Case Study: IgG4-Related Diseases from Diagnosis to Treatment

Announcer:
You're listening to Conference Coverage on ReachMD, captured on location at the Congress of Clinical Rheumatology’s Annual Meeting in Destin, Florida. Your host is Dr. Madelaine Feldman, Clinical Associate Professor of Medicine at Tulane University Medical School and Vice President of the Coalition of State Rheumatology Organizations.

Dr. Feldman:
This is Dr. Madelaine Feldman, and I'm here at the Congress of Clinical Rheumatology, and I'm here with Dr. Marissa Sansone, who is an internal medicine resident at Jersey City Medical Center, Barnabas Health, and she is presenting a case of IgG4-related disease which strikes the kidney.

Dr. Sansone, what can you tell us about IgG4-related diseases?

Dr. Sansone:
Hi, so it's basically a fibroinflammatory disorder comprised of IgG4 positive plasma cells that cause obliterative phlebitis, storiform fibrosis, and symptoms usually occur depending on where they deposit, so basically, in which organs are affected.

Dr. Feldman:
What is the best way to diagnose IgG4 disease?

Dr. Sansone:
Usually, we order IgG4 levels like within the hospital. We usually like to see a level at least over 300 mg/dL, but sometimes it’s very nonspecific, so the best way is usually histopathology showing obliterative phlebitis, fibrosis, and then a high percentage of IgG4 positive plasma cells, usually around like 40, around 40 over a certain area.

Dr. Feldman:
I see your case has it striking the kidney. Is this a common area for IgG4?

Dr. Sansone:
So it’s not necessarily uncommon, but it’s usually hard to diagnose because a lot of the times there’s overlap with other diseases, and sometimes it’s misdiagnosed as either a lupus nephritis or a Sjögren’s, or there’s an overlap with GPA, so that’s why a lot of these patients usually with renal biopsy, that’s when it’s usually diagnosed more.

Dr. Feldman:
So, how do you treat IgG4?

Dr. Sansone:
So usually, steroids are first-line. They usually respond well with steroids. But in cases of refractory IgG4, induction and maintenance therapy with rituximab has shown to be effective.

Dr. Feldman:
Thank you so much.

Dr. Sansone:
Thank you very much.

Announcer:
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