Immune Thrombocytopenic Purpura – A Case Report and Narrative Review

[1]Dr. Ilamurugan Ezhilarasi, [2] Mrs. N. Karthiga Priyadharshini, [3] Dr. Paramasivam Preethi, [4] Dr. K.R. Shakila, [5] Dr. Thalaimalai Saravanan

[1] Second year Post graduate Student, Department of Oral medicine and Radiology, Karpaga Vinayaga Institute of Dental Sciences.

[2] Assistant professor, Karpaga Vinayaga College of Nursing.

[3] Second year Post graduate Student, Department of Oral medicine and Radiology, Karpaga Vinayaga Institute of Dental Sciences.

[4] Professor, Department of Oral medicine and Radiology, Karpaga Vinayaga Institute of Dental Sciences.

[5] Professor, Department of Oral medicine and Radiology, Karpaga Vinayaga Institute of Dental Sciences

ABSTRACT

An autoimmune bleeding disorder, formerly known as idiopathic thrombocytopenic purpura is characterized by isolated thrombocytopenia which is not associated with any other systemic illnesses. ITP in adults is often considered an autoimmune disorder, as the body produces antibodies that damage some of its own products-in this case, blood platelets. It is recorded that immune thrombocytopenic purpura occurs in about 2 in 1,00,000 adults. The course is more chronic although spontaneous remission can also occur within months of initial diagnosis. A thorough and timely workup of thrombocytopenia is necessary as it is considered as a diagnosis of exclusion to rule out other differentials of ITP. The present case report emphazises a case report and review on immune thrombocytopenic purpura in a 24- years old female who was identified in our hospital who visited for a normal dental screening.

KEYWORDS: Purpura, Idiopathic thrombocytopenic purpura, Immune thrombocytopenic purpura, Blood platelets

INTRODUCTION

The acronym ITP stands for 'immune thrombocytopenia' and which has, by international agreement, replaced the term 'idiopathic thrombocytopenic purpura'. This is done as because the cause of ITP is no longer idiopathic. The Greek term 'ídios pathos' refers to a disease without a tangible cause, but today we know that ITP is caused by a dysregulation of the immune system [1]. It is a bleeding disorder which is characterized by the presence of isolated thrombocytopenia (platelet count <150,000 u/L), with no associated systemic illness. ITP is reported in approximately 2 per 100,000 adults with a mean age of diagnosis of 50 years [2]. It is more common in females of childbearing age and in pregnancy. Although the pathogenesis is still unclear, ITP is believed to result from the development of an immunoglobulin G autoantibody targeting structural platelet membrane glycoproteins IIb-IIIa [3]. This renders platelets susceptible to phagocytosis by splenic macrophages and Kupffer cells in the liver. These autoantibodies are detected in 40–60% of individuals. Thus, other mechanisms including impaired production of the glycoprotein hormone thrombopoeitin, a stimulant for platelet production as well as triggers such as childhood exposure to viruses, helicobacter pylori infection and pregnancy are thought to contribute to ITP. Diagnosis mainly relays on history, physical and clinical examinations, laboratory investigations such as complete blood count. This case report presents the findings of a patient with immune thrombocytopenic purpura.

CASE REPORT

A 24 years old female patient came to our hospital

with a chief complaint of bleeding gums for the past 2 months. Patient encountered bleeding of gums spontaneously and also while brushing. History of presenting illness revealed that the patient had spontaneous bleeding of gingival areas and did not have any pain or sensitivity in any area. Patient did not have any history of fever or associated symptoms. Past medical and dental histories did not reveal any abnormalities. All routine investigations were done. The medical, social and family histories were remarkable. On general examination, patient was calm, co-operative, well oriented to time and place, well built and nourished. On extra oral examination, erythematous pin point spots were spot on the upper limbs and shoulder for the past 3 months [Figure 1]. No other abnormalities such as facial asymmetry was seen. On intra oral examination, hard tissue examination revealed minimal calculus was seen on the lingual aspects of mandibular incisors. Soft tissue examination revealed presence of petechial spots on the right and left buccal mucosa, hard palate and soft palate [Figure 2,3]. Based on clinical examination, a

provisional diagnosis of idiopathic thrombocytopenic purpura was given. Differential diagnosis included dengue, thrombotic thrombocytopenic purpura, hemolytic uremic syndrome, disseminated intravascular coagulation were given. On laboratory investigations, complete blood count was done which revealed a decreased platelet count of less than 10,000 cells/cumm, bleeding and clotting time within normal limits, haemoglobin was 12.6% and INR was 1.1. Co relating the laboratory investigations with the clinical examination and provisional diagnosis, a final diagnosis of Immune Thrombocytopenic purpura was given.

The patient was then shifted to Department of General Medicine and further management with platelet transfusion was done.

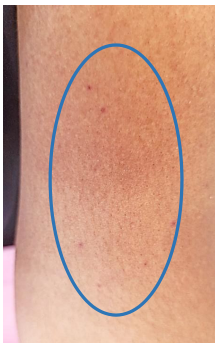

Figure 1: Extra oral examination

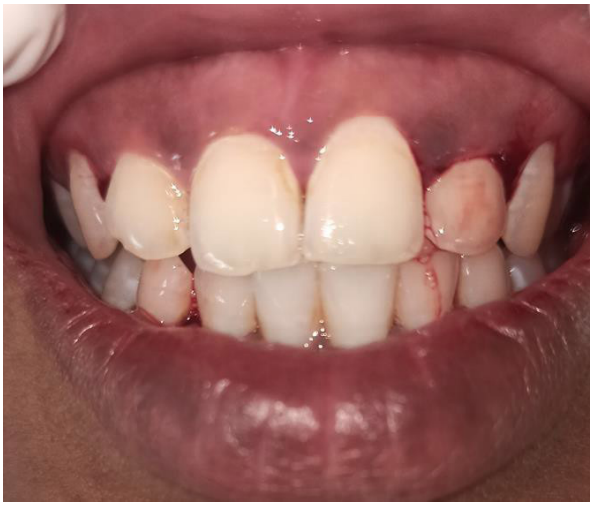

Figure 2: Intra oral examination - Bleeding of gingiva

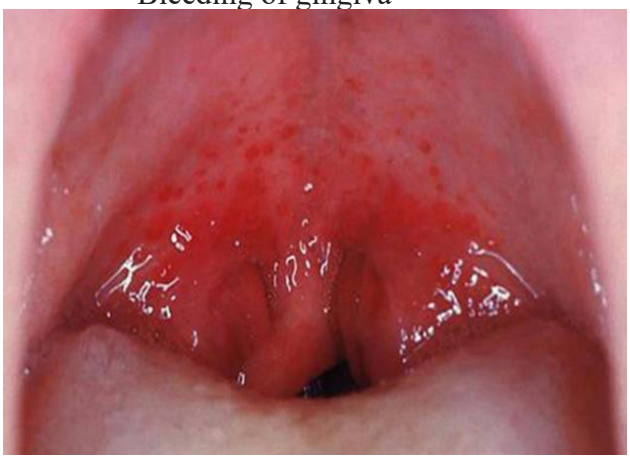

Figure 3: Intra oral examination – Petechial spots on hard and soft palate

DISCUSSION

ITP is a common condition which can have serious complications if not treated early and promptly. It is an autoimmune syndrome in which the blood platelets are coated with autoantibodies to the antigens present on the platelet membrane, resulting in splenic sequestration and phagocytosis by mononuclear macrophages. It results in shortened life span of platelets in the circulation, along with incomplete and poor compensation by increased platelet production by bone marrow megakaryocytes, results in a decreased number of circulating platelets. ITP is not hereditary but an acquired form of thrombocytopenia and needs to be distinguished from congenital thrombocytopenias, which are much rarer. The incidence of ITP in adults is between 0.2 and 0.4 new cases per 10,000 per year [4] and the prevalence is 0.9-2.6 per 10,000. In children and adolescents, the ITP incidence is 0.2-0.7 new cases per 10,000 per year [5] and the prevalence is 0.4-0.5 per 10,000 [6]. The prevalence is significantly lower in children

than in adults because pediatric ITP rarely becomes chronic (see the section 'Prognosis and Risk'). In pediatric ITP, boys are more often affected than girls. In middle age, women are more likely to develop ITP than men. After age 60 years, men predominate again.

The clinical manifestations of ITP are highly variable and range from a fairly common asymptomatic patient with mild bruising, mucosal bleeding to spontaneous or frank bleeding from any site. Overall, symptomatic bleeding is uncommon unless the patient has severe ITP

(platelet count <30,000/μL). Investigations in patients primarily focuses on excluding the other conditions that might cause nonimmune thrombocytopenia [7]. Acute thrombocytopenia with other signs of neurological manifestations may indicate a diagnosis of thrombotic ITP. Presence of lymphadenopathy and splenomegaly along with ITP in patients may suggest the presence of a lymphoproliferative disorder. On the management of patients who are considered for splenectomy, Bone marrow examination is recommended, if the patients are older than 60 years. Management of ITP varies according to the platelet counts.

When platelet counts drop below 30,000/uL in patients without any active bleeding, clinical observation is normally indicated. Patients with active bleeding are treated with the treatment of choice. The first line therapy usually involves glucocorticoids such as Prednisone 1 mg/kg po once/day with a slow taper in dose [8]. Second line therapies include monoclonal antibodies such as Rituximab or in refractory cases, thrombopoietin-like agents such as Eltrombobag (25–75 mg once/day) or Romiplostom (1–10 mcg/kg once/week) are indicated. Splenectomy can achieve complete remission in two thirds of patients however can increase the risk of thrombosis and infection with encapsulated bacteria. Intravenous immunoglobulin (IVIG) and anti D immunoglobulin (IG) are indicated for glucocorticoid-resistant ITP or for management of severe bleeding. In patients who are actively bleeding or in those with platelet counts <10,000 rapid phagocytic blockade through IVIG (1 g/kg once/day for 1–2 days) is attempted and IV anti-D IG in Rhesus positive patients can be tried [9]. In life threatening bleeds, platelet transfusion can also be initiated which otherwise would normally be ineffective due to rapid consumption.

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