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### Behcet's Disease (Malignant Aphthosis) and the Silk Route

I was recently visiting Istanbul for the Annual Scientific Meeting of the World Association of Medical Law.

This got me thinking about the famous Turkish dermatologist Dr Hulusi Behçet – born in Istanbul, who described Behçet's syndrome: a condition marked by oral and genital ulcerations, chronic relapsing uveitis and erythema nodosum.

The exact cause of this syndrome is unknown but there is an associated systemic vasculitis and it is thought that exposure to an infectious or an external agent prompts an auto-inflammatory response in genetically predisposed individuals. There is an increased prevalence of this condition along the Silk Route from China to Turkey and so it is thought genetics have a role to play especially as there is also a familial predisposition.

The average age of first presentation is between 20 and 40 years and both sexes are equally affected. In Arab populations male victims predominate whereas in Korea, China, the US and northern Europe female sufferers are in the majority of cases. Male victims have a more severe clinical course.

Biopsy of the mucocutaneous lesions shows a neutrophil-predominant reaction with endothelial swelling, extravasation of RBCs, and leukocytoclastic vasculitis with fibrinoid necrosis of the blood vessel walls. The neutrophilic vascular reaction is considered the most predominant reaction in Behçet disease.

The vast majority of patients with Behçet disease have oral ulcers which are usually painful, recurrent, and multiple and may involve the soft palate, hard palate, buccal mucosa, tongue, gingiva, lips, and tonsils. The recurrent genital lesions which are found in 80% of patients with Behçet disease involve ulcers of the scrotum, vulva and vagina and in the genitalia scarring is more permanent. Epididymitis can occur.

The involvement of the eyes with uveitis is more common in the initial phases of the disease and is more prominent in male patients. Anterior uveitis causes erythema and photophobia, while posterior uveitis causes vision loss. Retinal involvement with retinal vasculitis can be seen as a cause of blindness in these patients.

Erythema nodosum of the lower limbs is found in 50% of cases and 25% of patients develop superficial thrombophlebitis.

Pulmonary artery involvement with aneurysm formation is unique to Behçet disease and is the leading cause of death in these patients

Other manifestations include CNS involvement – mainly of the brainstem causing cerebellar, pyramidal and sensory symptoms including headache and papilledema. The gastrointestinal symptoms include ulceration of the ileum and caecum and can result in bowel perforation. Renal involvement is uncommon but can involve amyloidosis or glomerulonephritis. Cardiovascular manifestations include pericarditis, myocarditis, endocarditis, coronary artery vasculitis, and coronary aneurysms

#### Treatment

Oral and genital ulcers are usually treated with topical corticosteroids such as triamcinolone qid which can alleviate the pain in ulcers. Recurrences are prevented by the use of colchicine 1-2 g/day in divided doses.

From 1933 to 1948 Professor Hulusi Behçet was appointed as Head of the Department of Dermatology and Venereal Diseases at Istanbul University.



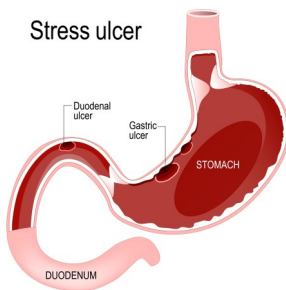
## The dangers of systemic corticosteroids

I was recently involved in a case of acute bowel perforation and peritonitis caused by high dose systemic corticosteroids being used to treat a severe case of bullous pemphigus .

Systemic corticosteroids increase the risk of bowel perforations by interfering with normal tissue repair, increasing susceptibility to ulcers (including peptic ulcers and diverticular perforations), and suppressing the immune system's inflammatory response. The medication can mask the symptoms of a perforation, such as abdominal pain leading to delayed diagnosis and treatment and leading to higher rates of morbidity and mortality, particularly in hospitalized patients.



Peptic ulceration is a known consequence of corticosteroids in high doses and these ulcers can result in GIT bleeding and /or bowel perforation. The other focus for bowel perforation with corticosteroids is diverticulae in the colon. Patients receiving corticosteroids for conditions like COVID-19, along with other medications like Tocilizumab, are at an increased risk of gastrointestinal perforation.



The evidence strongly suggests that corticosteroids can be responsible for ulceration and perforation of the colon either by direct injury or by interference with normal mechanisms of bowel repair.

One study (Narum et al,2017) of 33,000 patients receiving corticosteroids or placebo showed a 2.9% rate of bleeding or bowel perforation compared with 2% for placebo patients .The overall conclusion was that hospitalised patients receiving high dose corticosteroids had a 40% increased risk of GIT bleeding and/or bowel perforation.

It is usually at high doses of corticosteroid administration where multiple and especially severe adverse effects of glucocorticoids occur, ranging from mild suppression of hypothalamic-pituitary axis to severe, life-threatening infections However, long-term use of low to moderate doses of glucocorticoids can also lead to several serious adverse effects

### Further Reading:

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