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Massive pulmonary embolism- Is it because of Behcet's Disease?

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Abstract

A 34 year old female was admitted to the hospital with a chief complaint of recurrent abdominal pain. She had a past medical hisotry of Behcet's disease with multiple abdominal symptoms. She had undergone multiple abdominal surgeries, including an exploratory laparotomy, cholecystectomy and appendectomy. Three days into her admission, the patient syncopized in the bathroom. Vitals were significant for hypotension. CT of the head was negative. EKG showed normal sinus rhythm. Troponins continued to rise over the next 24 hours and an echocardiogram was done showing RV dilation. CT angio of the lungs showed bilateral pulmonary emboli. Lower extremity ultrasound was negative. She was started on anticoagulation and continued on her immunosuppressive agents for Behcet's disease, with significant clinical improvement. This case highlights vascular thrombosis in a patient with Behcet's disease. Behcet's is not well understood and it is thought that vascular inflammation predisposes these patients to developing blood clots. Treatment includes anticoagulation with ongoing immunesuppression for the underlying autoimmune process.

Keywords

behcet's disease; syncope; pulmonary embolism

Introduction

Behcet's Disease (BD) is a chronic relapsing and remitting vasculitis with multisystem involvement. It is commonly referred to as the "Silk Road" disease, due to its prevalence in the Asian and Mediterranean region of the traditional Silk Road [1]. Behcet's can affect arteries and veins of all sizes; involvement of the central nervous system and intestines is thought to result in a poorer patient prognosis [1]. Current genetic and environmental links to the disease are being evaluated, including upregulation of HLA-B27 and heat shock protein 60 as well as association with herpes simplex virus and streptococcal virus [1].

Case Report

A hydromorphone PCA along with continuing her home steroids and azathioprine. She was started on standard heparin subcutaneous injections for deep vein thrombosis prophylaxis while inpatient. Three days into her hospital stay, the patient syncopized in the bathroom. Vitals at the time showed a blood pressure of 70/40 with sinus rhythm CT head was negative for an acute bleed. She

continued to have rising troponins, peaking at 1.68. Transthoracic echocardiogram showed reduced RV systolic function and RA and RV dilation. D-dimer was elevated. CT angio of the chest showed bilateral segmental pulmonary emboli. Lower extremity DVT studies were negative. The Pulmonary Embolism Response Team was activated and suggested catheter directed tPA, which the patient refused. She was then started on a heparin drip and transferred to the intensive care unit for closer monitoring. Her hypercoagulable workup resulted negative. The patient was bridged to rivaroxaban and immunosuppressive agents were continued with significant improvement in clinical status.

Figures

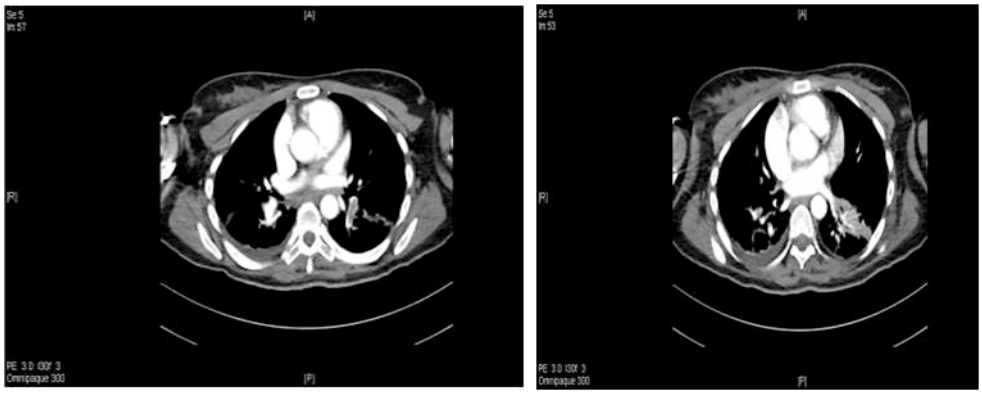

Figure 1 & 2: Sub-massive pulmonary embolism- filling defects seen in the pulmonary artery

Discussion

This case highlights an unprovoked massive pulmonary embolism in a young patient with Behcet's Disease (BD). BD is an autoimmune disease commonly presenting with recurrent oral and genital ulcers, as well as skin and eye involvement [2]. The etiology of this disease is not well-understood and it is thought vascular involvement is due to inflammation localized to the vasa vasorum [3]. Vascular involvement occurs in up to 40% of patients with a male predisposition [4]. Thrombosis is usually secondary to underlying vasculitis, with lower extremity venous thromboembolism presenting as the most common vascular abnormality in BD patients. Pulmonary artery involvement has a prevalence rate of less than 5%, presenting as pulmonary artery aneurysm and less likely as pulmonary artery thrombosis. In most patients, pulmonary artery embolism is isolated and does not occur in concordance with lower extremity venous thrombosis [5].Pulmonary artery embolism is considered one of the most severe and worst prognostic manifestations of this disease. In 2012, a retrospective cohort of 807 patients with BD and venous thrombosis showed that immunosuppressive agents significantly reduced the risk of venous thrombosis relapse rate; especially the use of glucocorticoids [5].

Conclusion

This patient presentation shows the importance of possessing high clinical suspicion of vascular involvement in BD patients presenting with new onset fever, dyspnea, hemoptysis or syncope. It also hopes to showcase the complex clinical management of these patients and the ongoing challenge to prevent recurrent vascular complications.

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