

Relapse and Treatment Resistance in Patients with Small-Vessel Vasculitis

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The full report is titled “Predictors of Relapse and Treatment Resistance in Antineutrophil Cytoplasmic Antibody–Associated Small-Vessel Vasculitis.” It is in the 1 November 2005 issue of *Annals of Internal Medicine* (volume 143, pages 621-631). The authors are S.L. Hogan, R.J. Falk, H. Chin, J. Cai, C.E. Jennette, J.C. Jennette, and P.H. Nachman.

What is the problem and what is known about it so far?

Vasculitis is blood vessel inflammation that develops when the immune system attacks the body. Inflamed blood vessels can decrease blood flow or cause small ruptures and bleeding into tissues. Several types of vasculitis primarily affect smaller blood vessels. Many of these types are associated with a kind of antibody called antineutrophil cytoplasmic antibody (ANCA). The condition commonly referred to as “ANCA-positive” small-vessel vasculitis most often affects the lung, sinuses, and kidneys. Doctors often treat patients with ANCA-positive small-vessel vasculitis with drugs that suppress the immune system. Most patients get better with treatment, but some do not. Some also relapse during or after treatment, meaning the disease and symptoms return and more treatment is usually required.

Because of fear of relapse, doctors often give drugs to patients with small-vessel vasculitis for a long time. Giving the patients drugs for a longer time increases the risk for serious side effects. If doctors could identify which patients were more likely to relapse, they could treat them for a longer time and spare patients who are unlikely to relapse from potential side effects from long-term treatments.

Why did the researchers do this particular study?

To see which patients with ANCA-positive small-vessel vasculitis would respond to treatment and which would relapse.

Who was studied?

334 patients treated for ANCA-positive small-vessel vasculitis in the southeastern United States. Most patients had their disease diagnosed by kidney biopsy.

How was the study done?

The researchers studied patients with ANCA-positive small-vessel vasculitis, most of whom had kidney disease. Most patients were treated with corticosteroids plus an additional immunosuppressive drug. The researchers observed what happened to the patients over about 4 years. Patients whose kidney disease worsened or who had symptoms of vasculitis that never went away during treatment were considered resistant to treatment. Patients who initially improved with treatment but then worsened were considered to have relapsed. The researchers compared the patients by age, ethnicity, and other factors to identify features that predicted treatment resistance and relapse.

What did the researchers find?

About 1 of every 4 patients was resistant to treatment. Most patients who were resistant to treatment reached end-stage kidney disease, meaning they needed dialysis or a kidney transplant to survive. Treatment with corticosteroids only, without the benefit of another immunosuppressive drug, was strongly correlated to treatment resistance. Women, African-American patients, and those with severe kidney disease before treatment were more likely to be resistant. Of the patients who did respond to treatment, about 40% had a relapse. Patients who had lung disease, upper airway (sinus) disease, and a specific type of ANCA called proteinase 3 (PR3) ANCA were more likely to have a relapse than patients without these characteristics.

What were the limitations of the study?

Most of the patients had their vasculitis diagnosed by kidney biopsy. These patients may differ from patients with small-vessel vasculitis who do not have kidney disease or who do not undergo kidney biopsy. Also, the patients were cared for by different doctors whose treatment decisions differed. As a result, the researchers’ findings might be the result of differences in doctors’ treatment decisions or other factors the researchers were not able to measure.

What are the implications of the study?

Among patients with ANCA-positive small-vessel vasculitis, adequate treatment with corticosteroids and other immunosuppressive agents is important at diagnosis. Women, African-American patients, and patients with severe kidney disease may be less likely to respond to initial treatment. Among patients who respond to initial treatment, those with lung or sinus disease involvement and specific patterns of antibodies (PR3 ANCA) in the blood are more likely to relapse. These findings require confirmation, but they suggest that doctors may be able to reserve long-term treatment for some patients with small-vessel vasculitis, sparing others the side effects of prolonged exposure to immunosuppressive drugs.

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