Case Report

ANNALS OF MEDICAL AND CLINICAL CASES

Pulmonary Thromboembolism Following Splenectomy In Hereditary Spherocytosis

Varun Reddy
Department of Medicine,UK

***Corresponding Author:**
Varun Reddy,
Department of Medicine, UK

Received Date: 27 Mar 2026
Accepted Date: 16 Apr 2026
Published Date: 23 Apr 2026

Citation: Varun Reddy. Pulmonary Thromboembolism Following Splenectomy In Hereditary Spherocytosis. AMCC. 2026; 19(2): 1-2

1. Abstract

Hereditary spherocytosis is a genetic hemolytic disorder characterized by spherical red blood cells and commonly presents with anemia, splenomegaly, and jaundice. Splenectomy remains the standard treatment for symptomatic patients. Thromboembolic complications following splenectomy in hereditary spherocytosis are rare. We report the case of a 36-year-old male who developed pulmonary thromboembolism along with right ventricular mural thrombosis six years after undergoing splenectomy, in the absence of identifiable prothrombotic risk factors. This case underscores a rare association and highlights the need for awareness of late thromboembolic complications in such patients.

Keywords: Hereditary spherocytosis, splenectomy, pulmonary embolism, thrombosis

2. Introduction

Hereditary spherocytosis (HS) is an inherited hemolytic anemia caused by structural defects in red blood cell membrane proteins, leading to decreased deformability and premature destruction of erythrocytes. The condition is most commonly inherited in an autosomal dominant manner, though autosomal recessive forms are also recognized.

Clinically, HS typically manifests as a triad of anemia, splenomegaly, and jaundice. Splenectomy is widely accepted as the treatment of choice in symptomatic individuals, as it significantly reduces hemolysis and improves hematological parameters.

Although splenectomy is generally safe, it has been associated with certain long-term complications, including an increased risk of infections and, less commonly, thromboembolic events. Pulmonary thromboembolism (PTE) in patients with hereditary spherocytosis is rare and not well documented.

3. Case Report

A 36-year-old unmarried male presented with a 7-day history of left lower limb swelling and tenderness, accompanied by progressive breathlessness on exertion.

Six years prior, he had been evaluated for anemia and diagnosed with moderate hereditary spherocytosis based on increased osmotic fragility and peripheral smear findings. Other investigations, including Coombs test, G6PD assay, hemoglobin electrophoresis, and coagulation profile, were within normal limits. He subsequently underwent splenectomy and remained asymptomatic until the current presentation. Screening of first-degree relatives was negative, and genetic testing was not performed.

4. Clinical Findings

On examination, the patient was tachycardic and tachypneic, with normal blood pressure and elevated jugular venous pressure. Cardiac examination revealed a right ventricular third heart sound. The lungs were clear on auscultation. Mild hepatomegaly with tenderness was noted. Examination of the left lower limb showed features of superficial thrombophlebitis along the course of the great saphenous vein. Homan's sign was negative.

5. Investigations

5.1. Peripheral blood smear: Presence of spherocytes, nucleated red blood cells, and Howell–Jolly bodies

5.2. Electrocardiogram: Sinus tachycardia with right ventricular strain pattern

5.3. Echocardiography:

- Dilated right atrium and right ventricle
- Elevated pulmonary artery pressure (~75 mmHg)
- Reduced right ventricular systolic function
- Normal left ventricular ejection fraction (~60%)

5.4. CT Pulmonary Angiography:

- Multiple emboli in the left main pulmonary artery and right descending pulmonary artery
- Wedge-shaped pulmonary infarction in the left lower lobe
- Right ventricular mural thrombus

5.5. Lower limb Doppler ultrasound: No evidence of deep vein thrombosis in proximal veins

5.6. Thrombophilia profile:

- Protein C, Protein S, Antithrombin III
- Factor V Leiden mutation
- Factor VIII levels
- Lupus anticoagulant

→ All within normal limits

6. Management and Outcome

The patient was started on low molecular weight heparin, followed by transition to oral anticoagulation therapy. He showed marked

clinical improvement with resolution of symptoms.

7. Discussion

Hereditary spherocytosis is not typically associated with a marked increase in thrombotic risk. While isolated reports of thromboembolic events exist, they are often linked to additional risk factors such as inherited thrombophilias or specific clinical conditions.

Splenectomy, however, has been implicated as a potential contributor to thromboembolic complications. The proposed mechanisms include:

- Persistent circulation of abnormal erythrocytes
- Increased platelet count and activation
- Altered vascular dynamics and endothelial interactions

There is also evidence suggesting an association between splenectomy and the later development of pulmonary hypertension, possibly mediated by recurrent thromboembolic events or in situ thrombosis.

Animal studies have demonstrated increased thrombogenic potential in the presence of abnormal red blood cells, although this relationship has not been conclusively established in humans.

In this case, no conventional prothrombotic factors were identified, making the temporal relationship between splenectomy and pulmonary thromboembolism particularly significant.

8. Conclusion

Pulmonary thromboembolism is an uncommon but important late complication in patients with hereditary spherocytosis following splenectomy. Clinicians should remain vigilant for thromboembolic events in such patients, even in the absence of identifiable risk factors. Early diagnosis and timely initiation of anticoagulation therapy are essential to improve outcomes.

References

1. Hayag-Barin JE, Smith RE, Tucker FC, Jr. Hereditary spherocytosis, thrombocytosis, and chronic pulmonary emboli: a case report and review of the literature. Am J Hematol 1998; 57(1): 82-84.
2. Nikol S, Huehns TY, Kiefmann R, Hofling B. Excessive arterial thrombus in spherocytosis. A case report. Angi-ology 1997; 48(8): 743-748.
3. Au WY, Ma ES. Peripartum pulmonary embolism in a patient with concomitant hemoglobin Newcastle, hemo-globin Constant Spring, and hereditary spherocytosis. Am J Hematol 2005; 80(2): 166.
4. Hoeper MM, Niedermeyer J, Hoffmeyer F, Flemming P, Fabel H. Pulmonary hypertension after splenectomy? Ann Intern Med 1999; 130(6): 506-509.
5. Wandersee NJ, Lee JC, Deveau SA, Barker JE. Reduced incidence of thrombosis in mice with hereditary sphero-cytosis following neonatal treatment with normal hema-topoietic cells. Blood 2001; 97(12): 3972-3975.
6. Stewart GW, Amess JA, Eber SW, Kingswood C, Lane PA, Smith BD, Mentzer WC. Thrombo-embolic disease after splenectomy for hereditary stomatocytosis. Br J Haematol 1996; 93(2): 303-310.
7. Ghosh K, Meera V, Jijina F. Pulmonary hypertension in patients with hematological disorders following splenec-tomy. Indian J Hematol Blood Transfus 2009; 25(2): 45-48.
8. Karnak D, Beder S, Kayacan O, Berk O. Postoperative pulmonary embolism in a young female accompanying with Factor V Leiden mutation and hereditary syphero-cytosis. J Thromb Thrombolysis 2004; 17(3): 213-217.