CASE REPORT

Splenectomy in Idiopathic Thrombocytopenic Purpura : A Clinical Experience

by

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Abstract

Since 1979 until 1987 there were 4 idiophatic thrombocytopenic purpura (ITP) cases who had undergone splenectomy, consisting of 2 males and 2 females. All patients had been treated with prednisone prior to splenectomy, 2 patients received additional cytostatics. The course of the disease prior to splenectomy had been followed in a period of time, varying from 2,5 to 8 years. The effect of treatment was not very statisfying, as both clinical and laboratory reccurrence often happened.

Post spelectomy, the administration of prednisone in 1 case was stopped immediately, in 2 cases it was stopped after 8 months and in 1 case it was continued. After more than 5 years follow up, 3 cases showed excellent clinical and laboratory findings, while 1 case failed.

Introduction

Idiopathic thrombocyopenic purpura (ITP) is a disorder of unknown etiology and pathogenesis. ITP in childhood is usually a brief, self limiting disease with an excellent prognosis for spontaneous and complete remission. Nevertheless 7 to 10 percent of these children fail to achieve sustained remission. (Carpenter et al., 1959; Ramos et al., 1978; Weinblatt and Ortega, 1982).

Platelet agglutination and complement fixing antibodies have been found to be present in a large proportion of patients suffering from ITP. The spleen may then be implicated in the genesis of the clinical entity, as a source of antibody production, the major sequestering agent for the sensitized platelets, or both. The spleen may

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also be implicated in the genesis of this disease by causing a production defect in platelets. This is evident by the frequent findings of immature megakaryocytes in the bone marrow.

This may, however, merely represent a "shift to the left" in response to increased destruction of platelets. It is very true that the spleen is the major sequestering site for platelets in this clinical entity (Block et al., 1966).

The treatment of ITP in childhood, especialy of chronic ITP, has been a matter of controversy in recent years (Wilde et al., 1967; Simon et al., 1975).

There are four modalities available for treatment of chronic ITP i.e.: splenectomy, corticosteroids, platelets transfusion and immunosupressive agents. Splenectomy is the oldest, firstly proposed and performed in 1916, and is still considered by many experts as the treatment of choice (Orringer et al., 1970; Bussel et al., 1983). The arguments in favor of splenectomy emphasize the high of permanent remission, with figures as high as 80 percent. (Orringer et al., 1970; Mc Millan, 1981), but according to others the permanent remission rate is only 50 to 60 percent (Wilde et al., 1967).

The purpose of this paper is to report the clinical experience of the efficacy of splenectomy for chronic ITP in our patients.

Report of Cases

In the past 8 years (from 1979) we had experienced only 4 cases of splenectomy in ITP, in the Child Health Department, Dr. Soetomo Hospital. The details of the four patients are recorded in table 1.

Table 1 : *Symptoms and signs*

	Case I	Case II	Case III	Case IV
Age :	7 years	11 years	3 years	3.5 years
Sex :	male	female	male	female
Symptoms :	epistaxis	gingival bleeding	petechiae	hematoma
Signs :				
- Ecchymosis :	+	+	—	+
- Petechiae :	+	—	+	+
- Liver & Spleen enlargment :	—	—	—	—
- Lymph node enlargment :	—	—	—	—
Other diseases :	—	—	—	—
Drug ingestion :	—	—	—	—

There were 2 males and 2 females, and their ages ranged from 3 years to 11 years. Epistaxis, bleeding of the gingiva, petechiae and ecchymosis or hematoma were the most common symptoms that occurred in our patients. On physical examination only ecchymosis (3 cases) and petechiae (3 cases) were found. Other signs and symptoms were not present.

The detail of the laboratory findings are recorded in table 2 and 3.

Table 2 : *Blood examination*

	Case I	Case II	Case III	Case IV
Hemoglobin (g/dl) :	10	11.1	10	10.7
Erythrocyte :	4110000	4340000	4600000	3120000
Leucocyte :	10.400	16.800	13.800	6.900
Platelet :	31.000	(—)	(—)	3.000
Diff. count				
Eosinophil :	—	—	2	7
Basophil :	—	—	—	—
Stab :	1	—	3	2
Segment :	65	40	55	23
Lymphocyte :	34	59	37	65
Monocyte :	—	1	3	3

No anemia was found in all cases. The platelet count was decreased and 2 patients had severe thrombocytopenia (platelets were not performed in the blood smears).

Table 3 : *Bone marrow examination and L.E. cell*

	Case I	Case II	Case III	Case IV
Bone marrow :				
- Cellularity :	normal	normal	hyper	normal
- Eryth. system :	good	moderate	increased	good
- Gran. system :	good	moderate	increased, Eo >	good, Eo >
- Megakar. system :	suffic.	suffic.	increased	increased
L.E. cell :	neg.	neg.	neg.	neg.

Bone marrow examination revealed normocellularity in 3 cases and hypercellularity in one. No abnormalities were found in the erythropoietic and granulopoietic systems. The megakaryocytes were sufficient in number in 2 cases and in the others they increased. L.E cells were not found in all cases (table 3).

Table 4 : *Treatment of cases before splenectomy*

	Case I	Case II	Case III	Case IV
Corticosteroid :	betameth.	betameth.	pred.	pred.
- duration :	2Yrs.5Mos	3Yrs.3Mos	3 Mos	10Yrs.3Mos
- response :	—	intermed	+	—
- relapse :			+ (after 8Mos)	
+ Immuno Suppressant :	imuran	imuran	(—)	cycloph/imuran
- duration :	1Yrs.3Mos	11 Mos		6Yrs.4Mos/2Yrs.4Mos
- response :	—	+		—
- relapse :		+ (after 5 Mos)		

Before splenectomy all patients received corticosteroid (betamethasone or prednisone) in a period of time, varying from 3 mos to 10 years 3 mos (table 4). No response were noted in 2 cases (case I and case IV), 1 patient had intermediate response, and only 1 patient had good response though then, after 8 months without corticosteroid administration the patient showed signs of relapse.

Only 1 patient had not received immuno suppressant (case IV) while the others had got immuno supressant administration varying from 11 months to 6 years. Response to immuno supressant was recorded only in one patient (case II) with good response though relapse then occurred after 5 months (table 4).

Table 5 : *Preoperation of splenectomy*

	Case I	Case II	Case III	Case IV
Period of diagnosis :	2Yrs.7Mos	4.5Yrs	9.5Yrs	9Yrs.7Mos
Treatment before splenectomy :				
- steroid :	+	+	+	+
- immuno suppr. :	+	+	—	+
Platelet count preop. :	5.000	40.000	1.000	1.000
Complication :	—	—	—	—
Infection (pre & post op.) :	—	—	—	—

The diagnosis of ITP was established after a period of time, varying from 2 years 7 months to 9 years 7 months. The platelet count before splenectomy was low especially in case III and IV. No infection or other complication were recorded before and after splenectomy (table 5).

Table 6 : *The course of the disease after splenectomy*

	Case I	Case II	Case III	Case IV
Corticosteroid :	stopped	continued (15 Mos)	continued (9 Mos)	continued
Immuno supressant :	stopped	continued (16 Mos)	—	continued
Response to splenectomy :	+	+	+	—
Period of platelet increase :	1 week	1 Mo	2 Mos	—
Relapse :	+ (after 4 Yrs)	+ (after 4 Yrs)	—	—
Reccurrent remission :	+	+		
Follow up :				
- period of time :	4 Yrs	8Yrs.4Mos	4Yrs.9Mos	4Yrs.10Mos
- clinic. appearence :	good	good	good	petechiaes

The course of the diseases post splenectomy had been followed up.
Corticosteroid was discontinued immediately, after 15 mos, and after 9 mos for case I, II, III, respectively, while in case IV corticosteroid was administered continously. Immuno supressive agent was administered continously only in case IV. Respons to splenectomy was recorded in case I, II, III with good responses and performed in time namely 1 week, 1 month, and 2 months respectively. But in case IV no response to splenectomy was noted.

In case I & II, relapse after 4 years occured, though followed by immediate reccurent remission with a good condition (table 6). The platelet count before and after splenectomy is recorded in table 7.

Table 7 : *Platelet count before and after splenectomy*

Case	Mean platelet count (X 1000) Before splenec.	After splenec.	Significancy : p (S/NS)
I	23 ± 6	163 ± 89	p : <0.001 (S)
II	63 ± 27.6	103 ± 39.7	p : <0.01 (S)
III	53 ± 44	168 ± 5.4	p : <0.001 (S)
IV	10 ± 11	7 ± 7.3	p : >0.05 (NS)

After splenectomy, mean platelet count increased significantly in cases I, II and III. In case IV the mean platelet count decreased, but not significantly different than before splenectomy.

Discussion

Chronic idiopathic thrombocytopenic purpura (ITP) is a syndrome of persistent thrombocytopenia secondary to an increased platelet destruction by the reticulo endothelial system, presumably caused by antiplatelet antibodies (Weinblatt and Ortega, 1982). According to Bussel et al. (1983), the syndrome at least lasts for 6 months, accompanied by an increase of megakaryocytes in the bone marrow and frequently by an elevated platelet associated immunoglobulin G (PAIgG). In our patients, thrombocytopenia had been recorded for more the 6 months.

The modalities of therapy for chronic thrombocytopenic purpura include the use of steroid, platelet transfusion or immunosuppressive agents and splenectomy. The results of steroid therapy vary and in reported series remissions range from 15 to 60 per cent. From the literature it appears that the percentage of remission at least in part, depend on the steroid dosage. Unfortunately, the adverse effect of longterm corticosteroid therapy represents the major argument against sustained usage of this drug.
The use of platelet transfusion seems to be of some benefit in a life threatening emergency, although evidence of their usefulness is sparse. A major sole cannot be ascribed to these transfusion for several reasons: (1) Heterologous platelets have a markedly shortened life span just as do autologous platelets. (2) The expense in preparing platelet concentration for use on a long term basis is prohibitive. (3) Patients receiving platelet transfusions quickly become sensitized and develop isoantibodies to the foreign platelets.
The use of immunosuppressive agent in the treatment of chronic ITP not yet achieved wide acceptance (Orringer et al., 1970). It therefore appears to us that splenectomy is the preferred treatment in chronic ITP, such as our cases. Some authors recommended splenectomy to be performed in patients with ITP that persists 6 to 12 months (Weinblatt and Ortega, 1982; Wilde et al., 1967; Ramos et al., 1978). In practice though it is recommended to operate after one year of illness (Schulman, 1964). According to Mc. Millan (1981), splenectomy should be postponed if possible until at least the age of six years, such as in our cases.

In patients with ITP that are refractory to corticosteroid, corticosteroid therapy should be continued before surgery, and in patients with a good response it should be tapered off rapidly and discontinued then after surgery (Mc. Millan, 1981). In our cases corticosteroid was continued in one patient, and discontinued in 3 patients (case I, II, III) immediately, after 15 Mos and 9 Mos post splenectomy, respectively.

Following splenectomy the platelet rise usually begins within the first 24 hours and it occasionaly will exceed one to two million percubic milimeter at the end of four to five days (Block et al., 1966).
According to Mc. Millan (1981), improvement may occur within hours and peaks achieved within two weeks. If relapse occurs it is usually note in the first six weeks. In our case improvement was

recorded in 3 cases (case I, II, III), and occurred after 1 week, 1 month and 2 months, respectively. Relapse occured until 4 years post splenectomy (case I, II) and afterwards reccurent remission occurred again. The improvement of platelet count in our cases was recorded significantly in 3 cases, but in one case the platelet count decreased post splenectomy though not significantly different.

Post operative (splenectomy) complication outlined, as subdiaphragmatic abscess, wound infection, septicemia, pneumonia, post operative hemorrhage and thrombocytosis (Wilde et al., 1967) were not at all found in our cases.

Conclusion

(1) Splenectomy is recommended in chronic ITP, especially when this disease is refractory to the medical approach.

(2) With close observation during splenectomy preparation, the post splenectomy complication could be prevented.

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