Association between Immune Thrombocytopenic Purpura and Guillain-Barré Syndrome

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September 8th, 2017
Case

21 yo Female who presents with a chief complaint of “raised bumps” or “blisters” on her tongue.
HPI

- 4-5 days of URI symptoms
  - cough
  - blood-tinged sputum
  - blood-tinged rhinorrhea
- Diffuse, mild headache
- Woke with non-painful “raised black bumps” on her tongue
- Later noticed faint rash on thighs, chest, shoulders
Past Medical & Surgical Hx

- Mild cognitive delay
- Guillain-Barré Syndrome 16 months prior
- Severe Scoliosis s/p Spinal fusion
Family Hx

• No pertinent family history including no:
  • Neurological disorders
  • Autoimmune disorders
  • Bleeding disorders
Social Hx

• Lives at home with mother and extended family

• Sexually active with long-term boyfriend

• No tobacco, alcohol or drug use
Medications/Allergies

• Progesterone only IUD

• No known drug, food, or environmental allergies
Physical Exam

• Vitals: T 97.5 F, HR 83, BP 129/83, RR 16, SpO2 98%
• Gen app: well appearing, alert, cooperative, in no acute distress
• Skin: scattered groups of petechiae on bilateral thighs and anterior chest, large ecchymosis on R lateral abdomen, large vertical, well healed surgical scar on posterior thorax
• HEENT: NCAT, MMM, oral mucosa with visible purpura and petechiae, sclera non-icteric, EOMI
Physical Exam- Cont

- CV/Resp: within normal limits
- Abd: soft, non-tender, non-distended, no masses, **no hepatosplenomegaly**
- Ext: warm, well-perfused, no cyanosis or edema
- Neuro: CN II-XII intact, **no focal neurologic findings**
Work-up

- CBC
  - 9.4
  - 11.7
  - 34.2
  - < 3

- CMP
  - 136
  - 102
  - 5
  - 106

- PT/INR and PTT: wnl
Management

• Hematology consulted due to concern for immune thrombocytopenic purpura
• Patient given 40 mg PO dexamethasone and transferred to our facility for further care
Management

Patient remained well appearing after transfer

CBC: 9.3 11.7 35 < 5

Additional labs:
• LDH 189, Haptoglobin 208
• HIV, Hep B, and Hep C neg
• ANA 1.0 (nl 0-0.9)
Management

• Treated with 1 dose of IVIG (1gm/kg)
• Repeat labs the next morning revealed Plt 10
• Pt discharged on 4 additional daily doses of 40 mg Dexamethasone
Hematology Clinic Follow Up

- 1 wk after discharge:
- Repeat Plt 223
- Platelet Antibody Screen Positive (IgG and IgM)
Clinical Question

• Is there an association between Immune thrombocytopenic purpura (ITP) and Guillain-Barré Syndrome (GBS)?
Immune Thrombocytopenic Purpura (ITP)

- Autoimmune reaction against platelets resulting in a platelet count <150
- Incidence of about 1-3/100,000 adults
- Prevalence of about 12/100,000 adults
- 1/5 to 1/3 asymptomatic
Guillain-Barré Syndrome (GBS)

- Autoantibodies against myelin sheath of peripheral nerve axons
- Often occurs after a viral or bacterial infection
- Ascending paresthesia, weakness, and paralysis
- Incidence of about 1/100,000
Concurrent Acute Motor and Sensory Axonal Neuropathy and Immune Thrombocytopenic Purpura

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ABSTRACT Background: Guillain–Barré syndrome (GBS) is a potentially life-threatening autoimmune disease causing demyelination of peripheral nerves. Multiple variants of GBS exist, with acute motor and sensory axonal neuropathy (AMSAN) being the most severe. GBS typically does not occur in the setting of other autoimmune diseases; however, few case reports do exist describing the occurrence. Methods: We describe a patient with acute motor and sensory deficits and thrombocytopenia, ultimately diagnosed with concurrent AMSAN and immune thrombocytopenic purpura (ITP). Results: A 75-year-old woman presented with new onset diplopia and gait instability, however, was found to have a severe thrombocytopenia. Corticosteroids were initiated for ITP and intravenous immunoglobulin for apparent GBS. Nerve conduction studies and her clinical course indicated that she likely had AMSAN. Although her platelet count recovered, her neurologic status remained poor, prompting therapy with plasmapheresis with subsequent mild improvement. Conclusion: A review of the literature revealed eleven previous cases of concurrent GBS and ITP; however, we report the first case of concurrent AMSAN and ITP. Among these cases, trends were noted to include sex, preceding infections, and cranial nerve involvement.
**Trends:** More often Female... Preceding URI... Responded to Steroids and/or IVIG

<table>
<thead>
<tr>
<th>Age/SEX</th>
<th>Preceding Infection</th>
<th>GBS Variant</th>
<th>Cranial Nerve Involvement</th>
<th>Treatment</th>
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<tr>
<td>30/F</td>
<td>URI</td>
<td>AIDP</td>
<td>CN VII</td>
<td>Steroids</td>
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<td>URI</td>
<td>AMSAN</td>
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Adapted from Sato et al.⁵
GBS and ITP Antibodies

- GBS antibodies are directed against myelin gangliosides
- Antibodies against platelet surface gangliosides have not been identified in GBS
- ITP antibodies target platelet surface glycoproteins GPIIa-IIIb or GPIb-IX

- To date, no overlapping antigens involved in the autoimmune process of both GBS and ITP have been found
Zika virus-associated GBS

• The incidence of Zika-associated GBS is estimated at 0.24 per 1000 infections

• Increased incidence of GBS 2.0–9.8 times higher than the pre-Zika era

• Molecular mimicry between glycolipids and surface molecules of infectious agents may explain most of the cases of GBS preceded by infection
Zika virus-associated ITP

• Several cases of Zika virus-associated severe thrombocytopenia have been reported\(^1\)

• All other main causes of isolated severe thrombocytopenia were excluded in these cases
Concluding thoughts

• Given the clear correlation between GBS and ITP, whether concurrent or delayed, further research into Zika virus may provide a better understanding of each of their etiologies.

• We may continue see an increase in both GBS and ITP with Zika virus infections on the rise.
References


Questions?