Caution Advised When Diagnosing Behçet’s

Dr. Kenneth Calamia, the department of medicine at the Mayo Clinic in Jacksonville, Fla., said that in patients diagnosed with Behçet’s disease, everyting will be attributed to the disease for the rest of their lives.

In the United States, true Behçet’s is quite rare (about 0.3-7.5 cases/100,000 populations), compared with places like Turkey and other “Silk Road” areas, which have a very high prevalence (100-370 cases/100,000 population).

In those areas, more severe forms are much more prevalent, and the benign mucocutaneous symptoms that comprise most of the cases in the United States are referred to as American Behçet’s disease, Dr. Calamia said.

The term “Behçet’s syndrome” also can be used to describe the types of cases typically seen in the United States, but in many cases, the diagnosis is actually “complex aphthosis,” he said, giving that in patients diagnosed with Behçet’s disease, everyting will be attributed to the disease for the rest of their lives.

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In a study conducted by an oral dermatologist several years ago, only 9% of 269 patients with severe complex aphthosis – 16% of whom also had genital ulcers – had a Behçet’s diagnosis, he noted.

Some other diagnoses in the cohort included 25% of patients with Behçet’s disease in 16%, hematologic disorders in 5%, mucosal disease in 6%, smoking discontinuation in 4%, and drug-related ulcers in 3%.

“(Complex aphthosis) is the diagnosis I prefer in those who have mouth and genital ulcers, but nothing else to support a diagnosis of Behçet’s,” he said.

Consider the other possible causes of the ulcers, and also consider the differential diagnoses for recurrent aphthous stomatitis, which include recurrent intraoral herpes simplex virus, Wegener’s granulomatosis, Crohn’s disease, psoriasis, oral ulcerations, erythema multiforme, lichen planus, mucous membrane pemphigoid, and pemphigus vulgaris, he said.

A diffuse, widespread, and chronic presentation, which is not characteristic of recurrent aphthous stomatitis or Behçet’s disease, can help differentiate between those conditions and those differential diagnoses, he said.

Dr. Calamia disclosed that he has received research support from Genentech and Celgene, and has served on an advisory board for Centocor.

In another aortitis patient who had been treated with steroids but couldn’t tolerate the side effects – and whose IgG4 levels increased when the steroids were discontinued – rituximab had an equal or abrupt effect. At 1 month following treatment, her IgG4 levels had fallen to 31 mg/dL.

In a 68-year-old man who previously responded to steroids, but who flared and was being treated with various disease-modifying antirheumatic drugs, a single rituximab dose decreased his IgG4 level with each dose until it normalized.

Ten patients with aortitis, including seven with IgG4 elevation, have been treated with rituximab as part of this series, and IgG4 levels declined quickly in all seven, while all other IgG subcategories remained stable, he said.

IgG4-related aortitis was first described in 2009 by Dr. Stone and his colleagues, who published on the case of a 67-year-old patient who developed dissection of the ascending aorta in the setting of IgG4-related disease, thereby linking IgG4-related systemic disease with this newly recognized subset of noninfectious aortitis, and adding to a growing list of conditions, such as autoimmune pancreatitis, that are associated with IgG4-related systemic disease.

At surgery, a transmural lymphoplasmacytic infiltrate was detected in the aorta, and on immunohistoch-