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Behçet's disease associated with diarrhea and secondary amyloidosis

İshal ve sekonder amiloidoz ile ilişkili Behçet Hastalığı

To the Editor,

The symptoms associated with gastrointestinal manifestations of Behçet's disease (BD) are abdominal pain, nausea, vomiting, diarrhea, and constipation (1). Prevalence of secondary amyloidosis in BD has been reported to vary between 0.04 and 3% (2). Mutations in the MEFV gene might be among the risk factors for more severe disease and development of amyloidosis (3). We describe a patient with BD, chronic diarrhea, amyloidosis, and heterozygous M694V mutation (gene mutation for familial Mediterranean fever) on exon 10.

A 54-year-old male patient with BD was admitted with arthralgia of both knees, ankles, elbows, and hips, diarrhea, and swelling of both legs. On physical examination, pretibial edema (+++) and scrotal ulcer scars were detected. Laboratory analysis revealed: erythrocyte sedimentation rate: 84 mm/h, C-reactive protein: 91 mg/L, hemoglobin: 8.4 g/dl, blood urea nitrogen: 32 mg/dl (6-20 mg/dl), creatinine: 7.2 mg/dl (0.6-1.3 mg/dl), albumin: 2.6 g/dl (3.4-5 g/dl), total protein concentration: 5.4 mg/dl (6.4-8.3 mg/dl), and urinary protein excretion: 11 g/24 h.

A percutaneous renal biopsy showed deposition of amyloid by gentian violet in the glomeruli and walls of the blood vessels. Immunohistochemically, the glomeruli and walls of the blood vessels showed cytoplasmic reactivity for P component and amyloid A (Figure 1). The case was diagnosed as renal amyloidosis and interpreted as consistent with reactive systemic (secondary) amyloidosis.

DNA sequence analysis showed heterozygous M694V mutation on exon 10. Upper and lower gastrointestinal endoscopies were performed for prolonged diarrhea, and prepyloric ulcers on the duodenum were detected. Ileal biopsy also showed amyloid deposition in the submucosal blood vessels.

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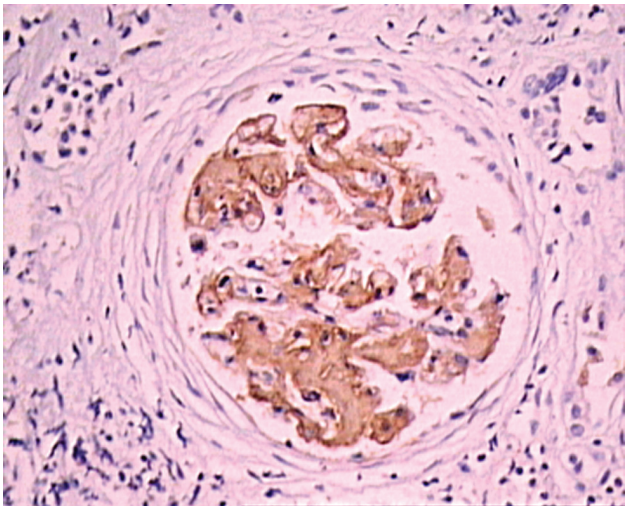

Figure 1. The glomerulus demonstrating cytoplasmic reactivity for amyloid A (Diaminobenzidine, x400).

Intestinal involvement constitutes the major gastrointestinal localization of BD. Colonic or ileo-colonic lesions may present in the form of acute complications (perforation, massive hemorrhage) or by prolonged diarrhea (4).

Genetic and environmental factors play a role in the pathogenesis of amyloidosis, such as MEFV mutations and serum amyloid A(SAA) protein gene polymorphism. M694V, M694I, V726A, M680I, and E148Q are the most frequent MEFV mutations (5).

Eprodisate, a specific drug for secondary amyloidosis, has been developed and evaluated, though it has not yet been approved by the Food and Drug Administration (FDA) (6).

In patients with BD and prolonged diarrhea, systemic amyloidosis should be kept in mind, although it rarely occurs. Coexistence of a genetic mutation such as in the MEFV gene may predispose patients to develop amyloidosis. More studies are needed to clarify the role of MEFV mutations in BD-related amyloidosis.

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Cancer of the transverse colon revealed by a gastrocolic fistula

Gastro-kolik fistül ile prezente olan transvers kolon kanseri

To the Editor,

Gastrocolic fistula (GCF) secondary to colon carcinoma is a rare entity. Establishing the diagnosis of GCF is difficult because it has nonspecific symptoms on admission. We report a case of GCF due to adenocarcinoma of the transverse colon who had only a one-month history of nausea, vomiting and weight loss.

A 50-year-old man admitted to our hospital complaining of nausea, vomiting and weight loss. Labo-

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