Case Report

A YOUNG GIRL WITH ACUTE IMMUNE THROMBOCYTOPENIC PURPURA

Ayesha Choudhry[1], Arsham Najeeb[2], Ehsan ul Haq[3], Agha Shabbir Ali[4]

ABSTRACT

Immune thrombocytopenic purpura (ITP) is an immune-mediated acquired disease in both adults and children. It is characterized by transient or persistent reductions in the platelet count. We report a case of ITP presenting with intermittent fever and oral hemorrhagic symptoms. The patient was a 9-year-old girl with no significant past medical history. She presented with sudden onset gum bleeding and hemorrhagic bullae on the buccal mucosa. During night pinpoint purple spots (petechiae) appeared on the body mainly, on the lower legs. Laboratory tests revealed severe thrombocytopenia with a platelet count as low as 2000/mm^3. Under a provisional diagnosis of Acute ITP, she was treated with 1 mega unit platelets transfusion and high dose immunoglobulin therapy. Her platelets rapidly increased, and no bleeding complications were reported.

Key Words: Gingiva, Hematoma, Hemorrhage, Purpura

INTRODUCTION

Immune thrombocytopenic purpura (ITP) is an acquired hematological disorder that is developed due to the production of auto-antibodies against platelets leading to isolated thrombocytopenia, in the absence of other causes of thrombocytopenia such as drugs, infections, malignancy, or other autoimmune diseases.[1,2] ITP commonly affects children between one and seven years of age. Parents are much concerned as they see such symptoms first time in children. Severe life-threatening bleeding is rare (0.2–0.9%).[3,4] Childhood Primary ITP usually is a self-limiting disease, with or without treatment. Complete remission occurs within six months after diagnosis, commonly within 6–12 weeks, in the majority of children. However, 20–30% of children continue to have persistent low platelets count with bleeding symptoms beyond six months after the diagnosis.[5,6]

CASE DESCRIPTION

A 9 years old girl student of 3rd class with no significant past medical history presented in peads emergency at Farooq Hospital on 21-08-2020 with a history of intermittent fever, gingival bleeding for the last 4 days. She was taking some oral medicine from a local practitioner for this complaint without any improvement. A day before admission, she developed pinpoint reddish-purple spots (petechiae) more marked on the lower limbs (Figure-1) and face, than gum bleeding was persistent and was getting difficult to control thus she was brought to peads emergency. There was no personal history of any bleeding problem and her family history was also insignificant regarding bleeding disorder or coagulopathy.

Examination at the time of admission revealed a young girl looking comfortable regardless of multiple new and old purpuric spots on the face and both lower limbs. She had stable vital signs including blood pressure. Her weight was 20kg with 130cm height and was at 25th centile for her age.

There was no lymphadenopathy and the liver, the spleen was not palpably enlarged. The rest of the systemic examination was unremarkable.

First Complete blood count revealed Hb 11.9 g/dl, TLC 5.3/mm^3, neutro 37%, lympho 54%, and severe thrombocytopenia with

[1-3]House Officers, Department of Pediatric Unit 2, Farooq Hospital, West Wood, Lahore.
[4]Professor Pediatric Unit 2, Farooq Hospital, West Wood, Lahore.

platelet count 2000/mm^3. X-ray chest and ultrasound abdomen were normal. Blood culture revealed no growth.

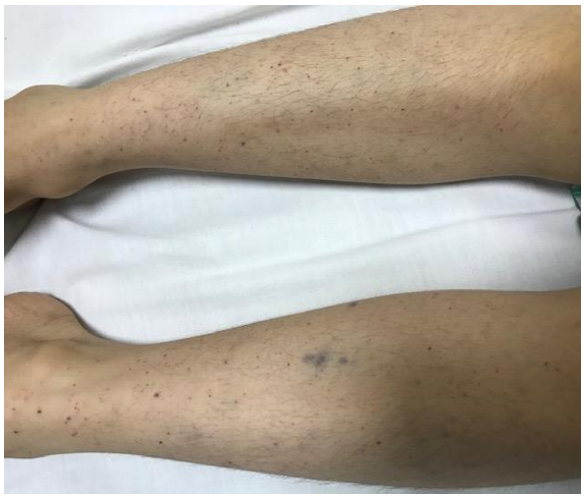

Figure-1. Multiple bruises and a petechial rash over both shins

Differential diagnoses before investigation included ITP, aplastic anemia, acute leukemia, and dengue fever. The dental consultant's opinion was also taken on this case. Considering the clinical behavior, absence of lymphadenopathy, and hepatosplenomegaly with isolated thrombocytopenia, the diagnosis of immune thrombocytopenia was made.

The patient was initially managed by IV fluids and oral antibiotics for stomatitis and gum bleeding. Due to the critically low platelet count (2000/mm^3), 1 Mega unit of platelets was transfused. We planned to treat her with immunoglobulin IgG as immunosuppressive therapy. The total dose decided was 2 gm/kg. Twenty (20) gm IVIG was given to the patient on two consecutive days. Clinically she improved within 24 hours as no new bruise or purpuric spot appeared after the first IgG infusion. CBC report showed improvement in platelet count within 48 hours.

The patient was discharged on the 4th day when she was clinically stable and platelet count was >70,000/mm^3. She was to follow up after 3 days.

After one week on her 1st follow-up, bruising completely disappeared and platelet count was 222,000/mm^3.

DISCUSSION

Idiopathic (autoimmune) thrombocytopenic purpura (ITP) is the most common cause of thrombocytopenia in children. Peak age is 2-5 years but also seen in adolescents as well as in younger children. The exact cause of ITP is not known but in 60-70% of babies, there is a history of viral throat infection 1-2 weeks ago. Antibodies produced against these viruses destroy host platelets as well. These antibodies fix on surfaces of platelets and make them easily sequestered in the spleen. Thus the number of platelets decreases (thrombocytopenia) which is responsible for the bleeding manifestation of the disease.[1]

Cutaneous bleeding i.e. bruises, purpura, and hematomas are common clinical manifestations in the baby with ITP who is otherwise well looking. Some children do have bleeding from mucous membranes e.g. epistaxis, gum bleeding, or hematuria. Fever, bone pains, lymphadenopathy, and splenomegaly are not regular features of ITP in children.[1,3]

ITP needs to be differentiated from other causes of thrombocytopenia in children e.g aplastic anemia, acute leukemia, and inherited platelet disorders. Most babies with these disorders are sick-looking and have lymph node enlargement or splenomegaly and bone pain.

In ITP CBC depicts reduced platelets count while red cell and white cell number is normal. Anti-platelets antibodies are on the rise but are generally not required in clinical practice. Bone marrow examination is subjected to the clinical experience of the pediatrician. If any clinical association creates doubt of an alternate diagnosis e.g acute leukemia then he may ask for bone marrow biopsy.

Two issues are important in the management of ITP in children

1. When to treat
2. Which drug to be used in the treatment of these babies.

ITP in children is a self-limiting disease in most of cases.[5] But the number of platelets count and site of bleeding decide the timing of treatment in many cases. Platelets count <

10,000/mm^3 and bleeding from mucous membranes e.g. nose, gums or hematuria also are indications of platelet transfusion. In this case, platelets dropped to 2000/mm^3 and she was bleeding from oral mucosa at the time of admission. It necessitated platelet transfusion and IgG therapy.

Steroids, immunoglobulins; IgG, IgD, and other Immunosuppressive drugs are used in acute ITP. These three drugs are taken as first-line drugs in the management of ITP. IgG is the first choice in children and steroids are used in adults. Oral prednisolone 2-4 mg/kg/day for 2 to 3 weeks is commonly used. This therapy is cheap and easy but needs bone marrow biopsy before therapy. IgG immunoglobulin is costly and associated with the risk of aseptic meningitis. In our case report platelets, the number started rising within 24 hours. Clinically, bleeding manifestations were improved within 48 hours.[2,3]

Anti D therapy is also cheap and of short duration but associated with risk of anemia. Thus Hb should be adequate before this therapy and is only carried out in positive blood group patients. Monitoring of response to therapy is carried out by the rise in platelets number. Complete response (CR) is platelets count more than 100,000/mm^3. Response (R) means a platelet count of 30,000/mm^3 on two occasions in a week. In both conditions, clinical symptoms should also disappear.[7,8]

AUTHOR'S CONTRIBUTION

AC: Collection of data
AN: Writing of article
EH: Critical review
ASA: Supervisor of the study

REFERENCES

1. Faki Osman ME. Childhood immune thrombocytopenia: clinical presentation and management. Sudan J Paediatr. 2012;12(1):27-39.
2. Jayaraman A, Das S, Biswal N, Dillikumar CG, Bade BA. High dose, short course prednisolone for acute idiopathic thrombocytopenic purpura (ITP) in children. Int J Contemp Pediatr. 2017 Sep;4(5):1705-8. doi:http://dx.doi.org/10.18203/2349-3291.ijcp20173770
3. Ahmed M, Martinez AY. Idiopathic Thrombocytopenic Purpura (ITP) Topic Review and Case Report. Int J Oral Craniofac Sci. 2017 Feb 8;3(1):008-11. doi: https://doi.org/10.17352/2455-4634.000024
4. Aytekin G, Yıldız E, Çölkesen F, Yılmaz S, Tekinalp A, Demircioğlu S, et al. Reverse Angle: Immunological Evaluation of Patients with Idiopathic Thrombocytopenic Purpura: A Retrospective Cohort Study. Asthma Allergy Immunol. 2019 Dec 28;17(3):152-9. doi: 10.21911/aai.500
5. Lambert MP, Gernsheimer TB. Clinical updates in adult immune thrombocytopenia. Blood. 2017 May 25;129(21):2829-35.
6. Neunert C, Terrell DR, Arnold DM, Buchanan G, Cines DB, Cooper N, et al. American Society of Hematology 2019 guidelines for immune thrombocytopenia. Blood Adv. 2019 Dec 10;3(23):3829-66. doi: 10.1182/ bloodadvances.2019000966.
7. Rodeghiero F, Stasi R, Gernsheimer T, et al. Standardization of terminology; definitions and outcome criteria in immune thrombocytopenic purpura of adults and children: report from an international working group. Blood. 2009 Mar 12; 113(11): 2386-93.
8. Heitink-Pollé KM, Uiterwaal CS, Porcelijn L, Tamminga RY, Smiers FJ, Van Woerden NL, et al. Intravenous immunoglobulin vs observation in childhood immune thrombocytopenia: a randomized controlled trial. Blood. 2018 Aug 30;132(9):883-91. doi: https://doi.org/10.1182/blood-2018-02-830844