DISCUSSION

Non-amyloid glomerulopathies with Congo red-negative organized immunoglobulin (Ig) deposits display two classical patterns on electron microscopy (EM): randomly arranged (pseudo-amyloid) fibrils, or microtubules, which are usually larger and ordered in parallel arrays. It is currently unclear whether this morphological distinction is relevant. Certain investigators consider this distinction to have no clinical or pathophysiological basis; they suggest that fibrillar and microtubular Ig deposits should be considered simply as variants of the same glomerulopathy, referred to as fibrillary-immunotactoid glomerulonephritis (GN). They also advocate limiting this diagnosis to patients with monoclonal gammapathy, systemic diseases or lymphoproliferative disorders. Others claim that it is essential to distinguish between immunotactoid (microtubular) GN and fibrillary GN (FGN). In immunotactoid GN, defined by the presence of orderly arranged microtubules that are usually more than 30 nm in diameter, glomerular Ig deposits are often monotypic. Immunotactoid GN may be associated with monoclonal gammopathy and/or hematological malignancy with improved renal prognosis upon chemotherapy. Interestingly, monoclonal microtubular Ig deposits with a microtubule diameter smaller than that commonly observed in immunotactoid GN have been described in a few patients with chronic lymphocytic leukemia or related B-cell lymphoma. In contrast, in FGN patients, the fibrils are consistently less than 30 nm in diameter and there is usually no underlying hematological disease. A predominance of polyclonal IgG4 in glomerular deposits also has been reported in patients with FGN, but not in those with microtubular deposits.

RENAL PROGNOSIS

FGN and immunotactoid GN generally respond poorly to corticosteroids and cytotoxic drugs, with an incidence of end stage renal disease of 50%.