TANAZ KERMANI:

Hello my name is Tanaz Kermani. I'm a rheumatologist and an assistant professor of medicine at Mayo Clinic in Rochester. Our article titled "Idiopatic Retroperitoneal Fibrosis, A Retrospective Review of Clinical Presentation, Treatment, and Outcomes" has been published in the April 2011 issue of the *Mayo Clinic Proceedings.* Retroperitoneal fibrosis is an uncommon disorder characterized by fibro-inflammatory infiltrates surrounding the abdominal aorta and adjacent tissues.

Periureteric involvement can result in acute renal failure. Most cases are idiopathic. We were interested in evaluating all cases seen at Mayo Clinic between 1996 and 2006 who had a diagnosis of idiopathic retroperitoneal fibrosis. Our objective was to evaluate the clinical features, treatment, and outcomes in these patients. We identified 185 patients. In contrast to prior studies where a greater proportion of men have been reported, our study only found a slight male predominance. 61% of patients in the study were men.

Symptoms at presentation were often nonspecific. The most common presenting symptom was abdominal, back, or flank pain. Laboratory tests were also nonspecific. Markers of inflammation, including sedimentation rate and C-reactive protein were elevated in 58% of patients at diagnosis. In other words, 42% had normal tests at diagnosis.

Renal dysfunction was observed in about 42% of cases with a median creatinine of 2.3 milligrams per deciliter. Majority of the cases were diagnosed by imaging findings, which included the CT of the abdomen in most cases. This showed that the primary retroperitoneal mass was around the abdominal aorta. Additionally, hydronephrosis was observed in 57% of patients. Most patients in the series were treated with a combination of medications and urologic intervention.

Most common urologic procedure performed was ureteral stenting. Common medications used were corticosteroids, methotrexate, and tamoxifen. Follow-up was available for 151 patients. And at last follow-up, 68% had normal creatinine. Additionally, we followed these patients with serial imaging studies. And imaging studies showed stabilization or improvement of the mass in 97%. However, relapses despite treatment were observed in 12% of cases.

Our study included a large cohort of patients with idiopathic retroperitoneal fibrosis with a long duration of follow-up. There are several clinical implications to our findings. Firstly, laboratory tests can be normal at presentation and also at the time of relapse. Therefore, imaging studies appear to be a reliable way to diagnose and follow these patients over time. Patients with retroperitoneal fibrosis often require a multidisciplinary approach. Most patients in the study were treated with a combination of urologic procedures and medications.

Finally, given that relapses were observed in 12% of our patients, long-term follow-up of these patients is warranted. In terms of implications for patients, our findings showed that overall prognosis is favorable in patients with idiopathic retroperitoneal fibrosis. However, given the relapse rate, these patients do require periodic medical assessment of their disease. There are several aspects of disease management that warrant further investigation.

While several medications, primarily immunosuppressive medications, have been tried to treat this disease, the optimal treatment strategy is unknown. Additionally, the frequency with which patients should be monitored also needs to be clarified. In summary, while renal dysfunction is common at presentation in retroperitoneal fibrosis, overall renal outcomes are favorable. Given the relapse rate observed in our study, which was 12%, these patients do warrant close follow-up. Our practice has been to follow these patients with serial imaging studies, although the optimal frequency with which these need to be performed remains to be clarified.

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