Goodpasture's disease in a patient with Kartagener's syndrome: An unusual association

Sir,

Kartagener's syndrome is a rare disorder characterized by the triad of situs inversus, bronchiectasis, and sinusitis.[1] Goodpasture's disease is an autoimmune disease characterized by the production of anti-glomerular basement membrane (GBM) antibodies, pulmonary hemorrhage, and glomerulonephritis.[2] Association of these two disorders in the same patient is not reported so far.

A 55-year-old lady presented with shortness of breath, fever, productive cough, streaky hemoptysis, and oliguria of 10 days duration. She was told to have situs inversus totalis 15 years back when she was evaluated for recurrent respiratory tract infections. Findings on examination were pallor, clubbing, and bilateral basal crackles. Laboratory investigations revealed hemoglobin of 5 g/dl, total leukocyte count of 12,000/mm^3 with normal differential counts, and platelet count of 2.3 lakh/mm^3. Urine examination showed active urinary sediment with red blood cell casts. Her serum creatinine was 14.1 mg/dl, 24 h urine protein of 1.8 g/day, and sonography showed normal sized kidneys. The antinuclear antibody, double-stranded deoxyribonucleic acid, antineutrophil cytoplasmic antibody, human immunodeficiency virus, and anti-hepatitis C virus antibodies were negative. Complement levels (C3, C4) were within normal limits. A percutaneous renal biopsy revealed 100% cellular crescents and linear pattern of immunoglobulin deposition on direct immunofluorescence, pathognomic of Goodpasture's disease [Figure 1]. Serum anti-GBM antibodies were negative. She was treated with intravenous pulse methyl prednisolone and intravenous cyclophosphamide. She was also given 3 sessions of plasma exchange with fresh frozen plasma as a replacement. On further evaluation, high-resolution computed tomography (CT) chest revealed situs inversus totalis [Figure 2] with bilateral lower lobe bronchiectasis with alveolar hemorrhage. CT scan of paranasal sinuses revealed bilateral maxillary and ethmoid sinusitis. During the course of hospital stay, she had massive hemoptysis and succumbed to her illness.

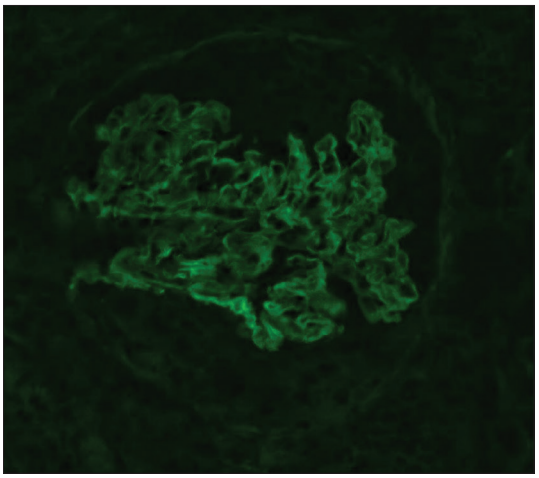

Figure 1: Immunofluorescence picture of renal biopsy showing linear immunoglobulin G pattern along glomerular basement membrane-pathognomic of Goodpasture's disease

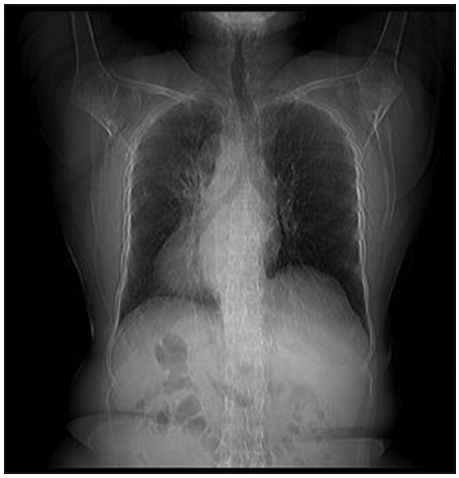

Figure 2: Scout film of high-resolution computed tomography chest showing situs inversus totalis

Our patient presented as rapidly progressive glomerulonephritis (RPGN) and was diagnosed as Goodpasture's disease based on typical histopathology. Anti-GBM antibodies were negative in this case, but serological assays are not reliable as their sensitivity varies from 63% to 100%.[3] Good prognostic factors for Goodpasture's disease are a histopathological percent of crescents (<30%) and plasma creatinine (<3 mg/dl).[4] Our patient presented with RPGN and was dialysis dependent and died of pulmonary alveolar hemorrhage. The various precipitating factors for Goodpasture's disease are increased capillary hydrostatic pressure, bacteremia, endotoxemia, exposure to volatile hydrocarbons, upper respiratory infections, and tobacco smoking.[5] We hypothesize that the recurrent respiratory tract infections associated with Kartagener's syndrome in our patient could be a precipitating event for Goodpasture's disease.

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Conflicts of interest

There are no conflicts of interest.

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