Letter to Editor Rheumatology
www.lettertoeditor.org

An international open-access and peer-reviewed online journal

Letter

Lett Ed Rheumatol
doi:10.2399/ler.12.0006

# Comments on the risk of pulmonary embolism in Behçet's syndrome

**Serdal Ugurlu[1]; Yusuf Yazici[2]; Gulen Hatemi[1]; Sebahattin Yurdakul[1]***

[1]Division of Rheumatology, Department of Internal Medicine, Cerrahpasa Medical Faculty, University of Istanbul, Istanbul, Turkey

[2]Department of Rheumatology, NYU Hospital for Joint Diseases, New York University, New York, NY, USA

**Abstract**

A recent study showed that the risk of pulmonary embolism in Behçet's syndrome (BS) is increased. Although there is venous thrombosis in BS in up to 40% of BS patients, thromboembolism does not occur because of the strong adherence of thrombi to the diseased veins.

**Key words:** Behçet's syndrome, pulmonary embolism, venous thromboembolism, autoimmune diseases

***Correspondence:**
*Prof. Sebahattin Yurdakul*
*Division of Rheumatology,*
*Department of Internal Medicine,*
*Cerrahpasa Medical Faculty,*
*University of Istanbul,*
*Istanbul, Turkey*
*Tel: +90 212 414 30 00*
*e-mail: profsyurdakul@gmail.com*

Dear Editor,

Zöller et al. recently reported on the risk of pulmonary embolism in patients with 33 different autoimmune diseases from Sweden.[1] In this study they obtained data from the MigMed[2] database, constructed from several national Swedish data registers. They studied the risk of venous thromboembolism in patients without previous hospital admission for venous thromboembolism. In addition, these patients had a history of admission for pulmonary embolism with a primary or secondary diagnosis of an autoimmune disorder between Jan 1, 1964, and Dec 31, 2008. The risk of pulmonary embolism was classified into five categories; patients who had <1 year, 1-5, 5-10, >10 years of follow up and all combined. The risk of pulmonary embolism was reported as 6.38 (95% CI: 6.19-6.57) for 33 autoimmune disorders in the first year after hospital admission. On the other hand, the overall follow-up time risk was determined as 1.59 (95% CI: 1.56-1.61). The highest risk for pulmonary embolism was reported in patients with polymyositis and dermatomyositis; 16.4 (95% CI: 11.57–22.69). They showed that the risk of pulmonary embolism during the first year after admission for BS was 9.03 (95% CI: 6.51-12.22), but the overall follow-up time risk was less at 1.68 (95% CI: 1.45-1.93).

Several issues need further discussion: It was suggested in the introduction of the manuscript that BS was associated with a high risk of venous thromboembolism with reference to a review by Yazici Y et al.[2] Although venous thrombosis is present, thromboembolism does not occur in BS probably because of the strong adherence of thrombi to the diseased veins.[3] Moreover, pulmonary embolism was rarely found in a large Japanese autopsy registry.[4] In another study, pulmonary embolism was not observed in 47 patients with pulmonary artery involvement of BS.[5] Pulmonary artery involvement included either pulmonary artery aneurysms or 'in situ' pulmonary artery thrombosis which developed as a complication of underlying vasculitis. Furthermore, it is to be noted that, immunosuppressive agents are essential in treating venous thrombosis and preventing relapses in BS. As reported in retrospective studies,

**Citation:** Ugurlu S, Yazici Y, Hatemi G, Yurdakul S. Comments on the risk of pulmonary embolism in Behçet's syndrome. Lett Ed Rheumatol 2012;2:e120006. doi:10.2399/ler.12.0006

**Received:** May 28, 2012; **Accepted:** September 21, 2012; **Published:** September 26, 2012

**Conflicts of Interest:** The authors have declared that no conflicts of interest exist.

solo anti-coagulation is ineffective in treating venous disease of BS.[6,7] Vascular involvement occurs in up to 40% of BS patients, and the lower extremity deep vein thrombosis is the most common type with a frequency of 60-80%.[8] Thus a SIR of 1.68 in such a population would indeed point out to the scarcity of pulmonary embolism in BS patients. Finally, BS is most active during the third decade of life, burning out after the fifth decade. The tendency for the disease to abate with the passage of time has been reported by several centers around the globe.[8-12] We had observed that mucocutaneous and joint lesions decreased in frequency after 20 years of disease onset and in at least 60% of the patients the disease had been observed to fade away.[8] Thus it is unlikely that an increased risk of pulmonary embolism in men between the ages of 70-79 and in women between 50-69 could be related to BS itself. This could be attributed to another cause rather than BS and older patients with pulmonary embolism should be investigated further and anti-coagulated if needed.

## References


1. Zöller B, Li X, Sundquist J, Sundquist K. Risk of pulmonary embolism in patients with autoimmune disorders: a nationwide follow-up study from Sweden. *Lancet* 2012;379:244-9.
2. Yazici Y, Yurdakul S, Yazici H. Behçet's syndrome. *Curr Rheumatol Rep* 2010;12:429-35.
3. Hamuryudan V, Melikoglu M. Vascular involvement in Behçet's syndrome. In: Yazici Y, Yazici H, editors. Behçet's syndrome. New York: Springer; 2010. p. 115-134.
4. Lakhanpal S, Tani K, Lie JT, Katoh K, Ishigatsubo Y, Ohokubo T. Pathologic features of Behçet's syndrome: a review of Japanese autopsy registry data. *Hum Pathol* 1985;16:790-5.
5. Seyahi E, Melikoglu M, Akman C, et al. Pulmonary artery involvement and associated lung disease in Behçet disease: a series of 47 patients. *Medicine (Baltimore)* 2012;91:35-48.
6. Ahn JK, Lee YS, Jeon CH, Koh EM, Cha HS. Treatment of venous thrombosis associated with Behcet's disease: immunosuppressive therapy alone versus immunosuppressive therapy plus anticoagulation. *Clin Rheumatol* 2008;27:201-5.
7. Desbois AC, Wechsler B, Resche-Rigon M, et al. Immunosuppressants reduce venous thrombosis relapse in Behçet's disease. *Arthritis Rheum* 2012;64:2753-60.
8. Kural-Seyahi E, Fresko I, Seyahi N, et al. The long-term mortality and morbidity of Behçet syndrome: a 2-decade outcome survey of 387 patients followed at a dedicated center. *Medicine (Baltimore)* 2003;82:60-76.
9. Kaklamani VG, Vaiopoulos G, Kaklamanis PG. Behçet's disease. *Semin Arthritis Rheum* 1998;27:197-217.
10. Mamo JG, Baghdassarian A. Behcet's disease; a report of 28 cases. *Arch Ophthalmol* 1964;71:4-14.
11. Pivetti Pezzi P, Gasparri V, De Liso P, Catarinelli G. Prognosis in Behcet's disease. *Ann Ophthalmol* 1985;17:20-5.
12. Shimizu T, Ehrlich GE, Inaba G, Hayashi K. Behçet disease (Behçet syndrome). *Semin Arthritis Rheum* 1979;8:223-60.


 