CASE REPORT

Fibrillary glomerulonephritis: a case report

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Fibrillary glomerulonephritis is a recently recognised condition. The usual presentation is heavy proteinuria. The diagnosis is established by demonstration of characteristic Congo-red negative, randomly arranged microfibrils in the glomeruli by electron microscopy. At present, there is no proven effective treatment for this condition and the prognosis is generally poor. The first case of fibrillary glomerulonephritis diagnosed in Hong Kong is reported here in a 38-year-old woman.

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Introduction

Fibrillary glomerulonephritis is characterised by deposition of Congo-red negative, randomly arranged microfibrils in the glomeruli. The deposited microfibrils are approximately 18 to 22 nm in diameter. This is a relatively new disease entity, first being noticed in the mid-1970s. We report the first case of fibrillary glomerulonephritis diagnosed in Hong Kong, and review the clinical and pathological features of this condition.

Case report

A 38-year-old Chinese woman with previously good health, presented with an initial incidental finding of proteinuria; the patient was asymptomatic. A physical examination yielded normal findings except for mild ankle oedema. Her blood pressure was 110/60 mmHg and serum albumin was reduced to 37 g/L. Otherwise, the complete blood count, liver and renal biochemistry profiles were normal. The creatinine level was 87 μmol/L. Tests for autoimmune antibodies which included antinuclear factor (ANF), antiDNA and serum immunoelectrophoresis (IEP) were all negative and complement levels were normal. Urine microscopy revealed microscopic haematuria and elevated urine protein excretion of 3.65 gm/day. Ultrasonography of the abdomen showed the kidneys to be of normal size, but with increased parenchymal echogenicity. A renal biopsy was performed.

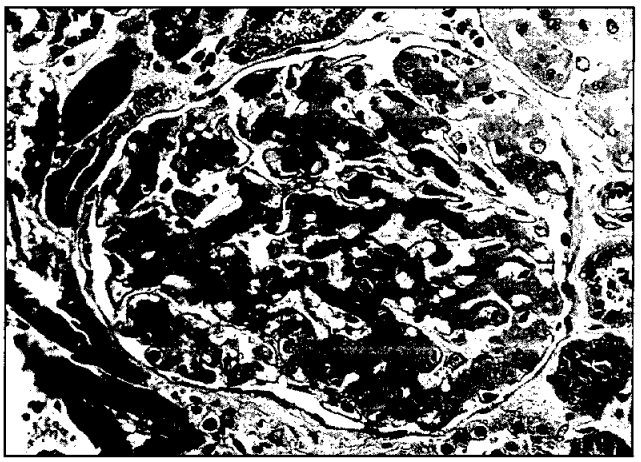

Fig 1. Glomerulus showing marked expansion of mesangial matrix associated with mild increase in mesangial cells. The capillary walls are not thickened (H & E, x 280).

Examination of the biopsy tissue showed all of the glomeruli to be enlarged, with massive mesangial deposition of eosinophilic, Periodic acid-Schiff (PAS) positive material (Fig 1). Peripheral loop involvement was segmental with obliteration of a few capillaries and formation of double contours. There was also a mild increase in mesangial cellularity. Both renal tubules, interlobular arteries and the interstitium were normal. Congo-red staining was negative, while immunofluo-

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rescence studies showed strongly positive staining for IgG, C3, κ- and λ- light chains, and weak C1q staining in the expanded mesangium. Electron microscopy showed that coarse non-branching microfibrils made up the mesangial deposits. The microfibrils ranged from 20 to 30 nm in diameter, diagnostic of fibrillary glomerulonephritis (Fig 2).

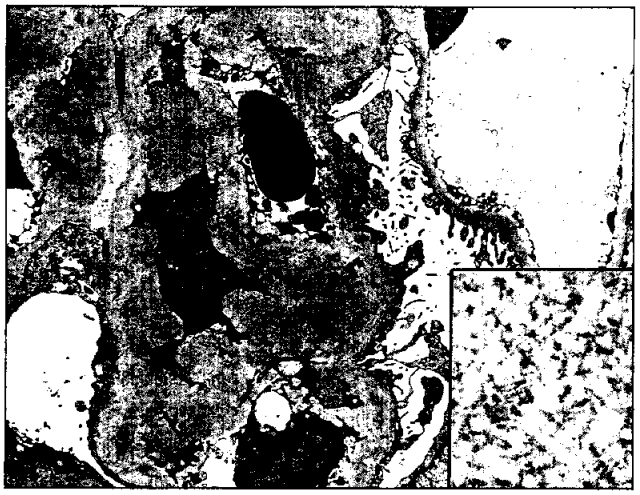

Fig 2. Electron microscopy showing massive mesangial deposits (x 4500). Inset: the deposits are composed of randomly arranged fibrils which are 20 to 30 nm in diameter (x 50 000).

Discussion

Glomerulopathy characterised by the infiltration of Congo-red negative fibrils into the glomeruli was first described in 1977.[1] In 1983, Duffy reported eight patients with this condition and used the term glomerulonephritis with fibrillary deposits.[2] Subsequently, the term fibrillary glomerulonephritis was adopted.[3] Currently, approximately 150 cases of fibrillary glomerulonephritis have been reported in the literature.

Fibrillary glomerulonephritis is a pathological diagnosis. The light microscopic appearance is variable, with capillary wall thickening, mesangial matrix expansion and hypercellularity being the most frequent findings. Crescent formation has been noted in approximately 20% of cases.[4] Immunofluorescence studies show intense staining for IgG and weaker staining for C3 in a predominantly mesangial pattern with IgG4 as the dominant immunoglobulin subclass.[5] Electron microscopy shows characteristic randomly arranged microfibrils with a diameter from 18 to 22 nm.[4]

Differential diagnoses of fibrillary glomerulonephritis include amyloidosis and immunotactoid glomerulopathy. Amyloidosis differs from fibrillary glomerulonephritis in that it stains positively with Congo-red and the microfibrils are smaller in size. In immunotactoid glomerulopathy, the microfibrils are larger in size—being 30 to 40 nm in diameter—and they are usually arranged in an orderly fashion. In addition, patients with immunotactoid glomerulopathy have a higher incidence of lymphoproliferative disorders. The pathogenesis of fibrillary glomerulonephritis is still unknown, although the homogenicity of the immune deposit in the glomeruli could be the basis for the fibrillar ultrastructural appearance. It has been demonstrated by immunoelectron microscopy that the fibrils of fibrillary glomerulonephritis actually contain the immunoglobulin seen in immunofluorescent microscopy.[6]

The average age at presentation of patients with this condition is approximately 50. Clinical features include heavy proteinuria which is usually in the nephrotic range, microscopic haematuria and renal impairment.[4] Extrarenal manifestations are rare.[5] The frequency of this condition among patients undergoing renal biopsy has been reported to be around 1%.[4,5]

The optimal treatment for this condition remains to be established. Response to steroid therapy alone has been inconsistent, although there have been isolated case reports which showed a favourable response to immunosuppressive therapy using high dose steroids and cyclophosphamide.[7] The renal prognosis for this condition is generally poor. Two years from diagnosis, approximately 50% of patients have progressed to end-stage renal failure.[3]

Although fibrillary glomerulonephritis is not a rare condition, this appears to us to be the first case of fibrillary glomerulonephritis reported in Hong Kong. The condition is probably locally underdiagnosed. A diagnosis of fibrillary glomerulonephritis relies on its characteristic electron microscopic appearance. With an increased awareness of this condition and the more frequent use of electron microscopy when examining renal biopsy material, we can expect more cases to be diagnosed in the future. Further studies are needed in order to elucidate the underlying mechanism of glomerular injury in this condition. This may enable us to develop more effective treatment for this condition.

References

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