

Hypersplenism: Clinical Evaluation and Management

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Abstract

Hypersplenism is a clinical syndrome characterized by splenomegaly, peripheral cytopenias, preserved or increased bone marrow cellularity, and improvement in blood counts following splenic intervention. It is most commonly associated with portal hypertension secondary to chronic liver disease but may also occur in hematological malignancies, chronic infections, autoimmune disorders, and storage diseases. Patients may present with symptoms related to anemia, leukopenia, or thrombocytopenia, which can significantly affect morbidity and quality of life. Differentiating hypersplenism from primary bone marrow disorders is essential, as management strategies differ substantially. This narrative review outlines the pathophysiology, etiological spectrum, clinical evaluation, diagnostic approach, and management of hypersplenism, with an emphasis on practical clinical understanding.

Keywords: Hypersplenism; Splenomegaly; Cytopenia; Portal hypertension; Spleen

Introduction

The spleen is an important organ involved in immune function and the regulation of circulating blood cells. Under normal conditions, it contributes to the clearance of aged or damaged cells while maintaining adequate levels of erythrocytes, leukocytes, and platelets in the peripheral circulation. Hypersplenism is a clinical syndrome in which exaggerated splenic activity leads to excessive sequestration and destruction of blood cells, resulting in peripheral cytopenias. While splenomegaly is commonly associated with this condition, the diagnosis is primarily based on functional abnormalities rather than splenic size alone.

Hypersplenism is frequently encountered in clinical practice, particularly in patients with portal hypertension due to chronic liver disease, as well as in those with hematological disorders, chronic infections, autoimmune diseases, and storage disorders. The resulting cytopenias may manifest as fatigue, susceptibility to infections, or bleeding tendencies, often leading to diagnostic confusion with primary bone marrow disorders. Early recognition of hypersplenism is therefore essential to avoid unnecessary investigations and to guide appropriate management strategies.

Pathophysiology

Hypersplenism develops as a consequence of increased splenic blood flow, congestion, and heightened phagocytic activity. In conditions such as portal hypertension, elevated portal venous pressure leads to splenic congestion, resulting in prolonged retention of blood cells within the splenic cords. This

prolonged exposure increases the likelihood of interaction between circulating blood cells and splenic macrophages, leading to their premature destruction.

Under normal physiological conditions, approximately one-third of circulating platelets are sequestered within the spleen. In hypersplenism, splenic pooling may increase markedly, sometimes involving up to 80–90% of circulating platelets, resulting in thrombocytopenia. Similar mechanisms contribute to the development of anemia and leukopenia. In certain disorders, immune-mediated processes may further accelerate peripheral blood cell destruction.

Despite the presence of peripheral cytopenias, bone marrow function typically remains normal or shows compensatory hyperplasia, reflecting an appropriate hematopoietic response. This preservation of marrow activity is a key feature that distinguishes hypersplenism from primary bone marrow failure syndromes.

Etiology

Hypersplenism is usually secondary to an underlying systemic disorder. Common etiological categories include:

- Portal hypertension, most frequently due to liver cirrhosis
- Hematological disorders such as lymphomas, leukemias, and myeloproliferative neoplasms
- Infectious diseases including malaria, visceral leishmaniasis, tuberculosis, and infective endocarditis
- Autoimmune conditions such as systemic lupus erythematosus and rheumatoid arthritis
- Storage disorders including Gaucher disease and other inherited metabolic disorders

Clinical Evaluation

History

Patients may present with manifestations related to cytopenias, including fatigue, dyspnea, recurrent infections, or easy bruising and bleeding. Symptoms suggestive of the underlying cause should be actively elicited, such as jaundice, abdominal distension, fever, weight loss, joint pain, or night sweats. A thorough history should include alcohol consumption, risk factors for viral hepatitis, travel to endemic regions, and features suggestive of autoimmune disease.

Physical Examination

Splenomegaly is the most consistent clinical finding and is typically firm, smooth, and non-tender. Massive splenomegaly raises suspicion for hematological malignancies, chronic infections, or storage disorders. Additional findings may include hepatomegaly, ascites, dilated abdominal veins, lymphadenopathy, or stigmata of chronic liver disease.

Diagnostic Approach

Laboratory Investigations

- Complete blood count demonstrating anemia, leukopenia, thrombocytopenia, or pancytopenia
- Peripheral blood smear usually showing normal morphology
- Reticulocyte count, which may be elevated due to compensatory erythropoiesis
- Liver function tests to assess underlying hepatic disease
- Serological tests guided by clinical suspicion

Bone marrow examination, when indicated, typically reveals normal or increased cellularity, supporting the diagnosis of hypersplenism.

Imaging

Ultrasonography is the preferred initial imaging modality for confirming splenomegaly and assessing portal hypertension. Doppler studies can evaluate portal and splenic venous flow. Computed tomography or magnetic resonance imaging may be used to assess splenic architecture, vascular anatomy, and associated abdominal pathology.

Differential Diagnosis

Conditions to consider include aplastic anemia, myelodysplastic syndromes, bone marrow infiltration, hemolytic anemias, and drug-induced cytopenias. The presence of splenomegaly along with preserved or increased bone marrow cellularity favors hypersplenism.

Management

Treatment of the Underlying Cause

Management is primarily directed toward addressing the underlying etiology. Effective treatment of portal hypertension, eradication of infections, or appropriate therapy for hematological malignancies often leads to improvement in cytopenias.

Medical Management

Medical treatment is largely supportive and focuses on correction of anemia, prevention and treatment of infections, and management of bleeding complications. In cirrhosis-related hypersplenism, non-selective beta-blockers may be used to reduce portal pressure. Immunosuppressive therapy may be indicated in autoimmune conditions when appropriate.

Interventional and Surgical Management

Splenectomy is reserved for patients with severe, symptomatic cytopenias that are refractory to medical therapy. Partial splenic embolization may be considered as an alternative in selected patients who are poor surgical candidates. In portal hypertension-related hypersplenism, transjugular intrahepatic portosystemic shunt placement may help reduce splenic congestion and improve cytopenias.

Complications

Potential complications include severe cytopenias, recurrent infections, bleeding episodes, and thrombotic events following splenic interventions. Patients undergoing splenectomy are at increased risk of overwhelming post-splenectomy infection and require appropriate vaccination and long-term preventive measures.

Prognosis

The prognosis of hypersplenism largely depends on the underlying cause. Cytopenias related to portal hypertension may improve with effective management of liver disease, whereas those associated with malignancies or storage disorders often carry a more guarded prognosis.

Conclusion

Hypersplenism is an important clinical syndrome with diverse etiologies and presentations. Recognition of its characteristic features allows differentiation from primary bone marrow disorders and helps prevent unnecessary investigations. Management should be individualized, with primary emphasis on treating the underlying condition, while splenic interventions should be reserved for selected cases.