positive in heterozygosis, diagnosing the HIDS. The patient started therapy with Anakinra, Interleukin -1 antagonist.

DISCUSSION

Renal amyloidosis comprise a spectrum of vascular, glomerular, and tubulointerstitial deposition. The reason for the preferential localization to one or the other compartment is not well established. It seems very likely that the varying chemicophysical properties of the amyloid fibrils determine the tropism. Currently, the distribution patterns do not aid in the management of the patients.[3] At least 25 different precursor proteins are known and are associated with a variety of inflammatory, immune, infectious, and hereditary conditions. Most renal amyloidosis is either the result of primary fibrillar deposits of immunoglobulin light chains known as amyloid L (AL) or secondary to fibrillar deposits of serum amyloid A (AA) protein fragments. Renal involvement can be found in some monogenic diseases, the hereditary periodic fever syndromes, which present with recurrent inflammation and unexplained fevers as part of their phenotype. Familial Mediterranean Fever (FMF) is the most widely known and the most prevalent of these inherited disorders.[4] The most dreaded complication of untreated FMF is amyloidosis, which eventuates in renal failure in as many as 20 % of patients in some populations. HIDS, which mimics FMF, is a much rarer disease; it has been reported mainly in families of European ancestry, most of whom are clustered in The Netherlands. It is inherited in autosomal recessive manner. Mutations in the gene encoding MVK constitute the molecular defect in HIDS. The defective gene resides on chromosome 12q. MVK is a key enzyme in the biosynthesis of cholesterol and isoprenoid. HIDS is caused by a defect in the isoprenoid pathway; presumably, intermediary metabolites of the isoprenoid pathway (or a shortage of certain metabolites) influence the immune system in such a way that high levels of IgD are produced.[5] HIDS is characterized by recurrent, self-limiting attacks of fever occurring since early childhood. Febrile episodes usually last 3–7 days and are variably associated with headache, arthralgias, lymphadenopathy, abdominal pain, diarrhoea, vomiting, and skin lesions. The diagnosis of HIDS is based on clinical criteria and elevated serum immunoglobulin D (IgD) levels (100IU/ml). Amyloidosis has been reported only rarely in HIDS. In 2006 Obici et al. did the first report to describe the occurrence of renal AA amyloidosis causing severe nephrotic syndrome in a young Italian man affected with HIDS.[6]

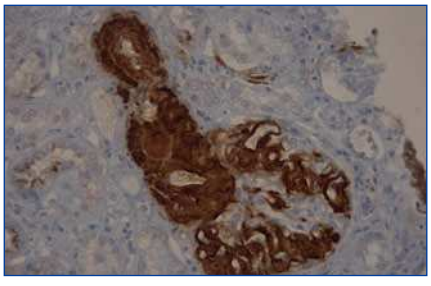

Figure 3. Kidney biopsy
The involvement of a glomerulus and its urinary pole by amyloid deposits intensively and specifically stained with an anti-amyloid A antibody (Peroxidase-Antiperoxidase/Diaminobenzidine stained, 40x).

Conflict of interest

The authors declare that there is no conflict of interest associated with this manuscript.

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fe de erratas

En el número anterior de la Revista NEFROLOGÍA, volumen 32, número 5 del año 2012, se publicó un obituario dedicado al Doctor Xosé Manuel Lens Neo. Por error, se omitió uno de los firmantes: **Miguel A. García González,** Complexo Hospitalario de Santiago de Compostela (CHUS), Instituto de Investigación Sanitaria (IDIS), Grupo de Genética y Biología del Desarrollo de las Enfermedades Renales.

Pedimos disculpas a los lectores de la Revista.