotting factors or inhibitors. This may simply reflect low grade, weak or intermittent activating stimulus, in which case DIC is often mild and asymptomatic[7]. Destruction and production of coagulation factors and platelets are balanced. The pathophysiology of such chronic, subacute or compensated DIC is fundamentally the same as that in the acute case. Nevertheless, the distinction is valuable because the clinical pictures and laboratory findings in the chronic form are quite variable and may be diagnostically confusing[6].

In chronic DIC, superficial but extensive ecchymosis of extremities may develop intermittently or may persist for weeks or months. Recurrent episodes of epistaxis or more serious internal mucosal bleeding may punctuate the course. DIC, caused by carcinoma, may cause bleeding or recurrent deep and superficial venous thrombosis[8]. Intrauterine fetal death may produce chronic DIC, particularly if the fetus is retained for several weeks[9]. In chronic or subacute DIC, prothrombin time, activated partial thromboplastin time and thrombin time are prolonged but platelet count may be normal or low. Fibrinogen concentration may be normal or low. However, there is usually an increase in fibrin degradation product (FDP) and increased level of D-dimer[5].

Conclusion:

Chronic or subacute DIC is a rare, catastrophic haemorrhagic disorder or sometimes shows a thrombo-embolic manifestation. We diagnosed her as a case of chronic DIC due to retained product of conception. But we failed to explain why her ANA was increased. For this reason she should further followed up as she may develop any connective tissue disease. It is further to see whether ANA disappear spontaneously.

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