End-Stage Renal Disease in a Patient with Werner's Syndrome

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Dear Sir,

Werner's syndrome is a rare autosomal recessive disorder characterized by premature aging. The clinical feature of this syndrome includes short stature, premature cataracts, skin atrophy, osteoporosis, graying and loss of hair, neoplasia, diabetes mellitus and atherosclerosis [1]. Kidneys with gross impairment are rarely involved and, as far as we know, only 1 case with widespread vascular disease has been reported to die in uremia [2]. We herein present a case of Werner's syndrome complicated by end-stage renal disease (ESRD) maintained on hemodialysis.

A 51-year-old male was referred to our hospital because of chronic renal failure. He had a medical history of bilateral cataract extractions at the age of 37, established proteinuria at the age of 44, partial thyroidectomy due to a thyroid carcinoma at the age of 46, intracranial meningioma removed at the age of 49 and had been on a diet for mild diabetes mellitus for the last 7 years. Consanguineous marriage had allegedly been done for generations in his family and his parents were cousins. His height was 149 cm, weight 39 kg and blood pressure 150/70 mm Hg. He had a squeaky hoarse voice, very sparse and gray hair on his scalp, extremely slender extremities and scleroderma-like alterations of the skin. Vascular calcification of the lower extremities was noted radiologically. A diagnosis of Werner's syndrome was made. Laboratory studies

showed: hemoglobin 8.7 g/dl, hematocrit 25.4%, white blood cell count 7,200/ μ l, platelets 27.6×10^4 / μ l, blood urea nitrogen 84 mg/dl, serum creatinine 11.9 mg/dl, sodium 140mEq/l, potassium 3.6 mEq/l, calcium 6.4 mg/dl, phosphorus 8.4 mg/dl, total blood proteins 5.5 g/dl, albumin 3.1 g/dl, blood glucose 84 mg/dl, hemoglobin A_{1c} 6.6%, creatinine clearance 6 ml/min. Urinalysis showed nephrotic proteinuria (7.4 g/24 h) and urinary sugar (2.0g/24h) with a normal sediment. An ultrasonographic examination showed contracted kidneys (7.2, 6.5 cm) with decreased visualization of the corticomedullary junction. Ophthalmoscopy showed mild arteriolar changes

but no evidence of diabetic retinopathy. The renal function continued to worsen and uremic symptoms appeared. The hemodialysis treatment was initiated in our hospital and has been maintained on an outpatient basis for 4 months until this day.

Diabetes mellitus in Werner's syndrome, reportedly appearing in 44-45% of all cases, is usually mild and rarely develops diabetic nephropathy [1,3]. Although the exact cause of ESRD with nephrotic proteinuria in our patient was unknown, as a renal biopsy had not been performed, absence of diabetic retinopathy considerably raised the possibility of nondiabetic glomerulopathies as the cause [4]. It has also been reported that the kidneys in diabetic subjects for any degree of renal failure are larger than nondiabetic kidneys [5, 6], whereas the kidneys in our patient were small. Again, we speculate that ESRD in our patient might be attributable to a nondiabetic glomerulopathy. To our knowledge, this is the first case of ESRD with Werner's syndrome maintained on hemodialysis.

References

Epstein CJ, Martin GM, Schultz AL, Motulsky AG: Werner's syndrome: A review of its symptomatology, natural history, pathologic features, genetics and relationship to the natural aging process. *Medicine* 1966;45:177-221.

Valero A, Gellei B: Retinitis pigmentosa hypertension and uraemia in Werner's syndrome. *Br Med J* 1969;ii:351.

Goto M, Tanimoto K, Horiuchi Y, Sasazuki T: Family analysis of Werner's syndrome: A survey of 42 Japanese families with a review of the literature. *Clin Genet* 1981;19:8-15.

Parving HH, Gall MA, Skott P, Jorgensen HE, Lokkegaard H, Jorgensen F, Nielsen B, Larsen S: Prevalence and causes of albuminuria in non-insulin-dependent diabetes patients. *Kidney Int* 1992;41:758-762.

Wirta OR, Pasternack AI: Glomerular filtration rate and kidney size in type 2 (non-insulin-dependent) diabetes mellitus. *Clin Nephrol* 1995;44:1-7.

Wirta OR, Pasternack AI, Laippala P, Turjanmaa V: Glomerular filtration rate and kidney size after six years disease duration in non-insulin-dependent diabetic subjects. *Clin Nephrol* 1996;45:10-17.

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