

CHILDHOOD IDIOPATHIC THROMBOCYTOPENIA: CLINICAL PROFILE AND MANAGEMENT

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ABSTRACT

Objective: To find out frequency of various clinical findings in children with idiopathic thrombocytopenic purpura and to evaluate the response of different management options.

Material and Methods: This study was conducted at Paediatric Department, LRH, Peshawar from 1st October 2003 to 30th September 2004 (One year). A total of 20 patients of ITP were encountered during this period and were evaluated. Our panel decided to use the American Society of Haematology definition for ITP, which reads ITP as "Isolated thrombocytopenia with no clinically apparent associated condition or other causes of thrombocytopenia". After taking detailed history and thorough examination, relevant investigations like peripheral smear, bleeding time, clotting time and coagulation screen were carried out in all cases. Bone marrow examination was done only in selected cases. Patients were given chance of spontaneous recovery except in cases with platelet count <20,000 and with heavy bleeding from any orifice. Such cases were treated with platelet transfusion and steroids.

Results: The study included 20 patients of whom 12 were males and 8 were females in the age range of 18 months to 14 years (mean age = 5.2 years). The commonest clinical symptoms and signs were bruises, epistaxis and gum bleeding. The commonest laboratory findings were Thrombocytopenia and prolonged Bleeding Time (in all cases). Bone marrow examination was done in three cases and was consistent with the diagnosis of ITP. Fifteen patients were offered chance for spontaneous recovery. Five patients received platelet transfusions and steroids. These were the cases who bled heavily and platelet counts less than 20,000. None of our patients had chronic ITP on their reviews so splenectomy was not performed in any of 20 cases.

Conclusion: The results of our study highlight the importance of wait and see policy and not to do bone marrow aspiration in every child with typical presentation of ITP as 90% of cases resolve spontaneously. Special smear to be done in all cases and treat the child rather than number of platelets.

Key Words: ITP, Platelets, Bone Marrow.

INTRODUCTION

Idiopathic thrombocytopenic purpura (ITP) is also known as immune thrombocytopaenic purpura and is the commonest cause of Thrombocytopenia in childhood¹. It results from an immune mediated destruction of circulating platelets within the reticuloendothelial system, mainly in the spleen. The reduced platelet count is accompanied by a compensatory increase in megakaryocytes within the bone marrow.

ITP is important cause of concern to parents as one-day child is well and all of the sudden develops bruises/epistaxis without any warning. It mainly affects children between 2

and 10 years of age with onset often 1-2 weeks after a viral infection. Affected children may develop purpura and superficial bruising and they may have epistaxis and other mucosal bleeding². The most common severe bleeding results from heavy nose bleeding but even these only occur in minority.

In about 90% of the children, the disease is acute and self-limiting³. Most will require only a brief stay in hospital to confirm the diagnosis and assess its severity. There has been much debate about the need to perform a bone marrow aspiration to exclude malignant infiltration or aplasia. If the clinical features are characteristic, with no abnormality in the blood other than a low

platelet count and no intension to treat, there is no need to examine the bone marrow⁴.

Treatment depends upon the condition of child with which he presents. If the child has minor purpura and platelet count are greater than 30,000 the child should be treated conservatively⁵. Children with platelet counts <20,000, significant mucous membrane bleeding and with moderate purpura should be treated with a specific regimens of glucocorticoids or intravenous immunoglobulins (IVIg)⁶. Children who have life-threatening bleeding should be hospitalized. They should receive conventional critical care measures along with treatment of ITP. Appropriate regimens included high dose parental glucocorticoids therapy, platelet transfusions and IVIg⁷.

This study was conducted to find out frequency of various clinical findings in children with ITP and to evaluate the response of different treatment options.

MATERIAL AND METHODS

This study was conducted at Department of Paediatrics, Postgraduate Medical Institute, Govt. Lady Reading Hospital, Peshawar Pakistan, over a period of one year from 1st of October 2003 to 30th September 2004.

A detailed proforma was made which comprised of all important information required to make the diagnosis of ITP. We looked into detail history of presentation, history of recent viral infection, family history of bleeding disorder, history of drug ingestion and bone pains etc.

After taking detail history physical examination was done to look for evidence of bleeding in the skin and mucous membranes. In clinical examination we specifically looked for lymph nodes enlargement, hepatosplenomegaly, weight loss and bony tenderness.

Investigations included peripheral smear, bleeding time, clotting time and coagulation screen in all cases. Bone marrow examination was done only in selected cases.

We defined ITP as "Isolated thrombocytopenia with no clinically apparent associated condition or other causes of thrombocytopenia" (definition of the American Society of Haematology).

Patients were given chance of spontaneous recovery except in cases with platelet count <20,000 and with heavy bleeding from any orifice. Such cases were treated with platelet transfusion and steroids.

INCLUSION CRITERIA

Any child with isolated thrombocytopenia with bleeding from any site.

EXCLUSION CRITERIA

1. Patient with aplastic anaemia
2. Patient with other causes of thrombocytopenia such as SLE, Wiskott Aldrich Syndrome.
3. Neonatal thrombocytopenia.

RESULTS

Department of Paediatrics, Lady Reading Hospital, Peshawar comprises of 110 beds. The total number of admissions during the study year was 7216 out of which twenty cases (0.38%) were diagnosed as ITP.

Male children were 12 in number while female patients were 8. The age range was between 18 months and 14 years with a mean age of (5.2 years) {table-1}. The commonest clinical findings were bruises in 08 cases (40%), anaemia, 5 cases (25%); epistaxis, 4 cases (20%) and gum bleeding in 3 cases(15%) {table-2}.

The history of preceding upper respiratory tract infection was recorded in 7 cases. Lymphadenopathy and organomegaly

Age and Gender-wise Distribution

Age	Patients No. (%Age)	Male	Female
18 months to 5 years	8 (40%)	5 (25%)	3 (15%)
> 5 years to 10 years	4 (20%)	3 (15%)	1 (5%)
> 10 years to 14 years	8 (40%)	4 (20%)	4 (20%)
Total	20 (100)	12 (60%)	08 (40%)

Table-1

Frequency of various Presentation

Signs & symptoms	No. of patients	%Age
Bruises	8	40
Anaemia	5	25
Epistaxis	4	20
Gum bleeding	3	15
Total	20	100

Table-2

Frequency of Laboratory Investigations on Arrival

Lab Findings	No. of Patients	(%) age
Platelet count	< 150,000	20
	< 20,000	4
Bleeding Time	Prolonged	20
Haemoglobin	< 9 Gm/dl	09
Bone marrow	Megakaryocytosis	03
Prolong PT/CT/APTT	Nil	0

Table-3

was not found in any case which are all consistent with diagnosis of ITP.

Laboratory findings were anaemia i.e. Hb% < 9 Gm/dl, (9 cases), Low Platelet count i.e. < 150,000/Cmm, (20 cases), Prolong BT (20 cases). CT, PT & APTT were normal in all cases. Bone marrow was performed in 3 rather atypical cases with a family history of aplastic anaemia. In two of these children the bone marrow was aspirated from the iliac crest (age above 2 years) while in one case (age 1 year), the specimen was taken from the medial tuberosity of tibia (table-3). The bone marrow examination revealed abundant megakaryocytes with normal budding in all the three cases. No abnormal cells were seen. These findings were consistent with the diagnosis of ITP.

Out of 20 patients, 15 were not treated with any form of treatment and they were observed for a day or two in hospital and were discharged home with advice of no contact sports and to see us in 2 weeks time. All these patients were reviewed at 2 weeks, 4 weeks and 8 weeks time. At each visit peripheral smear and platelet count was rechecked. All the patients showed a consistent rise of platelet count above 150,000/cmm, with no abnormality in the differential count.

Steroids were used in four patients; immunoglobulins were not used, as they were

expensive form of treatment. Platelet transfusion was given to five patients as they were bleeding heavily.

DISCUSSION

ITP is the commonest cause of thrombocytopenia¹. Although there is no reliable epidemiological data on the incidence of ITP, estimates are that 10-25/100,000 children and adults develop ITP each year⁸.

ITP affects more males than female children⁹. This fact is obvious in our study as the males outnumber females in a ratio of 3/2. The same results were encountered by Shahnaz I et al¹ where the predominant affected of this disease were males.

This disease is known to affect children in younger age group¹⁰. This fact is clearly highlighted in our study where children less than 5 years constitute 50 % of the cases while the age range of patients was between 18 months to 15 years. The same results are found by Hann IM where the major affected age range was between 2 to 5 years^{11,12}.

The commonest clinical presentation in our study was bruises and ecchymosis constituting a total of 12 cases. In contrast to our findings, in the series of Shanaz et al¹, all cases presented with easy bruisability and ecchymosis. Other major findings were

epistaxis, gum bleeding, haematemesis and malena.

A study conducted in Peshawar in 1995 reported petechiae and ecchymosis as initial presentation in 100% cases. Epistaxis, gum bleeding and bleeding per rectum were reported in eleven cases (34%), 3 cases (9%) and 2 cases (6%) respectively. There was no case documented with intracranial bleed¹².

For anaemia our cut of point was Hb% 9 Gm %. This was microcytic hypochromic anaemia which was also reported by Frank et al⁷. A total of 9 patients presented with Haemoglobin less than 9 Gm%. Platelet count was < 150,000/Cmm in all the cases and that is why bleeding time (BT) was prolonged in all these cases. Clotting time (CT) and coagulation profile was normal in all the cases. Study done by Frank⁷ also showed same results. Bone marrow was performed in 3 cases and results of all three were consistent with diagnosis of ITP.

Four patients had platelet counts <20,000/Cmm and all of them were treated with oral steroids for a period of four weeks after which all of them had responded to treatment and the platelet counts had returned to normal. Same response to treatment with steroids was shown by Arman^{13,14} in 1993.

Immunoglobulins and Anti-Rh (D) was not given to any patient as these modalities of treatment are quite expensive and beyond the buying capacity of our patients.

Study done by American Society of Haematology on both treated and untreated group were as that their best data on untreated disease come from two series in which about 75% of patients were not treated initially. Most patients had platelet counts <50,000 at presentation, and in one of the reports most had platelet counts <20,000 of the 221 untreated children, 2(0.9%) had fatal bleeding associated with the acute presentation, and 191 (87%) had a complete remission from ITP. The platelet count normalized in 2 to 8 weeks, with one half to two thirds of the patients recovering within 4 weeks^{15,16,17}.

So our study confirmed that to treat the child rather than number of platelets which correlates with international recommendations. Bone marrow should not be performed in every

case with typical presentation of ITP. In case of atypical presentation one should go ahead for bone marrow examination.

CONCLUSION

1. Petechiae and bruises are the commonest clinical findings of ITP.
2. Platelet counts above 20,000/cmm usually do not require any special treatment if asymptomatic.
3. Bone marrow examination for diagnosis is not required in straight forward cases of ITP.

RECOMMENDATIONS

1. ITP should not be over treated with platelets and transfusion.
2. Bone marrow should not be done in typical cases of ITP and investigations like mean platelet volume, anti nuclear antibody, platelet antigen specific antibodies and direct Anti-Globulin test are un-necessary.
3. Belt and brace treatment is only required in severe life threatening bleeding and one should treat the child rather than the number of platelets.

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