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Journal Title: Pediatric Rheumatology
Volume: Volume 10, Number Suppl 1
Publisher: BioMed Central | 2012-07-13, Pages A73-A73
Type of Work: Article | Final Publisher PDF
Publisher DOI: 10.1186/1546-0096-10-S1-A73
Permanent URL: https://pid.emory.edu/ark:/25593/s5h6h

Final published version: http://dx.doi.org/10.1186/1546-0096-10-S1-A73

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Accessed April 2, 2019 8:49 AM EDT
Pulmonary thromboembolism in children with rheumatic diseases

Larry B Vogler4*, Sheila Angeles-Han3, Sampath Prahalad2, Egla C Rabinovich1

From 2011 Pediatric Rheumatology Symposium sponsored by the American College of Rheumatology
Miami, FL, USA. 2-5 June 2011

Purpose
To demonstrate the clinical features and predisposing factors of pulmonary thrombotic events in children with rheumatic diseases.

Methods
Chart review, observational.

Results
Thrombotic events have been associated with antiphospholipid antibodies in autoimmune diseases, including systemic lupus erythematosus (SLE). However, pulmonary thromboembolism (PTE) from deep vein thromboses (DVT) or in situ pulmonary arterial thrombosis is uncommon in rheumatic diseases, especially in children. The diagnosis and treatment of PTE may be delayed due to a paucity of symptoms or to symptoms attributed to more common manifestations such as pleuritis or pneumonia. We report findings in 6 children with PTE secondary to SLE (4), Systemic Sclerosis (SSc) (1) and Polyarteritis Nodosa (PAN) (1).

Conclusion
Although antiphospholipid antibodies are common in SLE, pulmonary arterial thrombosis is rare. These 4 cases of SLE represent only 1.7% of 234 pediatric lupus patients seen at Emory University over 18 years. Pulmonary thromboemboli may mimic pleuritis with effusion or pneumonia. Besides antiphospholipid antibodies, which were present in only 2 of these patients, other risk factors include lupus anticoagulants (4 cases) and a factor V Leiden mutation (2 cases). A full list of the clinical findings is provided in Table 1.

Table 1

<table>
<thead>
<tr>
<th>Pt/Gender</th>
<th>1/F</th>
<th>2/F</th>
<th>3/M</th>
<th>4/F</th>
<th>5/M</th>
<th>6/M</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dx</td>
<td>SLE</td>
<td>SLE</td>
<td>SLE</td>
<td>SLE</td>
<td>SSc</td>
<td>PAN</td>
</tr>
<tr>
<td></td>
<td>thrombocytopenia</td>
<td>nephritis (IV)</td>
<td>nephritis (IV)</td>
<td>nephritis (IV)</td>
<td>PAH</td>
<td>CVA</td>
</tr>
<tr>
<td>Age at Dx (yr)</td>
<td>12.6</td>
<td>14.1</td>
<td>9.0</td>
<td>12.0</td>
<td>12.8</td>
<td>0.3</td>
</tr>
<tr>
<td>Age at PTE</td>
<td>15.2</td>
<td>14.8</td>
<td>16.8</td>
<td>12.6</td>
<td>16</td>
<td>6/4</td>
</tr>
<tr>
<td>Symptoms</td>
<td>leg pain</td>
<td>chest pain</td>
<td>chest pain</td>
<td>chest pain</td>
<td>chest pain</td>
<td>leg pain</td>
</tr>
<tr>
<td></td>
<td>dyspnea</td>
<td>dyspnea</td>
<td>dyspnea</td>
<td>dyspnea</td>
<td>dyspnea</td>
<td></td>
</tr>
<tr>
<td>DVT</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Lupus AC</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>+/-</td>
</tr>
<tr>
<td>Anticardio AB</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>D-dimer  (ng/ml)</td>
<td>647 (nl &lt;220)</td>
<td>8770</td>
<td>1600</td>
<td>&gt;10,000</td>
<td>n/a</td>
<td>&gt;10,000</td>
</tr>
<tr>
<td>Albumin (g/dl)</td>
<td>4.7 (nl 3.7-5.5)</td>
<td>2.0</td>
<td>0.7</td>
<td>1.7</td>
<td>4.1</td>
<td>3.6</td>
</tr>
<tr>
<td>AT III (%)</td>
<td>105 (nl 77-132)</td>
<td>278</td>
<td>7/2</td>
<td>154</td>
<td>n/a</td>
<td>114</td>
</tr>
<tr>
<td>Fibrinogen (ng/dl)</td>
<td>718 (nl 180-394)</td>
<td>234</td>
<td>n/a</td>
<td>298</td>
<td>n/a</td>
<td>421</td>
</tr>
</tbody>
</table>

[PAH: pulmonary arterial hypertension, CVA: cerebral vascular accident, AT III: anti-thrombin III, N/A: not available] All patients were treated with heparin and improved. No patient had any other genetic risk factors predisposing to thrombophilia.

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associated findings include nephrotic syndrome, elevated D-dimers and elevated fibrinogen levels. Recognition of PTE in pediatric patients with rheumatic diseases and prompt anti-coagulation therapy is important and potentially life-saving.

Disclosure
Larry B. Vogler: None; Sheila Angeles-Han: None; Sanpath Prahalad: None; Eglia C. Rabinovich: None.

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Published: 13 July 2012

doi:10.1186/1546-0096-10-S1-A73
Cite this article as: Vogler et al. Pulmonary thromboembolism in children with rheumatic diseases. Pediatric Rheumatology 2012 10(Suppl 1):A73.