
IGG 4 Related Diseases, Are We Under or over Diagnosing?? Clinical Observations in UAE

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During our practice, over last 40 years we have seen many missed diseases related possibly to IgG4 related disease, an example of that, suspected pancreatic cancer proved to be inflammatory in nature (autoimmune pancreatitis), many cases of hepatobiliary diseases, and in one case diagnosis was made of sclerosing cholangitis with ulcerative colitis proved to be IgG4 related diseases after 7 years.

From our recent experience in Abu Dhabi in our center, we came across many cases of elevated IgG4 associated with inflammatory bowel diseases especially ulcerative colitis and hepatobiliary diseases simulating sclerosing cholangitis, multiple autoimmune diseases including thyroiditis, eczema, prostatitis and pachymeningitis.

In all these IgG4 was twice the normal but the real link to the disease whether it is primary cause of the disease or associated phenomena is not yet settled.

Our observation in patients with high IgG4 showed that it carries more severe diseases, more relapses and protracted course.

We believe awareness is needed to highlight the importance of this rising star in our practice

IgG4-RD is a fascinating clinical entity including a wide variety of diseases, formerly diagnosed as Mikulicz's disease, interstitial nephritis, prostatitis retroperitoneal fibrosis, autoimmune pancreatitis (AIP), and other auto immune diseases like Sjogren syndrome, SLE [1-5].

Universal criteria for IgG4-RD have not yet been established at present, making its diagnosis in some patient's ambiguous leading to many IgG4-RD mimickers [6].

A 3-year investigation by the Japanese IgG4 team, organized the Ministry of Health, Labor and Welfare (MHLW) of Japan, has reached a consensus, in that IgG4-RD can occur in various organs, with clinical symptoms depending on lesion location [6].

Characteristics common to all forms of IgG4-RD include elevated serum IgG4 concentration and tissue infiltration by IgG4-positive plasma cells, accompanied by tissue fibrosis and sclerosis [2].

Our observations over the last 4 years on patients with multiple auto immune diseases or inflammatory bowel diseases based on high IgG4 level above 135mg/dl [6].

1-Male 54 years old with pachymeningitis treated with steroids and azathioprine.

2-Female child 13 years old, pancreatic and liver & biliary involvement responded completely to steroids.

3-Female 34 with ulcerative colitis, Hashimoto thyroiditis with recurrent clostridium difficile, responded to steroids, azathioprin and adalimumab.

4-Male 54 Hashimoto thyroiditis, eczema, arthritis, latent celiac disease, ulcerative jejunitis, eosinophilic esophagitis, empty sella syndrome, on steroids & azathioprine, frequent relapses.

5-Male 65 cholestatic jaundice with pancreatic mass and dilated intra hepatic bile ducts ducts, responded to stenting and steroids

6-Male 57 diabetic 20 years, ulcerative colitis 15 years, recurrent venous thrombosis, liver cirrhosis, portal vein thrombosis, factor V leiden mutation, thyroid follicular cancer following treatment with azathioprine, adalimumab & golimumab.

7-Male 35 Crohns disease, psoriasis on azathioprine and infliximab

All the above patients have elevated IgG4 [1].

8-Some cases with IgG4 elevation below 135 but above our normal 84 included involuntary movements, celiac disease, arthritis, psoriasis [6].

Our observations showed that those with raised IgG4 have frequent relapses and poor responses to treatment [5].

The key issue is that we need to study more cases and associations and to determine if the high level is causative or a secondary phenomena.

More data from different specialties is needed.

We feel it is time to raise awareness about this associations especially in patients with multiple auto immune or inflammatory diseases [1].

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