Unusual Presentation of Chronic Idiopathic Thrombocytopenic Purpura

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Abstract :

A snakebite victim presented with normal clotting profile and a low platelet count. A routine CBC in his past records (February 2004) showed a platelet count of 20,000/microlitre, but the patient was not symptomatic. We report a case of chronic idiopathic thrombocytopenic purpura, incidentally found in a patient presenting with snakebite. The patient also has acquired primary testicular failure. After the diagnosis the patient was on regular follow up. He caused trauma to the right external auditory canal and perforated his tympanic membrane. His left tympanic membrane was also scarred and retracted. Establishing a diagnosis of an ITP early is important so that the patient can take precaution to avoid undue trauma and monitor proper follow up.

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Case Report

A fifty eight year old male Omani presented with snakebite in the right dorsum of foot and after admission to the hospital was found to have multiple petechiae and superficial ecchymoses. He had no history of vomiting, swelling of feet, hemetemsis or melena. Examination revealed a moderately built male with blood pressure reading of 140/ 70 mmHg. There was no pallor or organomegaly. His initial investigation showed hemoglobin of 12.1 gm/dl, WBC count of 7,800 /microlitre and platelet count of 26,000 /microlitre. Peripheral smear showed decreased platelets and few giant platelets. His blood sugar, liver function and renal function tests were normal. His prothrombin time, activated partial thromboplastin time and fibrinogen levels were within normal limits.

Coombs test, antinuclear antibody, Lupus anticoagulant, antiphospholipid antibody screen, HIV I & II, and free auto and allo immune antiplatelet antibodies were negative. Hormone profile showed raised serum FSH (33.6 IU/L) & LH (15 IU/L) with decreased testosterone level (5.2nmol/L). Serum TSH, Prolactin and Cortisol levels were within normal limits. Bone marrow examination showed a normoblastic marrow and adequate megakaryocytes with focal crowding of megakaryocytes. There were no dysplastic features. Hence, a diagnosis of chronic idiopathic thromobocytopenia was made and was on regular follow up.

Past medical history reveals that the patient had mumps and orchitis. He is married and has one child. There was history of erectyl dysfunction for 2 months. On examination his testes was small and firm. Serum FSH, LH was elevated and testosterone was decreased. Follow up visits showed platelet count of 41,000/ microlitre. He came to ENT clinic with history of trauma to right ear while cleaning by a small piece of wood and subsequently caused perforation of the tympanic membrane. His left tympanic membrane was also scarred and retracted.

Discussion

Platelets are anucleate cytoplasmic fragments of megakaryocyte and circulate for 8 – 10 days in the peripheral circulation. The normal platelet count in adult is 150,000 to 450,000 / microlitre. Idiopathic thrombocytopenic purpura (ITP) refers to thrombocytopenia in which apparent exogenous factors are lacking and in which disease known to be associated with secondary thrompbocytopenia is lacking.[1] In chronic ITP the median age is usually 40 – 45 years although in one large series of patients, 74% of 934 cases were younger than age 40 years (range 16 to 87 years).[2] The ratio of females to males in chronic ITP is 3:1. There is now convincing evidence that the syndrome of ITP is caused by platelet – specific autoantibodies that bind to autologous platelets, which are then removed by the phagocyte system via macrophage Fc receptors.[3,4] Platelet autoantibodies are either IgG or IgA with antigen specificity to the platelet glycoprotein IIb/IIIa or Ib / IX.[5] Anti cardiolipin antibodies are also seen commonly but its significance in the pathogenesis is not known.

The onset of chronic ITP is insidious with long history of hemorrhagic symptoms of mild to moderate severity. Bleeding manifestation was seen once with petechial rashes and superficial ecchymoses in his lower half of the body. The bleeding in the external auditory canal was induced due to trauma with a small piece of wood.

The initial step in the evaluation of a thrombocytopenic patient is the examination of the peripheral blood smear to confirm the decreased platelet count. Thrombocytopenia may be produced artefactually by clumping of the platelets in the blood

sample caused by EDTA – platelet agglutinins or the platelets may be unavailable for counting because they are bound in rosette formation to the surface of white blood cells. The diagnosis of ITP is usually a diagnosis by exclusion based on demonstration or peripheral thrmbocytopenia with a history, physical examination and complete blood count that do not suggest another cause of thrmobocytopenia.[6,7] Hypertension and bradycardia may be signs of increased intracranial pressure of undiagnosed intracranial hemorrhage. Distant low amplitude heart sounds accompanied by jugular venous distension may be evidence of hemopericardium.

Studies from Italy and Japan indicate that many persons with ITP have Helicobacter pylori gastric infection and that eradication of Helicobacter pylori results in increased platelet count. But, studies from USA and Spain do not support such views. In chronic ITP if the platelet count is more than 50,000 / micro litre, treatment is not indicated. If the platelet count is 20,000 to 50,000 /micro litre, treatment is usually not indicated but monitor closely. If Platelet count is less than 20,000 / microlitre treatment is indicated with one or more of the following: – Prednisone 1mg/kg body weight, forms the first line of therapy. Intra venous immunoglobulins or anti D Rho immunoglubulins if the patient is Rhesus positive is considered as second line of treatment. Splenectomy is reserved for relapsing severe ITP. There is no role for prophylactic platelet transfusion except during the perioperative period.

Conclusion

This case is reported because of the unusual presentation in which the patient presented with snakebite and subsequently discovered to have chronic ITP. Since the prothrombin time and activated partial thromboplastin time was normal, the clinician thought there was a laboratory error. But, subsequent repeat tests also showed similar results. Although the low platelet count was missed two years ago, the patient did not suffer from any serious bleeding manifestation. The patient also has associated acquired primary testicular failure. But following the diagnosis, the patient induced trauma to right ear resulted in hemorrhage and perforation of tympanic membrane. His left tympanic membrane was already scarred and retracted. Hence, proper advice and follow up is essential in a case of ITP.

References

1. Waters AH. Autoimmune thrombocytopenia: clinical aspects. Semin Hematol 1992 Jan;29(1):18-25.
2. Cortelazzo S, Finazzi G, Buelli M, Molteni A, Viero P, Barbui T. High risk of severe bleeding in aged patients with chronic idiopathic thrombocytopenic purpura. Blood 1991 Jan;77(1):31-33.
3. Hashino S, Mori A, Suzuki S, Izumiyama K, Kahata K, Yonezumi M, et al. Platelet recovery in patients with idiopathic thrombocytopenic purpura after eradication of Helicobacter pylori. Int J Hematol 2003 Feb;77(2):188-191.
4. Houwerzijl EJ, Blom NR, van der Want JJ, Esselink MT, Koornstra JJ, Smit JW, et al. Ultrastructural study shows morphologic features of apoptosis and para-apoptosis in megakaryocytes from patients with idiopathic thrombocytopenic purpura. Blood 2004 Jan;103(2):500-506.
5. Chan H, Moore JC, Finch CN, Warkentin TE, Kelton JG. The IgG subclasses of platelet-associated autoantibodies directed against platelet glycoproteins IIb/IIIa in patients with idiopathic thrombocytopenic purpura. Br J Haematol 2003 Sep;122(5):818-824.
6. Karpatkin S. Autoimmune (idiopathic) thrombocytopenic purpura. Lancet 1997 May;349(9064):1531-1536.
7. Raife TJ, Olson JD, Lentz SR. Platelet antibody testing in idiopathic thrombocytopenic purpura. Blood 1997 Feb;89(3):1112-1114.