

The Frequency, nature and predictors of serositis in patients with Behcet's disease: Systematic review and meta-analysis.

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Abstract

Background: Behçet's disease is a chronic inflammatory vasculitis that is not restricted to vessel size and may affect various organs. Behçet's patients commonly present with recurrent oral and genital ulcers. They may also develop uveitis, skin rash or deep vein thrombosis. However, serositis is not a common manifestation of the disease. We aim to investigate whether serositis is related to Behçet's disease or secondary to other etiology, identify predictors and prognosis of such presentation.

Methods: We systematically searched MEDLINE, EMBASSE and Cochrane Central Register of Controlled Trials from data base inception until April 2020 for all cases of Behçet's disease which reported any form of serositis, and excluded interventional studies. Our primary outcome is the frequency of primary vs. secondary serositis among Behçet's disease patients presenting with any form of serositis. Secondary outcomes include predictor of Behçet's related serositis, outcomes of serositis in Behçet's disease and factors associated with poor outcome defined as mortality or lack of response.

Results: We retrieved 517 study records and included 44 case reports, and 7 cohort studies that fulfilled the inclusion criteria. The mean age of included patients was 33.9 ± 11.7 years, 32(72.7%) male. The average disease severity was 7.16 ± 2.15 , and duration of 4.8 ± 8.9 years. Forty-four patients with Behçets disease were affected by serositis, of whom 28(63.6%) were primary to Behçets disease, 10(22.7%) were secondary to other diseases, and 4(9.0%) were undetermined. The secondary causes of serositis were Familial Mediterranean fever, cyclosporine-induced acute

pericarditis, Tuberculosis, Crohn's disease, T-cell large granular lymphocytic leukemia, pneumonia, and rheumatic fever. Serositis presented with peritonitis in 12(27%), Pericarditis 23(52.3%), Pleuritis 20 (45.5%). Four patients (11%) died immediately, while 21(47.7%) experienced full recovery, 7(15.9%) partial, 6(13.6%) not recovered.

Conclusion: Majority of serositis reported in Behçet's disease are related to disease itself rather than secondary etiologies. Most patients respond to the treatment, yet mortality remain high. High risk of bias due to low to moderate quality of reported cases mandate careful interpretation of the results.