Hope Beyond Flares: Escalating Therapy in Resistant Behçet's Disease

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Introduction:
Behçet's disease is a chronic inflammatory condition characterized as a primary systemic vasculitis that affects both arteries and veins of all sizes, leading to a highly variable clinical phenotype amongst patients. The most common initial symptom is recurrent oral aphthous ulcers or genital ulcers. Patients may also present with cutaneous lesions and ocular involvement, but other organ systems can be affected as well. While Behçet's disease is most prevalent in countries along the ancient silk road from the Mediterranean basin to East Asia, it has now been recognized worldwide due to migration and improved awareness of the disease. The mean age of diagnosis is 30 years, and the prevalence is equal between males and females. It typically follows a relapsing and remitting course, with young men often experiencing more severe manifestations. Prompt diagnosis and treatment is extremely important for all patients, as the diffuse nature of this disease can lead to severe and irreversible end-organ damage.

Case:
A 43-year-old female with Behçet's disease diagnosed at age 32 characterized by recurrent oral and genital ulcers presented with five days of worsening oral ulcers, severe throat pain, poor oral intake, medication intolerance, nausea, and dry heaving. She has had a complex disease course managed by rheumatology and was on apremilast, azathioprine, colchicine, and intermittent steroids at the time of her presentation. She was also recently treated for presumed oral thrush with fluconazole. During this hospitalization, an esophagoduodenoscopy showed a non-bleeding gastric ulcer and antral erosions, consistent with gastrointestinal involvement of her Behçet's disease. She was treated for suspected superimposed candidiasis with nystatin, received intravenous solumedrol, and was discharged on a steroid taper along with her home medications. Given her severe disease refractory to her current regimen, rheumatology started her on adalimumab, and she has been stable without flares since.

Discussion:
The pathological course of Behçet's disease often has relapsing and remitting episodes. Although there is no official cure, there are pharmacologic therapies used to prevent recurrent mucocutaneous lesions and further multisystem involvement. However, a subset of patients experience severe, debilitating flares that require careful initiation and titration of aggressive immunosuppressive therapy. Despite early initiation of azathioprine, our patient had frequent refractory symptoms affecting the oropharynx, genitals, and gastrointestinal tract over 10 years. Her medications have frequently been titrated to achieve remission prior to her recent hospitalization. However, escalating to adalimumab has resulted in the most clinically significant symptom relief for this patient. Our case illustrates the high degree of morbidity and debilitation that can be associated with Behçet's disease, even in patients without significant end-organ damage.