CLINICAL VIGNETTE

Infection Prevention in Adults with Asplenia

Kimberly J. Richardson, MD and Valerie S. Wong, MD

Case Presentation

A 32-year-old female without significant past medical history presents to the emergency department with abdominal pain and shortness of breath. She was riding her bicycle downhill at 20 mph when she hit a pothole and fell over the handlebars. She was wearing a helmet, and there was no loss of consciousness. She arrived via ambulance and experienced a witnessed syncopal episode while returning from the bathroom. Vitals signs included blood pressure 102/62, pulse 103, temperature 36.8°C, respiratory rate 17, SpO2 100%, height 5'8" and weight 185 Physical exam was remarkable for facial abrasions/ lbs. lacerations and a diffusely tender abdomen. Laboratory evaluation was significant for white blood cell count 16.0 x 10^3 , potassium 3.0 mmol/L, and random glucose 227 mg/dL. CT abdomen/pelvis showed splenic laceration with hemoperitoneum. She was admitted to general surgery and underwent emergent exploratory laparotomy and splenectomy.

The postoperative period was complicated by readmission for abdominal pain and fever. CT abdomen/pelvis revealed extensive thrombosis in the portal venous system with extension to the superior mesenteric vein. Hematology was consulted, and initiated on anticoagulation with apixaban for six months to be followed by hypercoagulable testing. Cardiolipin IgG was moderately positive but all other hypercoagulable testing was unremarkable. She had persistent sinus tachycardia and Cardiology was consulted. Zio Patch cardiac monitoring showed an exaggerated heart rate response to minimal activity in addition to occasional premature atrial and ventricular contractions. She was diagnosed with autonomic dysfunction and initiated on beta blocker therapy. She was advised to follow up with her primary care provider for immunization recommendations following splenectomy and initiated post-splenectomy penicillin prophylaxis for one year. She also received pneumococcal, meningococcal, Haemophilus influenzae type b (Hib), and seasonal influenza vaccinations.

Discussion

Surgical removal of the spleen following traumatic injury is the most common cause of asplenia.¹ Approximately 40,000 splenic traumas occur annually in the United States with a small percentage requiring emergent splenectomy.¹ Asplenia can be anatomic or functional.² Anatomic asplenia also occurs after therapeutic splenectomy, in the setting of immune thrombocy-topenic purpura (ITP), autoimmune hemolytic anemia and hereditary spherocytosis.² Rarely, anatomic asplenia is caused

by congenital asplenia syndromes.² Functional asplenia occurs with loss of spleen function in sickle cell disease or other hemoglobinopathies, oncologic, immunologic, or secondary to untreated HIV infection.²

The spleen is a secondary lymphatic organ with two main functions in two anatomical parts: blood filtering in the red pulp and mounting an immune response in the white pulp.^{1,2} It communicates with the digestive system via the hepatic portal venous system.¹ The spleen filters out senescent erythrocytes and protects against encapsulated bacteria through a B-cell mediated immune response.^{1,2} It also has an important role protecting against intraerythrocytic parasitic infections like malaria and *Babesia* due to removal of damaged red blood cells.²

Complications of asplenia include infection, thrombosis and pulmonary hypertension.² Infection is most concerning with a mortality of up to 50% without the recommended immunizations and antibiotic prophylaxis.² Severe and systemic infections in asplenia are referred to as overwhelming postsplenectomy infection (OPSI) most commonly caused by encapsulated bacteria including *Streptococcus pneumoniae*, *Neisseria meningitidis*, and *Haemophilus influenzae* type b.^{1,2} Severity of infection is determined by patient age, immune status, comorbidities, and the microorganism.¹ The risk of infection is highest in the first 3 years post-splenectomy, and although rare, infections can occur decades after the spleen is removed.¹

Daily antibiotic prophylaxis is recommended following splenectomy, without consensus regarding optimal duration of therapy.² This recommendation based on trials showing a 50-63% reduction in pneumococcal infection in children with sickle cell disease receiving penicillin prophylaxis.² Lifelong prophylaxis is recommended for immunocompromised patients or those with a previous episode of sepsis.² United States and Australian guidelines recommend one to three years of antibiotic prophylaxis post-splenectomy.²

Vaccination is the most effective infection prevention method. The Advisory Committee on Immunization Practices (ACIP) of the Centers for Disease Control and Prevention (CDC) recommends pneumococcal, meningococcal, *Haemophilus* influenzae type b (Hib) and influenza vaccinations for patients with asplenia.² The ACIP recently updated the pneumococcal

vaccination recommendations. Eligible adults are given either 15-valent pneumococcal conjugate vaccine (PCV15) in series with 23-valent pneumococcal polysaccharide vaccine (PPSV23) or 20-valent pneumococcal conjugate vaccine (PCV20) alone.^{1,3} There are two vaccines to protect against meningococcal disease.^{2,4} MenACWY protects against serogroups A, C, W and Y, while MenB protects against serogroup B.⁴ Despite recommendations from the ACIP, immunization rates among patients with asplenia is low.⁴ One American retrospective showed only 28.1% of individuals with newly diagnosed asplenia received the MenACWY vaccine and only 9.7% received the MenB vaccine in the first three years following diagnosis.⁴ Participants with at least one year of continuous enrollment in the study after the first dose, showed only 11.6% completion of MenACWY vaccination series and 51.3% completion the MenB vaccination series.⁴

Barriers for implementing vaccination recommendations include healthcare provider lack of knowledge, failure to identify at-risk patients, and poor access to vaccination opportunities for those at risk.⁴ Patients and their families should receive education regarding the risk of OPSI in asplenia and the importance of immunizations in infection prevention.^{1,2.} Countries including Australia, the UK and New Zealand have implemented registries for patients with asplenia to improve their overall management.¹

Conclusion

The most concerning complication of anatomic or functional asplenia is severe and systemic infection. The recommendations for adults with asplenia include antibiotic prophylaxis, immunizations and patient and family education for infection prevention. Clinicians should review immunization history at each visit and refer to the ACIP guidelines for updated recommendations. Spleen registries can improve patient management and quality of life.

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