

## Case 4: Polyarteritis Nodosa (PAN)

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**F**orty-four years old, male, familial Mediterranean fever (FMF) patient has appointed to the medical unit with the complaints of pain, cyanosis and tissue loss in the 3<sup>rd</sup> and 5<sup>th</sup> toes of left foot and 2<sup>nd</sup> toe of the right foot starting three days ago. He also suffers from increasing abdominal pain, widespread muscle pain and numbness in his left foot for three weeks before being admitted to hospital.

He has fever (38.2 C) on his first visit and his blood pressure was measured as 155/95 mm-hg. Additionally, in his first evaluation, there was widespread abdominal tenderness, allodynia in his left lower extremity and cyanosis and gangrene in the 3<sup>rd</sup> and 5<sup>th</sup> toes of left foot and 2<sup>nd</sup> toe of the right foot.

Laboratory evaluation revealed normochromic normocytic anemia, leucocytosis, increased levels of transaminases and cholestatic enzymes. Creatinine level has increased from 0.8 mg/dl to 1.8 mg/dl. Furthermore, although urinary sediment was normal, 24 hour urine protein level was found to be 480 mg/day. Then, we have ordered computed tomographic angiography to evaluate the circulatory problems in the distal extremity and abdominal pain possibly related to mesenteric ischemia. There were occlusions in inferior mesenteric, left anterior and posterior tibial and right anterior tibial arteries in tomographic angiography. We also performed biopsy next to the gangrenous part of the 3<sup>rd</sup> toe of the left foot. The pathologic examination revealed medium-small vasculitis with segmental and transmural necrosis. Additionally, sensorial neuropathy was observed in the left lower extremity on electromyography.

With these findings, he was diagnosed with polyarteritis nodosa. We then administered 1 gram/day prednisolone for three consecutive days, followed by 1 mg/kg/day prednisone equivalent steroid dose. Later, we started with 1 gram cyclophosphamide per month as an immunosuppressant. In addition, we applied debridement and amputation to the gangrenous part of the toes.

Vasculitis indices at the first admission:

### Birmingham Vasculitis Activity Score (BVAS)

1. Section (General) : 3 point

- Myalgia
- Fever $\geq$ 38

2. Section (Skin): 6 point

- Gangrene

7. Section (Abdominal): 6 point

- Ischemic abdominal pain

8. Section (Renal): 12 point

- Hypertension
- Proteinuria  $>1$  vs  $>0.2$  mg/24 hour
- Serum creatinine 1.41-2.82 mg/dl
- Increase in serum creatinine  $>30\%$  or decrease in creatinine clearance  $>25\%$

9. Section (Nervous System): 6 point

- Peripheral sensorial neuropathy

Total: 33 point

### Five Factor Score

Kartal Dr. Lütfi Kırdar  
Şehir Hastanesi  
Romatoloji Kliniği,  
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Gastro-intestinal involvement  
Renal involvement (Cr  $>1.69$  mg/dl)  
Total: 2 point

**Disease Extend Index**

Gastrointestinal system: 2 point  
Gastro-intestinal involvement in angiography  
Renal: 2 point  
Serum creatinine level  
Peripheral nerve system: 2 point  
EMG findings were related to sensorial neuropathy  
Skin: 2 point  
Digital gangrenes  
Arthritis/Arthralgia: 2 point  
Myalgia  
Constitutional symptoms: 1 point  
Fever>38 C  
Total: 11 point

At the third month visit, there was numbness in the left extremity. He had no complaints of fever, abdominal pain or musculoskeletal system. In addition, no circulation problems were observed in the extremities. Moreover, liver and kidney functional tests and all laboratory test including acute phase reactants were at normal levels. There was also no cytopenia in the hemogram. In addition, protein level in 24 hour urine was reduce to 250 mg/day. All examinations were normal except for diffuse allodynia in the left lower extremity and amputation scars on the toes. At the end of the evaluation we advised him to receive both cylophosphamide 1 gr per month and daily oral steroid therapy. We also diagnosed avascular necrosis in the left hip with the complaint of left groin pain.

Vasculitis indices at the third month of the treatment

**Vasculitis Damage Index**

Musculoskeletal: 1 point  
Avascular necrosis  
Peripheral vascular disease: 1 point  
Major tissue loss  
Neuropsychiatric: 1 point  
Peripheral neuropathy  
Total: 3 point