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Wegener granulomatosis and cancer

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To the editor: A 50 year-old male is presented, who is on periodic hemodialysis since April of 1984 because of chronic renal failure after rapidly progressive glomerulonephritis secondary to Wegener' s granulomatosis.

Wegener's granulomatosis was first described in 1931 by Klinger.[1] In 1936, Wegener clearly defined the clinical and pathological picture.[2] The etiology remains unknown. It is a systemic necrotizing vasculitis, which involves small arteries and veins, above all in the upper and lower respiratory tracts and the kidney.[3]

Renal involvement manifests with proteinuria, hematuria and red cell casts, and progressive renal failure. The most frequent pathological finding is ~~a~~ focal necrotizing glomerulonephritis. Interstitial nephropathy and vasculitis, as well as proliferative glomerulonephritis, can also be found.[4]

In January of 1984, a renal biopsy was performed and extracapillary necrotizing glomerulonephritis was observed. A biopsy of the nasal cavities was normal.

The patient received immunosuppressive therapy with steroids and cyclophosphamide, but the renal function did not recover.

In February of 1995, the patient presented a flare of vasculitis, which consisted in the development of a necrotizing ulcer at the internal malleolus of the right foot, pharyngitis, and diffuse intraalveolar hemorrhage with a radiological pattern of bilateral alveolar infiltrate.

In the lungs, nodules, atelectasis, pleural effusion and infiltrates can be seen. Cavitation is frequent.[5]

At that moment, the patient had positive anti-neutrophilic cytoplasm autoantibodies (c-ANCA) with specificity for anti-proteinase 3.

The four diagnostic criteria are nasal or oral inflammation, abnormal radiological image, signs of activity in the urinary sediment, and demonstration of granulomatous inflammation.[6] The presence of ANCA has also been shown to be useful for making the diagnosis.[7,8]

The patient reinitiated treatment with oral cyclophosphamide (1 mg/kg/day) and prednisone (60 mg PO with progressive dose reduction). The treatment was maintained for more than a year.

It is known that cyclophosphamide is associated with bladder cancer, due to the action of a drug metabolite, called acrolein, which has also been related to a higher incidence of skin cancer and myeloproliferative syndromes.[9]

In May of 2002, he was diagnosed with a stage IV A non-Hodgkin lymphoma by means of a lymph node biopsy. He was treated with 6 cycles of CHOP and rituximab with partial remission. Since 2003, he has been on maintenance therapy with periodic cycles of rituximab.

In September of 2007, twenty-three years after the diagnosis of Wegener's disease, he presented an upper respiratory tract infection and a chest X-ray film was performed, which showed left pleural effusion not present on the control CT scan performed in January (fig. 1). Immunological investigations were negative. A new CT scan was made, which confirmed the presence of left pleural effusion and multiple pulmonary nodules. Diagnostic and therapeutic thoracocentesis was performed. The fluid was hematic in appearance and the biochemistry was compatible with an exudate. The pathological study disclosed an adenocarcinoma of the lung and palliative therapy was initiated.

It is well known that Wegener's disease has a fatal prognosis if left untreated, but the treatment is on the long term associated to an increased cancer risk, as clearly illustrated by this case.

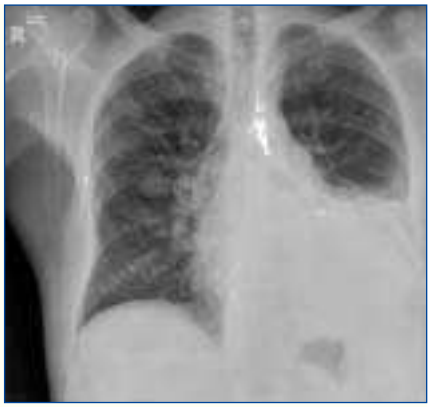

Figure 1. Left pleural effusion and multiple pulmonary nodes.

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