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A CASE OF RECURRENT THROMBOSIS IN CHRONIC IMMUNE THROMBOCYTOPENIC PURPURA- CLINICAL PARADOX AND THERAPEUTIC CHALLENGES.

Haematology

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ABSTRACT

ITP is characterized by humoral and cellular autoimmunity-mediated thrombocytopenia. Although clinical history is marked by bleeding manifestations, an increased thrombotic risk has been recognized in ITP1. Theories proposed for the same include platelet microparticles which are thrombogenic due to high surface expression of phosphatidyl serine, presence of antiphospholipid antibodies, effect of steroids, intravenous immunoglobulin, thrombopoietin receptor agonist causes sudden rise in platelet counts predisposing to thrombosis2. 29 years old female,36 weeks gestation, G2P1L1, known case of immune thrombocytopenic purpura presented with complaints of petechia in bilateral lower limbs and bleeding gums. In view of low platelet counts and bleeding manifestations patient was started on steroids following which patient developed bilateral deep venous thrombosis and pulmonary embolism. Patient was treated with steroids, anticoagulation as per platelet count monitoring and other supportive measures. Patient symptomatically improved and follow up reports showed normal platelet count with resolution of thrombosis It's important to recognize the complex nature of immune thrombocytopenic purpura (ITP) and the risk factors associated with it, such as increased thrombotic risk. The development of deep venous thrombosis and pulmonary embolism in the described case is concerning and underscores the need for a multidisciplinary approach in managing ITP, particularly in pregnant patients. Steroids are a common treatment for ITP, but their thrombogenic potential should be carefully considered, as seen in this case. Monitoring platelet counts and managing thrombotic risk with anticoagulation is essential for a positive outcome. This case highlights the importance of individualized care and close follow-up for patients with ITP, especially during pregnancy.

KEYWORDS

Immune thrombocytopenic purpura, steroids, deep venous thrombosis, pulmonary embolism.

Case Report:

29 years old female, 9 months of gestation, G2P1L1, known case of chronic ITP diagnosed 2 years back with past history of DVT managed with oral anticoagulant in 2020 presented with c/o lower limb petechiae and bleeding gums. Platelet count was 2000 and IPF was 29%.ANA came weakly positive (1:100 titer, speckled pattern) but ANA blot was negative. Fever profile, HIV, HBsAg, Antiphospholipid antibodies, ADAMTS 13 was negative. Renal function test and liver function test was within normal limits. Peripheral smear suggestive of severe thrombocytopenia with giant platelets. Patient received methyl prednisolone steroid pulse therapy for three days followed by oral steroids after ruling out secondary causes of ITP along with platelet transfusion.

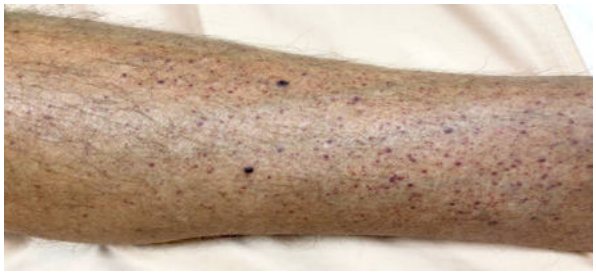

Figure 1: Petechia over lower limb

On day five of steroid therapy, patient developed swelling of lower limbs, pain while walking and breathlessness. Ultrasound doppler of lower limb showed deep venous thrombosis of bilateral lower limb and CT pulmonary angiography showed pulmonary thromboembolism. As ANA came positive (efficacy of NOAC not known) and patient being in third trimester of pregnancy, low molecular weight heparin was initiated and dose titrated as per daily platelet count monitoring. IVC filter was inserted to prevent significant pulmonary thromboembolism during peripartum period. Emergency LSCS was done in view of ITP with DVT with scar tenderness in labor under Single Donor Platelet (SDP) coverage. IVC filter was removed at 4 weeks postpartum. Patient was advised to take injection low molecular weight heparin as per weekly platelet monitoring.

INJ LMWH 0.6 cc SC twice daily if PLT count is > 50,000 or INJ LMWH 0.6 cc SC once daily if PLT count is 30,000 to 50,000 and withhold anticoagulation if patient has any bleeding tendencies or platelet count less than 30,000.

PLATELET COUNT AND/OR CLINICAL FEATURES	DOSE OF INJ LOW MOLECULAR WEIGHT HEPARIN(LMWH)
>50,000	0.6 cc s/c twice daily
30,000 – 50,000	0.6 cc s/c once daily
<30,000 or any bleeding manifestation	Stop LMWH and consult physician

Oral steroid was gradually tapered and stopped over 6 weeks. Patient was followed up every month and repeat ultrasound doppler of lower limb done at 6 months postpartum showed complete resolution. Patient is asymptomatic with current platelet count of more than one lakh.

DISCUSSION:

ITP is an acquired autoimmune disorder characterized by a low platelet count resulting from platelet destruction and impaired platelet production. The incidence of ITP is estimated to be 2 to 5 per 100 000 persons in the general population[3].

The clinical course of ITP may also be different depending on whether it is primary ITP (not associated with any other conditions), is the manifestation of a primary immunodeficiency or is associated with an underlying autoimmune condition or infection (secondary ITP). In the latter, the treatment of ITP is often directed at management of the underlying condition[4].

Chronic ITP is defined as ITP persisting for more than 12 months.[1] Therapy options for chronic ITP include glucocorticoids, splenectomy, rituximab, immunosuppression, thrombopoietin analogues (Eltrombopag and Romiplostim), and most recently, fostamatinib[5].

CONCLUSION:

Antithrombotic therapy in ITP is challenging as there are no recommendation for a minimum PLT count threshold. Some authors recommend that in patients with ITP and thrombosis, anticoagulation should be considered despite low PLT counts (< 20000) associated with significant risk of bleeding. Management of thrombotic events in ITP should be individualized taking into account of personal risk factors, risk of bleeding and severity of thrombosis

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