# Son Dönem Böbrek Yetmezliğinin Eşlik Ettiği Ainhum Hastalığı

## *Ainhum Disease Complicated by End Stage Renal Failure*

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**ÖZET**

Ainhum hastalığı beşinci ayak başparmağının otoampütasyonuyla karakterize bir hastalıktır. Ainhum hastalığına birçok tıbbi sorun eşlik edebilmektedir. Daha öncesinde bu az rastlanan hastalığa son dönem böbrek yetmezliğinin eşlik ettiği bir vaka bildirilmemiştir. Bu yazıda, son dönem böbrek yetmezliği gelişen 18 yaşındaki ainhum hastası bir bayan olgu sunulmaktadır.

**Anahtar sözcükler**: böbrek yetmezliği, ainhum hastalığı, otoampütasyon

**ABSTRACT**

Ainhum disease is characterized by the spontaneous autoamputation of the fifth toe. In ainhum disease there are many medical problems that might accompany this unknown disease. Ainhum disease associated with end stage renal failure was not reported before. A case of an 18-year- old female patient with ainhum disease who developed end stage renal failure was reported here.

**Keywords**: renal failure, ainhum disease, otoamputation

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### Introduction

Ainhum disease is characterized by the spontaneous autoamputation of the fifth toe after the occurence of a circular constriction located at the root of the toe. This progresive disease is not a well understood disease where its incidence is highest in tropical and subtropical climates especially in black population with a special predilection for middle-aged males in their fourth and fifth decades. Many medical diseases may complicate to ainhum disease. However, to date, no renal failure associated with this disease had been reported.

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### Case Report

On July 2004, an 18-year-old female patient was admitted to our clinic with complaints of paleness, weakness, nausea, vomiting and disuria. Medical history of the patient revealed with hyperemia in her fifth toe ten years before, then constriction and at last spontaneous and painless autoamputation. After a period of time, she had the same manifest deformities for her other toes and fingers. Her younger brother had some limited fissures only in his toes resembling the lesions of our patient's. In her physical examination arterial blood pressure was 120/70 mmHg and she had subfebril fever. Autoamputated toes and fingers, growth retardation and short stature were noticed. She had the face of an old female and her left foot was hyperkeratotic, thickened, brownish, fissured and infected in addition to the autoamputated toes (Figure 1,2,3,4,5,6). Urinanalysis revealed positive dipstick reaction for protein and blood. Spesific gravity and pH were 1020

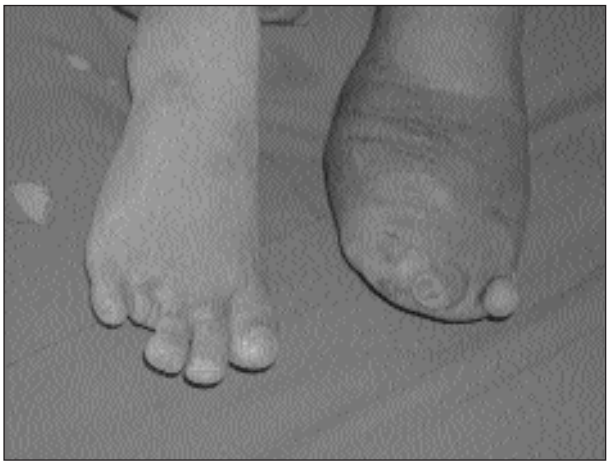

**Figure 1.**

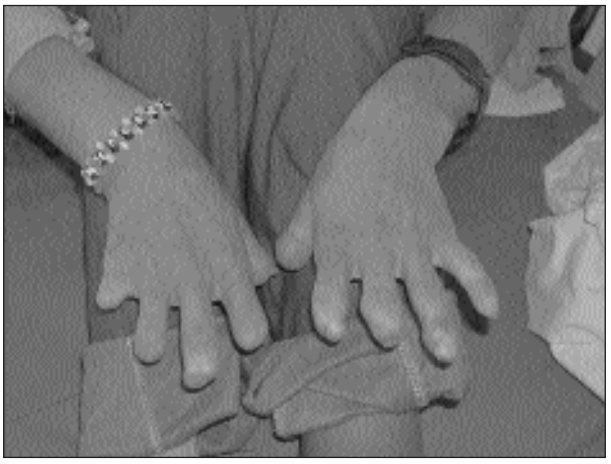

**Figure 2.**

**Figure 3.**

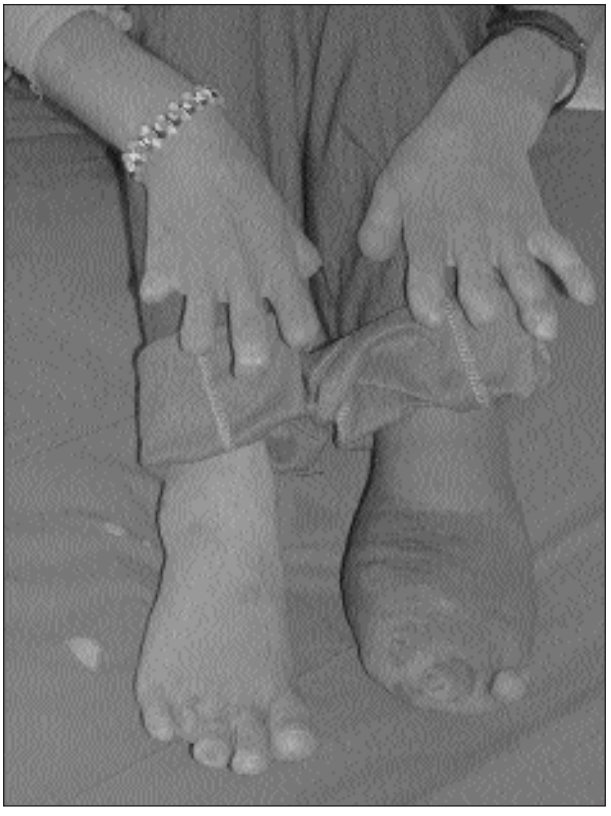

**Figure 4.**

and 6.0 respectively. The sediment of urine contained 2-3 epithelia, 10-12 leucocytes and 18-20 eritrocytes. In the 24-hour collected urine 0.26 gr protein was found. Peripheric blood count showed 26.500 leucocytes/$mm^3$, platelets of 894.000/$mm^3$, haemoglobin of 9.6 g/dL. Biochemical tests results were as follows: glucose 72 mg/dL, sodium 124 mmol/L, potassium 2.7 mg/dL, chloride 103 mmol/L, calcium 71 mg/dL, urea 42 mg/dL, creatinine 1.4 mg/dL, creatine phosphokinase (CK) 21 U/L (normal: 0-170), CK-MB 13 U/L (normal: 0-25), lactate dehydrogenase 530 U/L (normal: 220-450), aspartate aminotransferase (AST) 35 U/L (normal: 10-37), alanine aminotransferase (ALT) 21 U/L (normal: 10-37), albumin: 1.01 gr/dL, globulin: 2.06 gr/dL. Erithrocyte sedimentation rate was 61 mm/h, C-re-

**Figure 5.**

**Figure 6.**

active protein was 78 mg/dL (normal: 0.0-8.0 mg/dL). Prothrombin time (INR: 6.5) and aPTT were 80 and 58 respectively. Immunological studies including anti-nuclear antibody, anti- ds DNA, anti-scl70 and rheumatoid factor were either negative or with in the normal ranges. Nontreponemal reaginic test for syphilis was negative. Protein electrophoresis and the other laboratory analysis were in normal range. Levofloxacine sensitive E. coli was detected in urine culture. No pathology was found in audiography as well as in her eye and ear-nose-throat examination. The chest x-ray and thorax computerized tomography (CT) were normal except bilateral pleural effusion and vascular congestion. In abdominal ultrasonography (USG) the size of the kidneys were increased and grade 2 echogenity of the renal paranchyma in addition to intraabdominal effusion were detected which were also seen in the abdominal CT. Renal doppler USG demostrated no vacular resistance and echocardiogarphy showed only mild pulmonary hypertension with pulmonary arterial pressure of 35-40 mmHg. Upper gastrointestinal endoscopy revealed grade C esophagitis which was the reason of patient's intractable vomiting causing electrolyte imbalance. After oral flucanazol therapy was applied, this symptom was improved. We applied 7 units of erithrocyte suspension for her chronic illness anemia and 5 units of fresh-frozen plasma due to her high level of prothrombin time. Nearly three months after her admission, progresive increase in the urea and creatinin levels were established. Urea increased to 136 mg/dL, creatinine increased to 7.1 mg/dL. The glomerüler filtration rate was decreased to 8 ml/minute and 24 hours urine volume decreased to 500ml. So, we began hemodialysis. In control renal USG, we detected grade 2-3 echogenity of the renal parenchyma which were increased in size in contrast to usual expectation. No amyloid deposition was detected both in rectal and abdominal subcutaneous biopsies. After the beginning of hemodialysis and erithropoietin therapy, üremic symptoms were cured completely.

## Discussion

Ainhum disease is often associated with the hyperkeratosis of the skin, deformity of the nails and the pallor of the affected digits. Pain is especially pronounced after a pathologic fracture and is usually the presenting symptom causing the patient to seek medical attention. Abduction and hyperextension deformity of the toe, with loss of voluntary movement, may follow marked underlying bone involvement. The end result of untreated ainhum is often auto-amputation of the toe, with the stump of the proximal phalanx remaining intact with its metatarsophalangeal joint. Surgical amputation may be required in the later stages of ainhum when pain persists, even after autoamputation, because the

remnant of the proximal phalax sometimes protrudes through the skin and causes ulceration. It is well to remember that ainhum usually occurs in otherwise healty individuals, and what happens to be a trival lesion often causes prolonged and severe crippling disability. We differantiated ainhum disease from other similar diseases like leprosy, syphilis, tuberculosis, yaws, keratodermas, connective tissue diseases, congenital connecting bands and traumatic lesions by the lesions' typical appearrance and their clinical stages. No mental retardation was present. At present, the exact cause of ainhum remains a mystery, but two undisputed factors associated with its development are race and prolonged walking on bare foot. The fibroblastic diathesis or abnormal fibrogenesis commonly noted in the African, especially with regard to keloid formation, may be an associated factor. Mechanical rotation, strain of the digit, recurrent chronic fissuring and perhaps altered endarterial vascular supply or angiodisplasia may all play a vital role in its etiology. In our patient, only walking on bare foot was present from the probable causes. The most frequent causes of medical diseases that may complicate to ainhum disease were hearing problems, prone to infections maybe due to immunological disorders and growth retardation. However, no relation between ainhum disease and renal failure was defined before. In the presented case, at her first admission to hospital while we were examining the patient in our clinic for the other medical problems that may complicate this mysteries disease, we found limited failure of renal functions with high parenchymal echogenity. End stage renal failure was determined after the close follow-up of her clinical status. The interesting point was the increased in sizes of her kidneys in contrast to what the clinician expected in chronic renal failure (CRF). As we know, there are some exceptions in CRF with increased renal sizes like amiloidozis, diabetic nephropathy, hidronephrosis, policystic renal disease and infiltrative diseases of the kidney. Our patient was not diabetic, no hidronephrosis and policysts were found in imaging modalities and no sign of infiltration of the kidney was detected. As a result, main possible causes of increased renal sizes in CRF except amyloidozis were eliminated by the patient's history and imaging modalities. In order to find out whether amyloidozis was the cause of CRF or not, we did rectal and abdominal subcutaneous biopsies. No amyloid deposition was found out in these biopsies with the congo red and crystal violet histochemical staining. Renal biopsy was thought to be unnecessary owing to grade 2-3 echogenity of the parenchyma and risky because of the high level of prothrombin time.

## Conclusion

In conclusion, the relation between renal failure and a patient with ainhum disease was presented in this report. Depending on these findings, ainhum disease should be considered as one of the causes of chronic renal failure particularly with increased in their sizes.

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