

Clinical feature and management of immune thrombocytopenic purpura in a tertiary hospital in Northwest Nigeria

Abdulaziz Hassan, Adeshola Adebayo, Abubakar Umar Musa¹, Aishatu Maude Suleiman, Ismaila Nda Ibrahim, Ibrahim Usman Kusfa, Mohammed Sirajo Aminu

Department of Haematology, Ahmadu Bello University Teaching Hospital, Zaria, ¹Department of Haematology, Usmanu Danfodio University, Sokoto, Nigeria

ABSTRACT

Background: Immune thrombocytopenic purpura (ITP) is a rare bleeding disorder that may remit spontaneously. Life-threatening bleeding may require transfusion support, steroids, and other immunosuppressive therapy or splenectomy. **Objective:** To review the clinical presentation and laboratory features of ITP at Ahmadu Bello University Teaching Hospital (ABUTH), Zaria, Nigeria. **Subjects and Methods:** A retrospective analytic study of case notes and bone marrow (BM) records of patients diagnosed with ITP at Haematology Department, ABUTH, Zaria, from January 1, 2004, to December 31, 2012. **Results:** There were nine cases (six females, three males), aged 6–20 (mean 11.11) years. The presentations were epistaxis 8 (88.9%), purpura 4 (44.4%), gum bleeding 4 (44.4%), menorrhagia 2 (22.2%), and intracranial hemorrhage (ICH) 1 (11.1%). Only 1 (11.1%) had clinical splenomegaly. Platelet count of $<20 \times 10^9/L$ was found in 4 (44.4%) while 6 (66.7%) had packed cell volume of $<25\%$. All the nine cases had BM megakaryocytic hyperplasia. Six patients had blood transfusion support while 7 (77.8%) patients received oral prednisolone therapy with time to cessation of bleeding of 12–16 (mean of 8) weeks. One case had spontaneous remission while another had anti-D due to relapse after steroid therapy; this resulted in transient rise in platelet counts. None had other immunosuppressive therapy or splenectomy. Six (66.7%) cases were lost to follow-up after achieving remission and one died of ICH. **Conclusion:** ITP is not common in our center though its clinical presentations are varied. However, prednisolone and blood transfusion therapy are central to the management of these patients with favorable outcome.

Key words: Immune thrombocytopenia, Nigeria, purpura, Zaria

Address for correspondence:

Dr. Abdulaziz Hassan,
Department of Haematology,
Ahmadu Bello University
Teaching Hospital, Zaria, Nigeria.
E-mail: hassanyola@yahoo.com

INTRODUCTION

The platelet disorders are probably the most common cause of abnormal bleeding in patients. Platelet disorders may cause bleeding as a result of inadequate numbers (thrombocytopenia) or abnormal platelet function (thrombocytopathy). Bleeding resulting from platelet disorders is characterized by bleeding into the skin and mucosal membranes and prolonged bleeding, following trauma.¹

Immune thrombocytopenic purpura (ITP) also referred to as “acute” or “idiopathic” thrombocytopenic purpura is a bleeding disorder that results from peripheral autoimmune destruction of platelets.² It may also be defined as isolated thrombocytopenia with no clinically apparent associated conditions or other cause of thrombocytopenia.³ The mechanism underlying ITP is development of anti-platelet autoantibodies that

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leads to the destruction of platelets without adequate compensatory production of platelets.^{2,3}

ITP is characterized by thrombocytopenia (platelet count $<100 \times 10^9/L$), shortened platelet survival, presence of anti-platelet antibodies in plasma, and increased megakaryocytes in the bone marrow (BM). Presenting clinical features are spontaneous mild purpura with rarely severe bleeding in $<10\%$ of patients.^{4,5} The true incidence of ITP is unknown. However, in the USA, incidence was put at 1/10,000 in children and 5.5/100,000 populations in adults, while the male to female ratio is variable as there may be male preponderance in children and female preponderance in adults.^{4,5} Incidence of ITP in Nigerian population has not been documented, but a study in Southwest Nigeria reported an incidence of 0.005% of all hospital admissions.

The management of ITP in both children and adults involves several options including observation of nonbleeding patients, use of corticosteroids such as oral prednisolone or dexamethasone in mild cases, and use of intravenous immunoglobulin G, intravenous anti-D, transfusion support, and cytotoxic drugs in severe and recurrent cases. Recently, rituximab and dapsone have also been used while surgical therapy may be used in patients with intracranial hemorrhage (ICH) or children who may require splenectomy.^{6,7}

This study reviewed and analyzed the clinical presentation, laboratory findings, and management options and outcomes of ITP cases seen at the Ahmadu Bello University Teaching Hospital (ABUTH), Zaria, over a 10-year period.

SUBJECTS AND METHODS

This was a retrospective study involving the review of patients' case notes and BM register of all ITP cases seen at the Haematology Clinic and Haematology Day Centre of ABUTH from January 1, 2004, to December 31, 2013. Ethical clearance was obtained from the Health Research Ethical Committee of ABUTH.

Data on gender, age, presenting signs and symptoms, packed cell volume (PCV), platelet count, and BM aspiration finding as well as blood transfusion and drug treatment were extracted and analyzed using Microsoft Excel and Primer of McGraw-Hill. Descriptive statistics and *t*-test were used for data analysis with level of significance set at $P \leq 0.05$.

RESULTS

There were a total of nine cases (three males and six females) giving a male to female ratio being 1:2. The age range of the patients was from 6 to 20 years with a mean and SD of 11.1 ± 4.9 and median of 12.0 years. Eight (88.9%) patients presented with epistaxis, while other forms of presentations are depicted as given

in Table 1. At presentation, the overall mean, median, and range of platelet count were $27.3 \pm 15.2 \times 10^9/L$, $25 \times 10^9/L$, and $12-58 \times 10^9/L$, respectively, while those of PCV were $25.7 \pm 9.5\%$, 24%, and 15-44%, respectively. The presenting PCV, Platelet Count and treatment modalities used for each case are presented in Table 2. There were no significant differences between the mean ages, platelet counts, and PCV of male and female patients; 12.7 ± 7.0 versus 10.3 ± 4.1 years ($P = 0.527$), 25.0 ± 16.5 versus $26.8 \pm 16.9 \times 10^9/L$ ($P = 0.884$), and 34.4 ± 10.5 versus $23.0 \pm 4.7\%$ ($P = 0.051$), respectively, as shown in Table 3.

Thrombocytopenia with platelet counts of $<25 \times 10^9/L$ was found in 5 (55.6%) of the patients. Similarly, 5 (55.6%) of the patients had PCV of $<25\%$. All the nine cases had megakaryocytic hyperplasia in the BM. All the patients received oral steroid (prednisolone) therapy, six (66.6%) patients had blood transfusion support, but only one had anti-D immunoglobulin following relapse after therapy with prednisolone. Table 2 further highlights on some of the laboratory features and treatment options of the patients.

The mean time to cessation of bleeding was 8 weeks (6-10 weeks). None of the patients had other immunosuppressive therapy or splenectomy. Six (66.7%) cases were lost to follow-up after achieving remission and 1 (11.1%) died of ICH while 2 (22.2%) are followed up at the clinic.

DISCUSSION

The finding of only nine cases over 10 years or 0.9 cases per annum indicates that ITP is uncommon in Zaria. Similarly, with a total of 405,880 new patients seen at ABUTH during the study period, an incidence of 0.002% for ITP cases was obtained. This finding is not surprising as reports from earlier studies by Salawu and Durosini (2001) in Ile-Ife, Southwest Nigeria, reported an almost similar low incidence of 0.005%. This further corroborates the fact that ITP is a rare disease worldwide.²

The median age of the patients in this study was 12 years in conformity with most reports that ITP is commoner in childhood. Furthermore, 8 (89.9%) of the patients in this study were <18 years of age. However, this younger age is at variance with a previous Nigerian study in Southwest Nigeria where the median was found to be 21 years.⁸ This may be attributed to the likely difference in the population genetics between Northern and Southern Nigeria. The number of females doubled that of the males in this study. This is similar to most reports that indicate a female preponderance in ITP cases, especially in the younger age groups, even though no specific pathophysiologic mechanism has been attributed to this predilection toward female gender. However, this gender disparity is said to disappear with increasing age of the patients.⁹

Table 1: Clinical presentation of patients with ITP in ABUTH 2004-2013

Patient's S/No	Clinical presentation of patients with ITP n=9					
	Epistaxis	Purpura	Gum bleeding	Menorrhagia	Intracranial haemorrhage	Clinical splenomegaly
1	+	+	-	-	-	-
2	+	-	-	-	-	-
3	+	-	-	-	-	-
4	+	-	+	+	-	-
5	+	-	-	-	-	-
6	+	-	+	-	-	-
7	+	+	-	-	-	-
8	+	+	+	-	-	-
9	-	+	+	+	+	+
Total (%)	8 (88.9%)	4 (44.4%)	4 (44.4%)	2 (22.2%)	1 (11.1%)	1 (11.1%)

+: Indicates presence while, -: Indicates absence

Table 2: Laboratory parameters and treatment options for ITP cases in ABUTH Zaria, 2004-2013

SNo.	Age	Sex	PCV (%)	PLT(x10 ⁹ /L)	Treatment
1	8	Female	29	58	Pred, BT
2	6	Female	20	12	Pred, BT
3	6	Female	24	18	Pred
4	14	Female	15	31	Pred, BT and Anti-D
5	20	Male	44	44	Pred
6	12	Male	23	17	Pred, BT
7	13	Female	25	15	Pred, BT
8	6	Male	36	14	Pred
9	15	Female	24	27	Pred, BT

PCV: Packed Cell Volume, PLT: Platelet Count, Pred: Prednisolone, BT: Blood Transfusion, Anti-D: Anti-D immunoglobulin

Table 3: Comparison between Age, PCV and Platelets counts of male and female ITP cases

Variable	Sex		P
	Male	Female	
Mean Age±2SD	12.7 ± 7.0	10.3±4.1	0.527
Mean PCV±2SD	34.4±10.5	23.0±4.7%	0.051
Platelet x 10 ⁹ /L±2SD	25.0±16.5	26.8±16.9	0.884

SD: Standard deviation

Epistaxis was the most common form of presentation, followed by gum bleeding, while purpura and menorrhagia were rare. These presentations are in stark contrast to the purpuric bleeds that are common in Caucasian patients. Similarly, a Nigerian study reported gingival bleeding as the commonest presentation and purpura was rare. This may be explainable to the fact that our patients are black skinned and thus identifying early pinpoint petechial hemorrhages may be difficult.⁸

Eight (88.9%) of the patients had anemia at presentation and 5 (55.6%) had grade 4 thrombocytopenia (platelet count of <25 × 10⁹/L), but none had very severe thrombocytopenia or platelet count of <10 × 10⁹/L. The anemia may be explained by the bleeding episodes before presentation to hospital. On BM aspiration smears, all the patients had increased BM megakaryocytes which is

a cardinal finding in ITP and is considered reactionary to the peripheral thrombocytopenia.⁵

All patients were treated with oral steroids, but there was a high transfusion rate (67%) which may be indicative of the late presentation in our setting. However, only one patient had anti-D due to nonresponse to steroids, but none of the patients had splenectomy as a treatment modality. This could be due to the high rate of remission on oral prednisolone and the relatively high cost of the other modalities as most of our patients could not afford such treatment.

CONCLUSION

Immune Thrombocytopenic Purpura (ITP) is relatively uncommon in our centre. Majority of the cases achieved remission on oral steroids though there is a high rate of transfusion due to late presentation.

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Conflicts of interest

There are no conflicts of interest.

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