**Introduction:** Renal amyloidosis may lead to renal disease, and then nephrotic syndrome may develop. This is the first case report in which a patient presents with generalized aggressive periodontitis and nephrotic syndrome in conjunction with renal amyloidosis.

**Case presentation:** An 18-year-old male applied to the Periodontology department for his generalized gingival recessions. He was diagnosed as primary renal amyloidosis by his physician. The patient presented with severe gingival inflammation and alveolar bone loss. Biochemical tests were within normal limits except for serum albumin level. No amyloid deposition was found in a gingival biopsy, and the patient was diagnosed as generalized aggressive periodontitis. Non-surgical periodontal treatment, in combination with antibiotic treatment, was performed. In the 3rd year follow-up period, his systemic and periodontal conditions showed deteriorations.

**Conclusion:** The effects of systemic factors related to nephrotic syndrome in conjunction with renal amyloidosis, and deterioration in oral hygiene may play a significant role in the progression of periodontal disease. Even if there is no amyloid deposition in periodontal tissues, clinicians should consider that nephrotic syndrome associated with systemic amyloidosis may provide an important contribution to the periodontal destruction by the modifying conditions which affect the host response to the accumulation of dental biofilm in affected patients.