Adrenal Insufficiency in a Patient with End Stage Renal Disease due to Secondary Amyloidosis

Sekonder Amiloidoza Bağlı Son Dönem Böbrek Hastalıklı Bir Olguda Adrenal Yetmezlik

ABSTRACT

Amyloidosis is a group of disease characterized by the extracellular deposition of the fibrils called amloid in various organs. Progressive organ dysfunction is seen in the systemic form. It may infiltrate various endocrine glands but rarely causes endocrine dysfunction. In this report, we describe a case of symptomatic adrenal failure complicated with infection in a patient with end-stage kidney disease due to secondary amyloidosis.

KEY WORDS: Addison disease, Adrenal failure, Amyloidosis

ÖZ


ANAHTAR SÖZCÜKLER: Addison hastalığı, Adrenal yetmezlik, Amiloidoz

INTRODUCTION

The amyloidoses comprise a heterogeneous group of diseases in which proteins aggregates into characteristic fibrils with some unique properties (1). The prevalence of the disease varies in the world. In the developing countries, secondary amyloidosis (AA amyloid) is more frequent than primary amyloidosis (AL type), because of the higher burden of chronic infectious diseases such as tuberculosis and osteomyelitis. In some parts of the world, familial Mediterranean fever, a genetic disorder, accounts for a major part of secondary amyloidoses. Secondary amyloidosis is seen more frequently in our country than many others owing to the high frequency of familial Mediterranean fever and tuberculosis. In some kidney biopsy reports, it has been documented to be as high as 12% (2).

The kidney is the most commonly affected organ in patients with secondary amyloidosis. In clinical practice, renal involvement most often presents as proteinuria or nephrotic syndrome. However, primary amyloid deposition can be limited to the vessels or tubules and such patients present with progressive renal failure. Apart from the kidney, the liver, spleen and heart are frequently involved in systemic amyloidosis. Amyloid infiltration may be seen in endocrine glands but it does not always cause dysfunction. Primary adrenal insufficiency rarely occurs in amyloidosis, but if it occurs, life-threatening complications may develop. In this report, we describe a patient that presented as symptomatic adrenal failure due to amyloidosis and was on a regular hemodialysis programme.

CASE REPORT

A 52-year-old man on a regular hemodialysis programme due to end-stage renal disease presented to emergency department of our hospital with fever and generalized sei-
Glomerular amyloid deposition, light microscopy with Congo-red x40.

Table I: Results of Corticotropin stimulation test (250 mcg synachten, im).

<table>
<thead>
<tr>
<th>Test Duration</th>
<th>Serum Cortisol (μg/dL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-minute</td>
<td>7.57</td>
</tr>
<tr>
<td>30-minute</td>
<td>8.94</td>
</tr>
<tr>
<td>60-minute</td>
<td>10.79</td>
</tr>
<tr>
<td>6-hour</td>
<td>9.36</td>
</tr>
<tr>
<td>24-hour</td>
<td>7.13</td>
</tr>
</tbody>
</table>

Figure 1: Glomerular amyloid deposition, light microscopy with Congo-red x40.

Diagnosis of the adrenal failure is very critical to prevent life-threatening complications. Some symptoms and signs of the adrenal failure (such as nausea, vomiting, metabolic acidosis, hyperkalemia and hypotension) may be ascribed to uremia and autonomic neuropathy which can be seen in systemic amyloidosis. End stage renal disease due to secondary amyloidosis is reported 11% at the time of the diagnosis (9). Progression to end stage renal disease is reported nearly 23%, changing with some risk factors (9). The frequency of the adrenal involvement in the patients with end-stage renal disease due to secondary systemic amyloidosis is not known exactly. The diagnosis of the adrenal failure in these patients can be very difficult because of the high frequency of hypotension and the masking effect of hemodialysis on the electrolyte and metabolic disturbances. Furthermore, low cortisol response to corticotropin without clinically evident adrenal failure was observed in nearly 50% of the patients with systemic amyloidosis (12-14). In these studies, some of the patients had end-stage renal disease. Although a simple test...
can be enough for the diagnosis of adrenal failure, this is not the case in end-stage kidney disease patients with secondary amyloidosis. Thus, clinicians should consider both clinical and laboratory findings carefully to diagnose adrenal failure in this group of patients. Perhaps, it is more important to remember the disease.

In secondary systemic amyloidosis, abnormal response to corticotropin represents a dilemma as to whether adrenal failure exists or not unless symptoms or signs related with disease are present. Hypotension and hypoglycemia are important clues for suspicion for adrenal failure. The patient in this case had no frequent hypotension episodes during hemodialysis seasons. It may be speculated that our patient already had subclinical adrenal failure due to adrenal amyloid infiltration, and symptoms developed with a stress factor, infection, causing decrement of adrenal reserve to a critical level.

In our country, Familial Mediterranean fever (FMF) is the leading cause of secondary amyloidosis (15,16). Connective tissue disorders and chronic lung diseases are other common etiological causes (16,17). Control of underlying inflammatory disorders can result in prevention and regression of secondary amyloidosis (1). Colchicine is an effective treatment for FMF both in preventing amyloid formation and regression of amyloid deposition, especially when started early in the course of the disease (1,17,18).

In conclusion, we presented a patient with secondary systemic amyloidosis who developed adrenal failure, a clinical condition rarely reported in the literature. The endocrine system is frequently involved in systemic amyloidosis, but it rarely causes endocrine dysfunction. Clues for adrenal failure can be masked with hemodialysis in patients with end-stage renal disease. As in the present case, adrenal failure should be kept in mind in all patients with systemic amyloidosis, and special attention and evaluation should be done to prevent untoward results.

**REFERENCES**