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Unusual Case of Thrombotic Thrombocytopenic Purpura (TTP) in Capnocytophaga canimorsus bacteremia

Nicole Tong, DO
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Introduction:
Thrombotic Thrombocytopenic Purpura (TTP)

- Pentad: fever, thrombocytopenia, micro-angiopathic hemolytic anemia, transient ischemic deficits, renal failure
- If not treated, mortality >90%. Plasmapheresis decreases mortality <50%

- Causes:
  - Platelet ADAMTS 13 deficiency - when ADAMTS 13 level is less than 10%. A special enzyme ADAMTS 13, aka, VWF metalloprotease, normally degrades very high molecular weight multimers of von Willebrand factor
  - Idiopathic (37%); drug-induced (13%) - cyclosporine, tacrolimus, gemoitbone, bleomycin, cisplatin, clopidogrel, ticlopidine, quinine; autoimmune disease (13%) - SLE; Pregnancy (7%); bloody diarrhea prodrome (6%) - shiga toxin, E. coli O157:H7; hematopoietic stem cell transplant (4%)

- Treatment: plasmapheresis until platelet count remains normal persistently
- DIC is frequently in the differential with TTP

- Differentials diagnostiques: disseminated intravascular coagulation (DIC), hemolytic uremic syndrome; thrombotic thrombocytopenic purpura (HUS/TTP), polyarteritis nodosum (PAN), Henoch Schönlein Purpura (HSP), ANCA-positive vasculitis, anti-phospholipid antibody syndrome

- Working diagnosis: DIC with severe sepsis of unknown source, possible HUS

- Treatment plan:
  - DIC: reversed with fresh frozen plasma
  - Progressive thrombotic vasculopathy of bilateral lower extremities: neurologic work up for vasculitis
  - Systemic inflammatory response syndrome (SIRS) with hemodynamic instability → IV antibiotics, volume resuscitation
  - Acute ARF: continued veno-venous hemodialysis (CVVHD)
  - Adopted mental status due to severe sepsis: ventilator dependent respiratory failure (VWD)

- Chief complaint: worsening bloody diarrhea, 3 days of flulike symptoms, sudden onset of headache, fever and extensive ecchymoses
- PMHx: hypertension on ARB, hyperlipidemia on statin

Hospital Course:
LVHN MICU Day 1-2:

- Physical exam: declining Glasgow Coma Scale, purpural fulminates, red wine colored diarrhea with microclots
- Working diagnosis: purpura fulminans
ds multilayered red, blue, green, and yellow skin changes; extensive ecchymoses

- Lab results:
  - Fibrinogen 193 mg/dl
  - Hgb 7.0
  - Platelets 7,000
  - INR 1.7
  - Creatinine 3.2
  - Schistocytes
  - WBC 12,000
  - LDH 3,000
  - Platelet transfusions
  - ESR 103

- Differential diagnoses:
  - sepsis, hypotension, renal failure, purpura fulminans, DIC and TTP/HUS: mortality as high as 90%, worse if immunocompromised or asplenia

- Two documented successful cases of Capnocytophaga canimorsus infection treated with plasma exchange as the diagnosis of thrombotic thrombocytopenic purpura was reached.

Conclusion:
In cases of a TTP-like syndrome associated with sepsis and a history or dog or cat exposures or bites, consider Capnocytophaga canimorsus bacteremia even without evidence of a dog bite. Further investigation is needed to evaluate the benefit of plasma exchange in purpura fulminans and dog bite-related sepsis.

Learning Point:
Continual reassessment is imperative in critically ill patients with unclear diagnosis as refined diagnosis and treatment plan can potentially be life-saving.

Special thanks to:
- Nekom King, D.O., Department of Nephrology, LVHN
- Kirsten Bellucci, M.D., Department of Pathology, LVHN
- Luca V., D.O., Department of Dermatology, LVHN

References:

Discussion:
- A gram-negative bacillus, facultative aerobe. Particularly slow growth, difficult to grow by ordinary culture medium, need Wright-Giemsa stain
- Highly susceptible to β-lactam antibiotics, erythromycin, clindamycin, tetracycline. Resistant to aminoglycosides
- Associated with dog and cat bites or saliva exposure
- Reported to cause bacteremia, sepsis, hypotension, renal failure, purpura fulminans, DIC and TTP/HUS: mortality as high as 90%, worse if immunocompromised or asplenia

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