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Case Report

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ACQUIRED BLEEDING DISORDER DUE TO SECONDARY ANTIPHOSPHOLIPID SYNDROME IN A YOUNG FEMALE- CASE REPORT

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ABSTRACT
Antiphospholipid Syndrome (APLA)is an autoimmune disease which is more commonly presents with arterial and venous thromboembolism rather than bleeding.We present a case of Secondary Anti phospholipid syndrome without thrombocytopenia in a 16 year old unmarried female presented with isolated per vaginal bleeding with deranged PT/INR, aPTT. Serum ANA and Anti dsDNA were positive withthe presence of ACLA IgG/IgM antibodies. Mixing study and factor VIII activity assay showed the presence of antibodies against factor VIII.

KEYWORDS: Antiphospholipid syndrome, Lupus anticoagulant, Acquired Factor inhibitor, Acquired bleeding disorder.

INTRODUCTION
Antiphospholipid Syndrome is an autoimmune disease which is more common in females with a varied presentation ranging from recurrent pregnancy losses, arterial and venous thromboembolism to life threatening catastrophic APLA syndrome.[1] Since the clinical presentation can be varied the disease requires a high index of suspicion particularly in females of reproductive age group. Bleeding is not a common manifestation of antiphospholipidsyndrome. Only a small number of cases are reported, when an acquired thrombocytopathy or an acquired factor VIII inhibitors or a severe thrombocytopenia is present. Aquired factor VIII deficiency is very rare but this may present with catastrophic bleeding episodes, despite having no previous history of a bleeding manifestation.[2,3] Here we are presenting a case of secondary Antiphospholipid Syndrome with bleeding due to acquired deficiency of factor VIII.

CASE REPORT
Sixteen year unmarried female came to us with history of menorrhagia since menarche (2 years duration). The cycles were otherwise regular. For last 2 month she had continuous menstrual flow. There was no history suggestive for systemic bleeding diathesis in the form of hematemesis, melena, haemoptysis, hematuria, skin rash, mucosal bleed from other sites, vision loss or joint swelling. There was no history suggestive for local trauma/bruising/sexual intercourse. She did not complaint of any photosensitivity, joint pains, hair loss, syncope, seizures, Raynaud's phenomenon, apthousulcers. There was a history of three units blood transfusion and of taking oral haematinics. She was not on any other medications (NSAIDs (Mefenamicacid/Estrogen/Progestrogen therapy). Family history was not suggestive of any bleeding disorder. On examination she had severe anemia, there was no icterus, lymphadenopathy, fundal bleed, hepatosplenomegaly and sexual maturity index was normal. On investigations her Hb-7.6 g/dL,TLC-5000/µLwith normal differential distribution, Plt-1.89 lac/µl, MCV-92 fL. In urine Routine/Microscopy -alb 1+, sugar/RBC -Nil. Renal function and liver function tests were with in normal limits.Hormonal assay including Serum Prolactin, FSH, LH and thyroid function were within normal limit. Collagen profile shown raised ANA (6.4, N<1) and antidsDNA (585 IU/ML, N<40). Her plasma also showed presence of Lupus anticoagulant (LAC) and raised both anti CardiolipinIgM(48,N<10)/IgG(37,N<10).Her PT and APTT were prolonged (PT-13.8s/22.3s with INR 1.61, APTT-29.7s/43.7s. Mixing studies showed the prolonged APTT level, so possibility of factor inhibitor in patient plasma suspected. Further tests were carried out and she was found positive for coagulation factor VIII

inhibitors.In view of raised ANA and AntidsDNA along with presence of LAC and raised anti CardiolipinIgM/IgG diagnosis of Secondary Antiphospholipid Syndrome with Acquired bleeding disorder was made. She was given oral Prednisolone 1mg/kg for 4 week then dose tapered. After 12 weeks her Hb-12g/dL, ANA-5.5,AntidsDNA-168IU/ML,PT-INR12.5/13.6-1.08,aPTT-29.3/31,LAC-Negative and Anti Cardiolipin antibody IgM(45),IgG(27). Now She is totally asymptomatic with normal cycle and taking Prednisolone 5 mg alternate day.

DISCUSSION

The diagnostic criteria is based on pregnancy morbidity of recurrent fetal loss and gestational and comorbid illnesses like eclampsia/ preeclampsia. The lab criteria includes testing for Lupus anticoagulant (LA), anticardiolipin antibodies IgG/ IgM (aCL), Anti–beta-2 glycoprotein I (Anti β2GPI) on two occasions 12 weeks apart.[4] APLA syndrome usually manifests with thrombotic manifestation and very rarely with bleeding diasthesis unless complicated by thrombocytopenia. Some time very few cases had also reported with acquired factor VIII deficiency in secondary APLA. Factor VIII work as a cofactor for factor IX in the coagulation pathway and a deficiency of factor VIII thus decreases the generation of thrombin on the activated platelets. Factor VIII is synthesized as a precursor protein with an A1-a1-A2-a2-B-a3-A3-C1-C2 domain structure. Most of the time acquired factor VIII inhibitors bind to the A2,A3 or C2 domains. The binding of factor VIII with phospholipid is disrupted by anti C2 antibodies.[5,6,7]

CONCLUSION

Bleeding in acquired antiphopholipid syndrome can be easily missed, with some time disastrous results. Early recognition is critical, since early therapy directed to the antibodies formation can be life saving.

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