Sirs,

Although familial Mediterranean fever (FMF) patients with nephrotic syndrome (NS) receive colchicine regularly, one-third of them do not sufficiently benefit from colchicine and all FMF patients with chronic renal insufficiency and NS progress to end-stage renal failure (ESRF) (1). There is not sufficient knowledge in the literature about whether or not progression of NS to ESRF could be halted through a choice other than colchicine. I herewith report 3 FMF cases with nephrotic range proteinuria (NRP) who responded well to immunosuppressive drugs.

The cases fulfilled the criteria for the FMF described previously (2). The presence of amyloidosis was determined by the immunohistochemistry method. Though NS was caused by AA amyloidosis, the initial treatment was with colchicine and enalapril. Later, and after progression of NS to NRP, immunosuppressive drugs were added. As Table I shows, there were clear differences in the laboratory findings of the patients throughout the follow-up period.

Table I. Laboratory data during the follow-up period (mean ± SD).

<table>
<thead>
<tr>
<th>Months</th>
<th>Laboratory findings</th>
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<td>0-6</td>
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<td>6-12</td>
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<td>12-18</td>
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<td>24-30</td>
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**Case 1**

- Proteinuria (g/day): 4.8, 4.6 ± 0.5 (4-5), 4.2 ± 0.2 (4-4.5)*, 2 ± 0.5 (1.5 – 2.5), 0.8 ± 0.3 (0.3 – 1.2), 0.65 (0.6 – 0.7), 0.6 ± 0.4 (0.3 – 1).
- Albumin (g/dl): 2.6, 2.2 ± 0.8 (1.4 – 3), 2.9 ± 0.1 (2.8 – 3), 3.4 ± 0.1 (3 – 3.6), 3.7 (3.7 – 3.9), 4.3 (4.3 – 4.3), 3.8 ± 0.2 (3.7 – 4).
- Creatinine (mg/dl): 0.8, 0.5 (0.4 – 0.6), 0.3 (0.2 – 0.4), 0.35 ± 0.2 (0.2 – 0.4), 0.45 (0.4 – 0.5), 0.5 (0.5 – 0.7), 0.56 (0.5 – 0.62).
- ESR (mm/h): 95, 107 ± 1 (93 – 119), 92 ± 16 (81 – 104), 67 ± 7.5 (60 – 75), 50 ± 8 (38 – 61), 41 ± 1.4 (40 – 42), 36 ± 8 (30 – 42).
- CRP (mg/dl): 2.5, 2.4 ± 0.3 (2.1 – 2.8), not tested, 0.76 ± 0.5 (0.2 – 1.2), 0.3 ± 0.2 (0.1 – 0.6), 0.15 (0.1 – 0.2), 0.2 ± 0.1 (0.1 – 0.3).

**Case 2**

- Proteinuria (g/day): 7, 5.9 ± 0.2 (5.8 – 6.1), 4.1 ± 0.8 (3.2 – 5), 1.1 ± 0.1 (1 – 1.2), 0.8 (0.75 – 0.85), 0.7 ± 0.1 (0.6 – 0.8), 0.6 (0.6 – 0.7).
- Albumin (g/dl): 2.5, 2.6 ± 0.1 (2.5 – 2.7), 3.3 ± 0.2 (3.2 – 3.5), 3.7 (3.7 – 3.8), 3.8 ± 0.2 (3.7 – 3.9), 4.0 ± 0.3 (4 – 4.1), 5.4 ± 0.5 (5 – 5.8).
- Creatinine (mg/dl): 0.9, 0.5 (0.52 – 0.5), 0.58 (0.54 – 0.6), 0.45 (0.4 – 0.49), 0.6 (0.5 – 0.7), 0.65 (0.6 – 0.7), 0.7 (0.62 – 0.8).
- CRP (mg/dl): 1.2, 1.5 ± 0.1 (1.4 – 1.6), not tested, 0.7 ± 0.1 (0.6 – 0.8), 0.9 ± 0.8 (0.3 – 1.5), 0.8 ± 0.1 (0.7 – 0.9), 0.65 (0.6 – 0.7).

**Case 3**

- Proteinuria (g/day): 7.2*, 4.7 ± 1.5 (3 – 6.2), 2.7 ± 1.2 (1.4 – 2.4), 0.7 ± 0.2 (0.4 – 1.1), 0.55 (0.5 – 0.6).
- Albumin (g/dl): 2.7, 3.1 ± 0.2 (2.9 – 3.4), 3.8 ± 0.3 (3.5 – 4.3), 4.2 ± 0.1 (4.1 – 4.3), 4.8 ± 0.1 (4.7 – 4.9).
- Creatinine (mg/dl): 1.3, 1.2 (1.1 – 1.3), 1.3 (1.29 – 1.39), 1.5 (1.4 – 1.6), 1.4 (0.8 – 1.6).
- CRP (mg/dl): 1.4, 1.2 ± 0.3 (0.8 – 1.4), 0.8 ± 0.7 (0.2 – 1.9), 0.3 ± 0.1 (0.2 – 0.6), 0.5 ± 0.2 (0.4 – 0.7).

Case 3 was monitored for 24 months; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein; nt: not tested. Normal levels: albumin: 3.5 – 5.5 g/dl; Creatinine: 0.5 – 1.6 mg/dl; ESR < 20 mm/h; CRP < 0.8 mg/dl. Values in parentheses give the range; *refers to the point at which immunosuppressive treatment was commenced. Creatinine levels were shown as the mean (range).
added to his treatment, as with Case 1. Case 3 (37/M) referred to us for cutaneous vasculitis, confirmed by the skin biopsy. He had a history of recurrent fever, abdominal and chest pain, and erythematous-like erythema since he was 3. He was diagnosed with FMF at the age of 27 and colchicine was administered at the dosage of 0.5-1 mg/day. Since case 3 had a cutaneous vasculitis, I administered PRD and AZA in the aforementioned dosages in addition to colchicine (2 mg a day), enalapril maleat (10-20 mg/day) immediately after the diagnosis of AA amyloidosis and vasculitis. The dosage of PRD and AZA was adjusted as in cases 1 and 2.

Letters to the Editor

C. KORKMAZ
Associate Professor Division of Rheumatology, Department of Internal Medicine, Osmangazi University Medical Faculty, Eskisehir, Turkey.
Address correspondence to: Cengiz Korkmaz, MD, Vissalek M, A箕asit Gisme C, Akaya S, 11/11, Eskisehir, Turkey.
E-mail: ckorkmaz@ogu.edu.tr

References

Behcet’s disease associated with Trisomy 8 in a male Caucasian patient from Great Britain – A case report

Sirs,
We would like to report a 54-year-old Caucasian male from Great Britain who was referred to our Rheumatology unit with a history of increasing tiredness associated with polyarthritis, night sweats and recurrent punched out orogenital ulcerations. Further questioning revealed that he had suffered from arthralgia and occasional joint stiffness during attacks. At the time of presentation, his symptoms included a 10 kg weight loss during the previous 6 months, and symptoms of orogenital ulcerations. He was diagnosed with Behcet’s disease with a neutropenic bone marrow and raised ESR of 52 mm in 1 h. Treatment with steroids was followed by an improvement in the patient’s condition, and he was referred to our haematology service for further investigation.

We initially performed a bone marrow biopsy which showed dysplastic and hypoplastic areas, with a significant number of metaphases. In view of his persistent neutropenia, we performed a bone marrow biopsy which showed hypoplastic marrow with adequate number of megakaryocytes. Some of them showed dysplastic changes. The presence of dysplastic changes was confirmed by immunostaining for CD117 and CD34. The patient was also referred to the ophthalmology service for funduscopy, which showed signs of retinal vein occlusion.

We also performed a bone marrow biopsy which showed dysplastic and hypoplastic areas, with a significant number of metaphases. Further questioning revealed that he had suffered from arthralgia and occasional joint stiffness during attacks. At the time of presentation, his symptoms included a 10 kg weight loss during the previous 6 months, and symptoms of orogenital ulcerations. He was diagnosed with Behcet’s disease with a neutropenic bone marrow and raised ESR of 52 mm in 1 h. Treatment with steroids was followed by an improvement in the patient’s condition, and he was referred to our haematology service for further investigation.

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