International Journal of Research in Medical Sciences
Biyani G et al. Int J Res Med Sci. 2017 Apr;5(4):1697-1699
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20171290

Case Report

Recurrent immune thrombocytopenic purpura with excellent prognosis

Garima Biyani*, Syed Md Azad, Souvik Guha, Rohit Kapoor, Hriday De

Post Graduate Trainee, Institute of Child Health, Kolkata, West Bengal, India

Received: 06 February 2017
Accepted: 07 March 2017

***Correspondence:**
Dr. Garima Biyani,
E-mail: garimabiyani04@gmail.com

ABSTRACT

Immune thrombocytopenic purpura (ITP) is an autoimmune disorder characterized by increase in destruction of circulatory platelets and is the most common cause of acquired thrombocytopenia in childhood. ITP can be classified based on duration of thrombocytopenia as acute and chronic form. Recurrent ITP is defined as recurrence of symptoms, after at least three months of remission sustained without any treatment. It is a rare entity and seen in just 5% of all ITP cases. Further, its treatment is often cumbersome and warrants use of non-conventional drugs and splenectomy. Reported here is a case of ITP in a 10-year-old girl, who presented with three recurrences and all episodes were successfully treated with either oral Prednisolone or resolved spontaneously.

Keywords: Autoimmune disorder, Recurrent ITP

INTRODUCTION

Immune thrombocytopenic purpura (ITP) is an autoimmune disorder characterized by increase in destruction of circulatory platelets and is the most common cause of acquired thrombocytopenia in childhood.[1] ITP can be classified based on duration of thrombocytopenia as acute and chronic form. Recurrent ITP is defined as recurrence of symptoms, after at least three months of remission sustained without any treatment.[2] It is a rare entity and seen in just 5% of all ITP cases. Further, its treatment is often cumbersome and warrants use of non-conventional drugs and splenectomy.

CASE REPORT

A 10-year-old girl was admitted with complaints of petechial rashes over body for four days and bleeding from nose for two days. Nose bleed comprised only 5 to 6 drops of fresh blood and subsided spontaneously or with local pressure. There was no history of fever, pallor, joint pain and pain in abdomen. She had had no significant illnesses and no family history of any bleeding disorder. The girl was active and playful. On examination, petechiae and purpura were present over the chest, back, extremities, face and oral mucosa. There was no organomegaly and lymphadenopathy. His vitals were stable and blood pressure was within normal range for the age. On laboratory investigations hemoglobin was 11.9 gm%, total leukocyte count was 8200 cmm, with Polymorphs 60%, Lymphocytes 37% and Eosinophils 02%, erythrocyte sedimentation rate was 14 mm in 1st hour and platelet count was 8000/cmm. Renal function test, liver function test and coagulation profile were within normal limits. Antinuclear antibody and serology for HIV and hepatitis B surface antigen were negative.

On account of compatible history and supportive examination findings, a presumptive diagnosis of acute ITP was made. Bone marrow biopsy was done before starting treatment. Bone marrow examination revealed megakaryocytic thrombocytopenia without any other abnormality. Since the platelet count was less than 20000/cmm with mucosal bleed, the decision to treat the child was taken. Patient was started on tablet Prednisolone, in a dose of 2mg/kg/day for 7 days. Patient responded well to the treatment and her platelet count increased to 95,000/cmm after first week. Glucocorticoid

therapy was tapered over next 3 weeks and finally stopped. After 4 weeks of treatment, her platelet count reached 1,60,000/cmm.

She was not seen again for 10 months, when she presented in OPD with 4-5 petechial spots over chest, thigh and back (Figure 1 and 2) with 1 episode of epistaxis 1 week back. This time her platelet count was 30,000/cmm. She was only followed up weekly without any treatment and her platelet count increased to 1,10,000/cmm in 8 weeks.

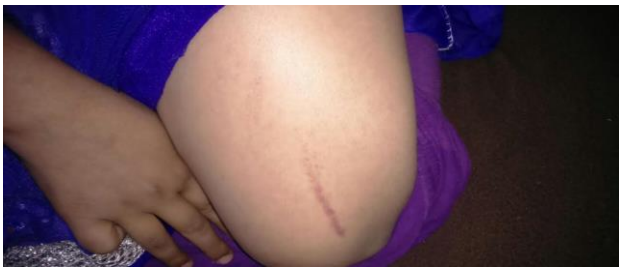

Figure 1: Ecchymotic spot over thigh.

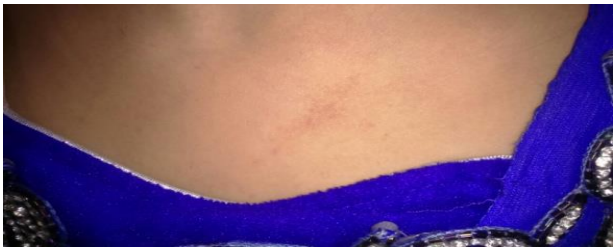

Figure 2: Petechial spots over back.

After 4 and 14 months following last episode, at the age of 11 years 2 months and 12 years respectively, patient again presented with epistaxis and petechial rashes over body and a diagnosis of ITP was made on similar ground both times. Every time patient was subjected to the same investigations (except ANA and HIV/HBs serology) and her platelet count was 20,000/cmm and 11,000/cmm respectively. Both the times, she was treated with Prednisolone (2mg/kg) for 1 week followed by tapering over next 21 days and had normal platelet counts after 4 weeks of treatment.

Now the patient is 14 years of age and is being followed up for the last two years. No further recurrence has been documented yet.

DISCUSSION

Immune thrombocytopenic purpura (ITP) is an autoimmune disorder characterized by increase in destruction of circulatory platelets and is the most common cause of acquired thrombocytopenia in childhood.[1] ITP can be classified based on patient age (adult or childhood ITP), and duration of thrombocytopenia (newly diagnosed ITP or chronic).[2] Newly diagnosed ITP is designation given for the 1st 3 months after diagnosis. Chronic ITP by definition persists for more than 12 months. In between acute and chronic category there is another entity called Persistent ITP, which is defined as ITP with low platelet count persisting for 3 to 12 months.[3] The annual incidence of ITP is estimated to be 5 cases per 100,000 children.[4]

In ITP, anti-platelet immunoglobulins bind to platelet membranes leading to premature death. The antibody coated platelets are rapidly removed by spleen and liver macrophages. Pediatric haematologists frankly don't know how to classify children with so called "recurrent" ITP. For practical reasons, the most acceptable definition is the recurrence of ITP after at least 3 months of remission sustained without any treatment.[5] An estimated 5% of paediatric patients have recurrent episodes of thrombocytopenia followed by variable periods of remission.[5] Although in some reviews recurrent ITP is included as a separate form, distinct from acute and chronic ITP, this is probably incorrect. Recurrent ITP is believed by most pediatric haematologists to be an exacerbation during the course of a compensated chronic ITP.[6] During periods of remission, increased platelet production balances the increased rate of platelet destruction, but during exacerbations, platelet production by the marrow is suppressed by viral infections or other factors and is unable to offset the rate of destruction.[6] Clinical and laboratory features did not differ from acute ITP (young age, preceding viral infection, abrupt onset of symptoms, low platelet count).

Treatments options available for recurrent ITP patients, not responding to glucocorticoids range from rituximab, azathioprine, cyclophosphamide, danazol, vinca alkaloids, ascorbic acid, colchicine, interferon-α, combination chemotherapy, protein A immunoadsorption, cyclosporine, ε-aminocaproic acid, plasma exchange to recently approved thrombopoietin receptor agonists.[7] Splenectomy is considered in patients who do not respond to these drugs. Corticosteroids were the mainstay of treatment in our patient. The main mechanism of action has been hypothesized as decrease autoantibody production, improve integrity of leaking capillaries and increase impaired clearance of antibody-coated platelets by mononuclear macrophages.[8-10]

Although recurrent episodes of ITP have been previously reported in literature, but these patients often don't respond to first line immunosuppressive therapy. Though in our case, all the recurrences were successfully treated with either oral Prednisolone or resolved spontaneously. The recurrence of thrombocytopenia did not influence an excellent prognosis in our patient.

CONCLUSION

In conclusion, ITP can present as multiple episodes of recurrences in children and can be treated satisfactorily with Prednisolone and a trial of Prednisolone should be given before resorting to other costly treatments.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Guerrero AP, Musgrave JE, Lee EK. Immune globulin-responsive thrombocytopenia in acute post-streptococcal glomerulonephritis: report of a case in Hawai'i. Hawaii Med J. 2009;68:56-8.
2. Stasi R, Evangelista ML, Stipa E, Buccisano F, Venditti A. Idiopathic thrombocytopenic purpura: current concepts in pathophysiology and management. Thromb Haemost. 2008;99:4-13.
3. Rodeghiero F, Stasi R, Gernsheimer T, Michel M, Provan D. Standardization of terminology, definitions and outcome criteria in immune thrombocytopenic purpura of adults and children: report from an international working group. Blood. 2009;113:2386-93.
4. Fogarty PF, Segal JB. The epidemiology of immune thrombocytopenic purpura. Curr Opin Hematol. 2007;14:515-9.
5. Choi SI, McClure PD. Idiopathic thrombocytopenic purpura in childhood. Can Med Assoc J. 1967;97:562-8.
6. Wilson DB. Acquired platelet defects. In: Orkin SH, Ginsburg D, Nathan DG, Look AT, Fisher DE, Lux S, eds. Nathan and Oski's Hematology of Infancy and Childhood, 7th ed. Elsevier, Philadelphia; 2008;1533-90.
7. Blanchette V, Bolton-Maggs P. Childhood immune thrombocytopenic purpura: diagnosis and management. Hematol Oncol Clin North Am. 2010;24:249-73.
8. Fujisawa K, Tani P, Piro L, McMillan R. The effect of therapy on platelet associated autoantibody in chronic immune thrombocytopenic purpura. Blood. 1993;81:2872-7.
9. Kitchens CS, Pendergast JF. Human thrombocytopenia is associated with structural abnormalities of the endothelium that are ameliorated by gluco corticosteroid administration. Blood. 1986;67:203-6.
10. Handin RI, Stossel TP. (1978) Effect of corticosteroid therapy on the phagocytosis of antibody-coated platelets by human leukocytes. Blood. 1978;51:771-9.

Cite this article as: Biyani G, Azad SM, Guha S, Kapoor R, De H. Recurrent immune thrombocytopenic purpura with excellent prognosis. Int J Res Med Sci 2017;5:1697-9.