Splenectomy for immune thrombocytopenic purpura

F Parray, N Wani, K Wani, N Chowdri, R Wani, M Wani

Citation

Abstract
The therapeutic effects of splenectomy in 126 patients with various hematological disorders managed from 1982 to 2007 at Sheri Kashmir Institute of Medical Sciences, Srinagar (Jammu & Kashmir), India, were studied. Immune thrombocytopenic purpura (ITP) was the main indication for therapeutic splenectomy among all the hematological disorders i.e., 53 patients (42%). The ITP group is the main study group for this paper; mean age was 30 years (range 7-66), and the male to female ratio was 1.03:1. The mean platelet count in the pre-operative period was 30,650/mm$^3$ (range 4000-85,000). All patients presented with thrombocytopenia i.e., platelet count of <100,000/mm$^3$. In addition, 8 patients presented with anemia i.e., Hb <10g%. None of the patients presented with leucopenia. The morbidity observed was 15% and the mortality was 2%. The response to splenectomy was complete for thrombocytopenia in 4 patients and partial in 7 patients, whereas 4 patients failed to show any response. The prognosis was excellent in patients with platelet counts of >50,000/mm$^3$, age <50 years, no concomitant disease and disease of shorter duration.

INTRODUCTION
Pliny, in the first century, recorded that the ancients would remove the spleen from athletes to improve their “Wind” but this seems most unlikely. Galen and Nicholas Mathias are credited with the first total splenectomy for trauma in 1678. It was in the mid 19th century that therapeutic splenectomy was performed for splenic enlargement. Splenectomy for thrombocytopenia dates back to the initial demonstration by Kaznelson in 1916 who proved that removal of the organ would return the circulating platelet count to normal. However, for a short period, the introduction of steroids gave the hope to obviate the need for splenectomy in patients with immune thrombocytopenic purpura (ITP), but these hopes did not last long. The effects of steroids rarely showed persistence, whereas splenectomy was immediately and permanently curative.

The present study describes the effects of splenectomy on ITP, the complications encountered and the prognosis following splenectomy in a consecutive series of 53 patients operated at our hospital.

MATERIAL & METHODS
Medical records of all the retrospective cases were reviewed. Preoperative assessment included detailed hematological and coagulation studies when relevant. Blood was cross-matched for the operation and platelet transfusions were prepared and administered preoperatively in patients with symptomatic thrombocytopenia. Preoperative blood transfusions and fresh frozen plasma and corticosteroids (prednisolone) were given in patients whenever indicated. RhIG/IVIG could not be given in any patients because of the cost factor. All splenectomies were performed under general anesthesia. A full laparotomy was carried out for accessory spleens in all cases.

RESPONSE CRITERIA
We modified the criteria established by Musser et al. for describing the response in a splenectomized patient for simplicity of calculation, gave them the name of “Wani & Parray Criteria” and used them in all groups of hematological disorders to assess the response (Table 1).

Patients were put on intravenous fluids for 24-48 hrs. Steroids (hydrocortisone) were given in all patients and tapered for 48-72 hrs. in the post-operative period. A preset routine was followed for taking blood samples on day 0, 3, and 7 and after the 1st, 3rd, and 6th month for assessment of hematological parameters which mainly included Hb%, total leukocyte count (TLC) and the platelet count. The mean of these post-operative and follow-up readings was taken and compared with pre-operative parameters to assess the response as described in table 1. Penicillin prophylaxis was
started on the day of surgery one hour before surgery in 41 patients and continued for a period of 1-2 weeks in adults and elderly patients and for a period of 3-5 years in all patients <16 years of age. Pneumococcal vaccine could not be given in this group of patients i.e., before year 1995 (first 41 patients with ITP) because of non-availability. But the said vaccine was given in all other patients i.e., 12 patients of ITP from 1995 to 2007. Follow-up was complete in 89% of patients. The clinical diagnosis of all patients was confirmed after surgery by histopathological examination of the spleen.

**Figure 1**
Table 1: Criteria Used to Describe Response to Splenectomy “Wani & Parray Criteria”

<table>
<thead>
<tr>
<th>Complete Response (CR)</th>
<th>Partial Response (PR)</th>
<th>No Response (NR)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Platelet count (per mm³)</td>
<td>≥ 100000</td>
<td>50000-100000</td>
</tr>
<tr>
<td>Leukocyte count (per mm³)</td>
<td>≥ 4000</td>
<td>2000-4000</td>
</tr>
<tr>
<td>Hemoglobin (g%)</td>
<td>≥ 10</td>
<td>8-10</td>
</tr>
</tbody>
</table>

Patients with platelet count 100000/mm³, Leukocyte count 4000/mm³ and Hemoglobin 10g% were all included in the category of CR.

**RESULTS**

The preoperative hematological parameters of our study group are described in Table 2. The mean platelet count in the preoperative period was 30,650/mm³.

**Figure 2**
Table 2: Preoperative Hematological Parameters in Patients with ITP

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Range</th>
<th>No. of Cases</th>
<th>% Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thrombocytopenia</td>
<td>Platelet &lt; 50000</td>
<td>40</td>
<td>75</td>
</tr>
<tr>
<td></td>
<td>Platelets 50000-100000</td>
<td>13</td>
<td>25</td>
</tr>
<tr>
<td>Anemia</td>
<td>Hemoglobin &lt; 8 g%</td>
<td>2</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td>Hemoglobin 8-10 g%</td>
<td>6</td>
<td>4</td>
</tr>
</tbody>
</table>

The response to splenectomy was described in terms of hematological parameters as explained earlier and is tabulated in table 3.

**Figure 3**
Table 3: Response to Splenectomy in 52 Patients with ITP

<table>
<thead>
<tr>
<th>Response</th>
<th>No. of Cases</th>
<th>% Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thrombocytopenia</td>
<td>CR</td>
<td>41</td>
</tr>
<tr>
<td></td>
<td>PR</td>
<td>7</td>
</tr>
<tr>
<td></td>
<td>NR</td>
<td>6</td>
</tr>
<tr>
<td>Anemia</td>
<td>CR</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>NR</td>
<td>2</td>
</tr>
</tbody>
</table>

One patient died in the post-operative period and is not included in the table.

Overall, 90% of patients benefited from the procedure. Mean splenic weight was 146g and an accessory spleen was found in 11 patients.

The prognostic factors for splenectomy were analyzed in relation to patient's age, preoperative platelet count, duration of illness, and any concomitant disease. The results are shown in Table 4.

**Figure 4**
Table 4: Analytical Response to Splenectomy: Prognostic Factors (n = 53)

<table>
<thead>
<tr>
<th>Complications</th>
<th>No. of Pts.</th>
<th>Morbidity</th>
<th>Mortality</th>
<th>CR</th>
<th>PR</th>
<th>NR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt; 50</td>
<td>41</td>
<td>6 (15%)</td>
<td>X</td>
<td>34 (84%)</td>
<td>5 (12%)</td>
<td>2 (5%)</td>
</tr>
<tr>
<td>≥ 50</td>
<td>12</td>
<td>3 (25%)</td>
<td>1</td>
<td>7 (64%)</td>
<td>2 (18%)</td>
<td>2 (18%)</td>
</tr>
<tr>
<td>Preop. Platelet Count</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt; 50000</td>
<td>40</td>
<td>9 (23%)</td>
<td>1</td>
<td>30 (77%)</td>
<td>5 (13%)</td>
<td>4 (10%)</td>
</tr>
<tr>
<td>≥ 50000</td>
<td>13</td>
<td>X</td>
<td>X</td>
<td>11 (85%)</td>
<td>2 (15%)</td>
<td>X</td>
</tr>
<tr>
<td>Duration of illness (months)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt; 1</td>
<td>20</td>
<td>2 (10%)</td>
<td>1</td>
<td>17 (85%)</td>
<td>2 (10%)</td>
<td>1 (5%)</td>
</tr>
<tr>
<td>≥ 1</td>
<td>30</td>
<td>7 (2%)</td>
<td>1</td>
<td>24 (75%)</td>
<td>5 (15%)</td>
<td>3 (9%)</td>
</tr>
<tr>
<td>Concomitant Illness</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Present</td>
<td>10</td>
<td>7 (70%)</td>
<td>1</td>
<td>8 (80%)</td>
<td>1 (11%)</td>
<td>X</td>
</tr>
<tr>
<td>Absent</td>
<td>43</td>
<td>2 (5%)</td>
<td>X</td>
<td>35 (81%)</td>
<td>6 (14%)</td>
<td>2 (5%)</td>
</tr>
</tbody>
</table>

X: No patient. See table 1 for definition of other abbreviations.

a. One patient died in the post operative period.

b. “Concomitant illness” included 6 patients with anemia and 5 patients with cholelithiasis (2 patients from the cholelithiasis group presented with anemia also).

**MORBIDITY**

The complications encountered were:

Three patients developed wound infection (6%), 3 patients
intra-operative bleeding (6%), 1 patient pneumonia (2%), and 1 patient post-operative bleeding (2%). The overall morbidity was 15%.

**MORTALITY**

Only 1 patient died on the 3rd post-operative day secondary to intra-cerebral hemorrhage (mortality 2%). The patient was female and 64 years old.

**DISCUSSION**

ITP is a disorder characterized by spontaneous and excessive bleeding, prolonged bleeding and normal clotting time, increased capillary fragility, and low platelet count. In patients with ITP, the spleen is not palpable in most of the patients and the bone marrow demonstrates the presence of megakaryocytes, sometimes in greater numbers than normal and sometimes showing unusual shapes or platelet budding. In our study of 53 patients with ITP, the mean age was 30 years (range 7-66), male to female ratio was 1.03:1. Most studies report a higher incidence for females but less than reported in another study. The reason can be lack of health education in our female population.

The mean platelet count in the preoperative period was 30,650/mm$^3$ (range 4000-85,000), consistent with other studies. All our patients received steroids (prednisone) in the preoperative period. Forty patients received prednisone for 3 weeks and were referred to our department when they did not show any therapeutic response and 13 patients showed only transitory therapeutic effect when given multiple cycles of prednisone therapy and were also referred for splenectomy.

Fifty patients (94%) were subjected to elective splenectomy and 3 patients (6%) to emergency splenectomy. Four patients had concomitant diagnosis of cholelithiasis and were treated for both diseases in the same sitting. The patients were put on hydrocortisone in the post-operative period and all were weaned off the drug without any complication in 48-72 hrs.

The splenic weight ranged from 130 to 650g (mean wt. 145g), which is less than observed by others, but less than reported in another study. However, our results are consistent with those of Little JM. A good number of accessory spleens was a proper and a meticulous search for spleniculi at the time of initial surgery. Many previous researchers have shown concern about accessory spleens in patients with ITP since evidence emerged that sustained remission was obtained after accessory splenectomy in patients with recurrent ITP. Relapses do occur following splenectomy for ITP, yet the rate of complete responses did not change over time in these case series. At this time, the published data including analysis of 21 case series with follow-up of more than 5 years suggest that the response to splenectomy is durable.

Laparoscopic splenectomy for ITP, however, has advantages, like: great visualization of the splenic pedicle, avoidance of injury to the pancreatic tail, easy identification of accessory spleens, it can be combined with other procedures like cholecystectomy, avoids muscle cutting incision, decreases post-operative pain, is cosmetically superior and allows faster post-operative recovery. Since our set-up is the only tertiary care institute available in the whole state, we had to deal with all cases by open method only because of the time factor and to avoid the back-log on our theatre lists.

The overall morbidity was 15% and the mortality was 2%. The morbidity reported in our series is consistent with one study, but less than reported in another study. The mortality is consistent with that of Albrechtsen and Ly, but less than in another study. The morbidity and mortality were decreased to a greater extent by proper patient selection and excellent post-operative care in our study group.

The response to splenectomy was excellent (Table 3). Our results are almost consistent with those of the study by Mintz et al., but are definitely better in comparison to another study. The reason was an excellent cooperation between the hematologist and the surgeon ensuring proper patient selection and proper peri- and post-operative care.

We also observed that prognosis was excellent in patients whose preoperative platelet count was >50,000, whose age was <50 years, whose disease was of shorter duration, and who had no concomitant disease (Table 4).

In 7 studies analyzed in a multivariate model, younger age was an independent variable for predicting response. Also the relatively high rates of death and complications despite advances in anesthesia and surgical care may be due to increased recognition of ITP among older persons; the greater risk of complications in older patients and the...
willingness of surgeons to perform surgery in older patients.

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