Case Report

Goodpasture's disease: A case report from South India

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Abstract

A 28-year-old male died following massive hemoptysis at Nizam's Institute of Medical Sciences, Hyderabad. He presented with recurrent hemoptysis and hematuria. The lungs showed alveolar hemorrhages with hemosiderin laden macrophages. Kidneys showed focal proliferative glomerulonephritis with crescents. Direct immunoflouresence of the kidney showed linear deposition of IgG and granular deposits of C3.

Key words: Goodpasture's disease, pulmonary renal syndrome

Goodpasture's disease is a rare cause of massive hemoptysis and is a form of antiglomerular basement membrane disease. Linear Ig G deposits along the glomerular and pulmonary basement membranes by direct immunoflouresence and demonstration of antiglomerular basement membrane antibodies, are diagnostic of the disease. The incidence is much less in Indian population. Available literature is from northern India. No cases have been reported from south India. This case report is an autopsy study of a patient who died of pulmonary and renal manifestations at Nizam's Institute of Medical Sciences, Hyderabad.

Case Report

A male patient aged 28 years presented with recurrent episodes of hemoptysis with dyspnea of 2 months duration. There was a single episode of massive hemoptysis, 20 days prior to the admission. He was a smoker. His brother had hematuria.

On examination he was pale and lungs showed bilateral diffuse crepitations. Two days after the admission, he developed massive hemoptysis and died. An autopsy was performed.

Investigations

Routine investigations showed hemoglobin of 9.3 g/dl, MCV 72.8fl, MCH 23.7 pg, red cell count 3.9 million/ mm^3 and PCV 28.5%. Total leukocyte count was 20,000/ mm^3, ESR 6 mm/1st hour and platelet count 4.4 lakhs/ mm^3. Urinanalysis revealed specific gravity of 1.030, albumin 4 +, red cells 120-150/high power field and granular casts 1-2/low power field. Serologic tests for HIV, HB S Ag and ANCA were negative. Antiglomerular basement membrane antibodies done by indirect immunofluorescence were negative. Chest radiograph and CT scan showed nodular opacities.

Autopsy findings

On gross evaluation, the lungs were congested and the trachea was filed with blood. Microscopic examination of routine H and E stained sections showed extensive alveolar hemorrhages with hemosiderin-laden macrophages and focal hyaline membrane. Kidneys showed focal proliferative glomerulonephritis with crescents in approximately five percent of glomeruli [Figure 1]. Direct immunofluorescence showed linear deposition of Ig G [Figure 2] and granular focal deposition of C3 along the glomerular basement membrane. A diagnosis of Goodpasture's disease was given.

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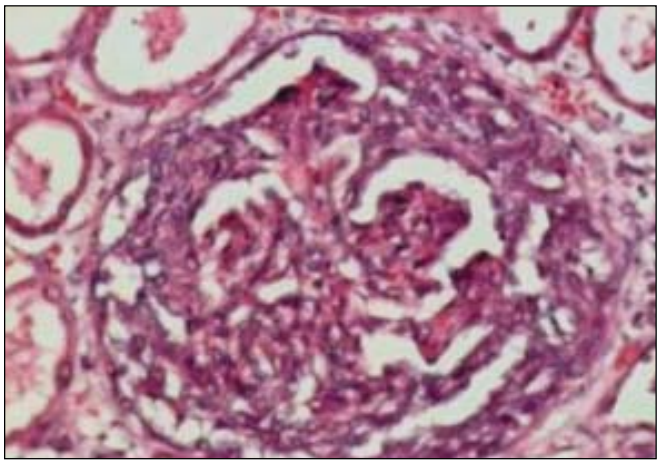

Figure 1: Glomerulus with circumferential cellular crescent (H/E, 200x)

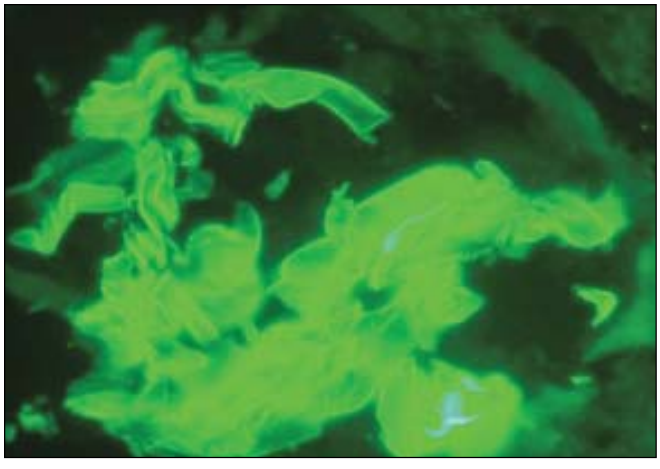

Figure 2: Linear deposits of Ig G along the glomerular basement membrane. Direct immunofluorescence (400x).

Discussion

Goodpasture's disease is a rare form of pulmonary renal syndrome, described in 1919 by Ernest Goodpasture. In United Kingdom, the incidence is one case in million population per annum. The incidence is much less in Asian Indians[1,2] hence, there are very few reported cases from India[3,4] and to the best of our knowledge, none from south India. It is possible that the disease is not so uncommon. But majority of the hospitals lack facilities for immunofluorescence, hence, it is usually suspected, but seldom confirmed.

Pathogenesis is known to be due to the presence of anti glomerular basement membrane antibodies, directed against the NC1 domain of α-3 type IV collagen.[5,6] The patients generally are young males. Inherited susceptibility is well documented through HLA complex. The disease is known to be associated with ANCA vasculitis, Alport's syndrome, membranous nephropathy, diabetes mellitus and lymphoma.

Immunofluorescence shows linear deposits of Ig G, even if there are no light microscopic glomerular changes and all the glomeruli show this characteristic immunofluorescence pattern. In autopsy setting, linear Ig G deposits should be interpreted with caution, as they can also be seen in diabetes mellitus, SLE, perfused kidneys, fibrillary nephritis and in addition to autopsy kidneys.[2] Presence of anti glomerular antibodies is diagnostic of the disease. Techniques for demonstration include indirect immunofluorescence, radio immuno assay and ELISA. The latter two techniques are considered superior to the former. False negative results are seen if the test is performed late in the course of the disease. All the patients of anti GBM antibody deposition in the tissues need not be symptomatic. Smoking, cocaine inhalation, exposure to organic solvents, hydrocarbons and Influenza A2 virus infection are known to predispose to hemoptysis.[7,8] Renal course is influenced by percentage of crescents and levels of circulating antibodies. Classical renal histology is a crescentic glomerulonephritis. However, if the presentation is pulmonary, the kidney may not show light microscopic changes or may show focal proliferation. Clinical presentation may be hemoptysis or rapidly progressive renal failure, based on the organ which is dominantly involved.

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