Secondary Amyloidosis with Renal Involvement in an Adult Patient with Cystic Fibrosis

Amiloidosis secundaria con afectación renal en paciente adulto con fibrosis quística

To the Editor:

Cystic fibrosis (CF) is a genetic disease from which survival has been increasing steadily over recent decades. Other complications associated with CF have also been increasing, such as secondary amyloidosis, which is associated with chronic inflammatory processes. Amyloidosis is a systemic disease characterised by the extracellular deposition of fibrillar proteins. Secondary AA amyloidosis consists of fibrils of protein A, an acute phase reactant produced by hepatocytes. Renal involvement is common in this condition. Secondary AA amyloidosis is a recognised complication of CF (mainly in patients with a long evolution of the disease and poor disease control), but very rare. Its incidence is not known in CF and it is associated with poor prognosis. In most cases it presents with proteinuria, thyromegaly, and/or hepatosplenomegaly. Furthermore, amyloidosis with renal involvement is frequent and evolves into kidney failure in a relatively short time (months or years), which is associated with a poor prognosis.

Concluding the recommendation, in this case we wish to show that a prolonged treatment with a combination of inhaled and systemic corticosteroids and hydroxychloroquine achieved an appropriate response in the case we have presented. The appearance of new episodes of haemoptisis when prednisone was first decreased prolonged the duration of treatment. The good response seen in our patient, although this was an isolated case, is an indication that it is possible to consider using this combined therapy for IPH.

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None.

Conflict of Interest

The authors affirm that they have no conflict of interest.

Appendix 1

Chest CAT: Multiple bilateral diffuse infiltrates, some of pseudonodular morphology, with ground glass areas and small sized hilar adenopathies.

References

We report the case of a 32 year old man, diagnosed with CF late when he was 20 years old (he consulted the doctors after his brother was diagnosed) and with: DF508/R334W genotype, bronchiectasis with chronic bronchial infection by Pseudomonas aeruginosa; pancreatic insufficiency; obstructive azoospermia; moderate malnutrition. A smoker of 15 packs/ year, occasional drug addict through inhalation, and social alcoholism. Stable up to 27 years old, despite irregular compliance with treatment and monitoring in the Unidad de FQ de Adultos de Málaga (Adult CF unit, Malaga), with 1-2 mild respiratory exacerbations per year. At that age, he was diagnosed with adjustment disorder with anxiety and depression and suicidal ideas, and from then on suffered progressive deterioration and 7 hospital admissions for severe exacerbations and persistent poor adherence to treatment. Depression-anxiety in chronic patients is related to the acquisition of poor health habits and a worse prognosis, and therefore diagnosis is crucial. The patient was admitted to our unit (months after abandoning follow-up) due to another episode of respiratory infection and with a clinical picture of periborial and lower limb oedema which had developed over several months with normal kidney function. The most important results were proteinuria of 7g/day, microalbuminuria 1172 mg/l, abdominal and thyroid ultrasound without alterations, chest HRCT with varicose bronchiectasis in LLL and cylindrical in RUL, and breathing pattern with moderate obstruction. As a result of these findings, a kidney biopsy was performed which showed: glomeruli with focal depositions of hyaline material; Congo red positive; positive staining for AA amyloid (figs. 1 and 2). The patient developed favourably and was discharged with diuretics and ACE inhibitors along with his basic treatment and a consultation programme in the CF, nephrology and mental health unit. After 10 months, he was re-admitted to the unit due to a new respiratory exacerbation, with multiple complications during hospitalisation (haemoptysis, acute pancreatitis, among others), and finally developed acute kidney failure which required haemodialysis (rejected for kidney transplant) with subsequent death due to multiple organ failure 60 days after admission (12 months after diagnosis of amyloidosis with renal involvement).

Secondary amyloidosis should be suspected in subjects with longstanding or poorly controlled CF (as was the case with our patient: with late diagnosis, poor adherence to treatment, alcoholism, and depressive syndrome), who have proteinuria, oedema, hepatosplenomegaly, and/or thyroid problems. Early diagnosis is important to make the necessary therapeutic adjustments with a view to kidney transplantation. In patients with CF, the transplantation of the lungs and kidney can be assessed. Annual microalbuminuria testing should be performed as a screening test, although a kidney biopsy is required to confirm the diagnosis. Iodine-123-labelled serum amyloid P component scintigraphy is also useful for diagnosis and to determine the location and extent of the amyloid deposits, although this is not available in most centres.

To date, no effective treatment is available and attention is given to the organ affected. Treatment focuses mainly on controlling chronic bronchial infection-inflammation.

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References