Predictors of Response and Outcome of Patients with Acquired Hemophilia A

Ifeoma Onwubiko  
*Henry Ford Health System*, IOnwubi1@hfhs.org

Gary Kaperek

Philip Kuriakose  
*Henry Ford Health System*, PKURIAK1@hfhs.org

Zaher K. Otrock  
*Henry Ford Health System*, zotrock1@hfhs.org

Follow this and additional works at: [https://scholarlycommons.henryford.com/merf2019clinres](https://scholarlycommons.henryford.com/merf2019clinres)

Recommended Citation
Onwubiko, Ifeoma; Kaperek, Gary; Kuriakose, Philip; and Otrock, Zaher K., "Predictors of Response and Outcome of Patients with Acquired Hemophilia A" (2019). *Clinical Research*. 55.  
PREDICTORS OF RESPONSE AND OUTCOME OF PATIENTS WITH ACQUIRED HEMOPHILIA A

Ifeoma Onwubiko, Gary Kaperek, Philip Kuriakose, Zaher Otrock
INTRODUCTION

- AHA is caused by auto antibodies against coagulation factor VIII.
- Incidence: 0.2 to 1.5 persons per million yearly
- Mortality rate up to 25%.
- Associated with malignancy, autoimmune disorders, pregnancy, post-partum period and lymphoproliferative disorders
- Elderly > Young
INTRODUCTION

- Presentation: melena, hematuria, hemorrhages (intracranial, retroperitoneal, soft tissue etc.)
- Treatment involves treating the underlying condition, and to remove/neutralize autoantibodies against FVIII.
- Therapies include: Immunosuppressives, cytotoxics, plasmapheresis, recombinant factors and complex concentrates used alone or in combination.
- Prognosis is unpredictable. Some achieve complete remission, others have a chronic course.
- Clinical characteristics and outcomes are difficult to establish due to rarity.
MATERIALS AND METHODS

- Study was approved by HFH IRB
- Retrospectively evaluated 25 consecutive patients with AHA managed at HFH main campus from 02/2007 to 01/2018 using Sunquest (HFHS LIS).
- Eligible patients had reduced FVIII activity levels, detectable FVIII inhibitors, no hx of congenital hemophilia A.
- All blood samples were collected in Na citrate tubes. FVIII inhibitor was assayed using Bethesda unit (BU).
- Complete response (CR) was defined when FVIII inhibitors were negative with resolution of bleeding signs.
RESULT

- Statistical analysis were performed using SPSS software version 2. Parameters were analyzed using Student’s t-test, Pearson’s chi-square test and Fischer’s exact test.
- Median age was 73.5 years (range 55.1-91.7 years)
- M > F. 13 (52%) were males.
- Underlying conditions: Malignancy (8 including GI CA, Prostate Adenocarcinoma, Lung carcinoma, Cervical carcinoma, follicular lymphoma, SLL/CLL, squamous cell carcinoma); infection (HIV and hepatitis)
Age distribution

<table>
<thead>
<tr>
<th>Age in years</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>55-59</td>
<td>4</td>
</tr>
<tr>
<td>60-64</td>
<td>1</td>
</tr>
<tr>
<td>65-69</td>
<td>4</td>
</tr>
<tr>
<td>70-74</td>
<td>5</td>
</tr>
<tr>
<td>75-79</td>
<td>3</td>
</tr>
<tr>
<td>80-84</td>
<td>5</td>
</tr>
<tr>
<td>85-89</td>
<td>2</td>
</tr>
<tr>
<td>90-94</td>
<td>1</td>
</tr>
</tbody>
</table>
Table 1: Initial characteristics of our patients with acquired hemophilia A

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender (M/F)</td>
<td>13/12</td>
</tr>
<tr>
<td>Age, median (Range), years</td>
<td>73.5 (55.1-91.7)</td>
</tr>
<tr>
<td>Ethnicity (Caucasian/African American)</td>
<td>14/11</td>
</tr>
<tr>
<td><strong>Underlying disease, N (%)</strong></td>
<td></td>
</tr>
<tr>
<td>Idiopathic</td>
<td>13 (52%)</td>
</tr>
<tr>
<td>Malignancy</td>
<td>8 (32%)</td>
</tr>
<tr>
<td>Infection</td>
<td>2 (8%)</td>
</tr>
<tr>
<td>Autoimmune</td>
<td>2 (8%)</td>
</tr>
<tr>
<td>Hgb, median (Range), g/dL</td>
<td>7.5 (3.8-13.5)</td>
</tr>
<tr>
<td>aPTT, median (Range), sec</td>
<td>80 (44-125)</td>
</tr>
<tr>
<td>Factor VIII activity, median (Range), %</td>
<td>1 (0.29-34)</td>
</tr>
<tr>
<td>Factor VIII inhibitor titer, median (Range), BU</td>
<td>30 (0.8-702)</td>
</tr>
</tbody>
</table>
RESULT

- FVIII activity was decreased in all patients by FVIII inhibitors (median=30 BU; range: 0.87-702 BU)
- 20% of patients required PRBC transfusion (median=7 units; range: 1-40 units).
- 76% patients achieved CR after initial immunosuppressive therapy with prednisone alone, in combination with cyclophosphamide and rituximab.
- Median time to CR was 50 days (range: 2-191 days)
- CR was achieved early (<1 month) in patients with low FVIII inhibitor titers (<20 BU); odds ratio= 4.33, CI: 1.61-11.69; p= 0.003.
- Pts with high FVIII inhibitors titers (>20 BU) received > 5 units of PRBCs compared to patients with low titer levels (odds ratio-4.33; CI: 1.14-38.83, p=0.047).
- 2 patients relapsed at 10 months and 2 years respectively.
- 1 patient died of bleeding complication
Factor VIII inhibitor titers and packed RBC transfusion

- Factor VIII inhibitor titer (Bethesda unit)
- RBC transfusion (units) x10^-1
REFERENCES