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UK IgG4-Related Disease Symposium

2-3 March 2023

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THE ROLE OF TRANSIENT ELASTOGRAPHY IN IgG4 RELATED HPB DISEASE

Oxford University Hospitals NHS



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Background & Aims

IgG4-Related Disease (RD) is associated with inflammation and fibrosis. IgG4-Hepatobiliary (HPB) disease affects the liver, bile ducts and pancreas. Vibration controlled transient elastography (VCTE) for liver stiffness has a high-performance characteristic for detecting advanced liver fibrosis in liver diseases such as metabolic, viral hepatitis, autoimmune and cholestatic liver diseases. The role of VCTE in patients with IgG4-HPB disease to assess disease activity, damage and progression has not been addressed.

Methods

Descriptive single centre study; 85 IgG4-RD with VCTE readings by Fibroscan. Data was collected retrospectively from patient electronic health records. Baseline and progression of liver stiffness readings, flar activity, organ involvement, serum biomarkers of activity were documented and studied.

Results

65 (76%) men, and the dominant phenotype was IgG4-HPB disease (79%). Median value of liver stiffness was 5.5kPa; IgG4-HPB cohort median was 5.4kPa.

There was no difference in VCTE during an active flar or remission (Table 1). 30.6% of the patients had serial readings over a median of 4 years.

Significant difference between baseline (median 5kPa, IQR 4.2 - 6.5) and follow up readings (median 6.4kPa, IQR 4.6 - 8.7) ($Z = 0.9$, $p = 0.02$, 97% CI 0.1 - 2.5).

29% (29%) patients had a liver stiffness reading ≥ 8 kPa, with 8 (9%) having a reading > 12 kPa. In this group 74% had a diagnosis of Metabolic Associated Fatty Liver Disease (MAFLD) with diabetes and obesity being the most significant risk factors. (76% and 32% respectively).

Table 1. Comparison of liver stiffness measurements (kPa) between patients with active disease and remission.

	Active disease (n=12)	Remission (n=10)	p-value
Median (IQR)	5.5 (4.2 - 6.5)	5.4 (4.2 - 6.5)	0.9
Range	4.2 - 8.7	4.2 - 8.7	
MAFLD	9 (75%)	8 (80%)	0.8
Obesity	8 (67%)	7 (70%)	0.8
Diabetes	7 (58%)	6 (60%)	0.8

Figure 1. Comparison of liver stiffness measurements (kPa) between patients with active disease and remission.



Conclusions

This is the first study to assess liver stiffness measurements with VCTE using Fibrosan in patients with IgG4-HPB and systemic disease.

Overall patients had a low liver stiffness reading suggesting low incidence of liver advanced fibrosis/cirrhosis. This is supported by clinical data showing low prevalence and rare progression to cirrhosis in IgG4-HPB disease. Liver stiffness measurements increased over time, and this may be a means to track fibrosis progression in the disease.

High liver stiffness readings ≥ 8 kPa were seen in majority of concomitant MAFLD patients. This is important as 50% of patients with IgG4-HPB disease will have exocrine diabetes mellitus and steroid-induced weight gain and worsening diabetes may be an additive risk factor for liver fibrosis.



Ritu...

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1. BACKGROUND

IgG4-RD is a multi-organ mediated immune-mediated disease. Corticosteroids are first-line response. Relapse on to 60%. Leads to organ damage. Rituximab (anti-CD20 antibody) received UK NICE approval for therapy for intolerance/immunomodulators.

2. AIMS AND OBJECTIVES

To assess the indications, clinical outcome to Rituximab in a multi-organ IgG4-RD patients.

3. METHODS

Retrospective data collected from tertiary centres for IgG4-RD patients. Rituximab therapy. Wilcoxon paired test was used for variables.

4. RESULTS

52 patients received Rituximab for IgG4-RD (and figures). Serum IgG4 levels fell with Rituximab (pre-treatment median 4.99g/L, post-treatment median 3.99g/L, $p < 0.001$). The majority of cycles received was 1 pair (range 0-2). 52% received maintenance therapy. Serum IgG4 levels before vs after Rituximab.



IgG4-RD Diagnostic and Classification Criteria



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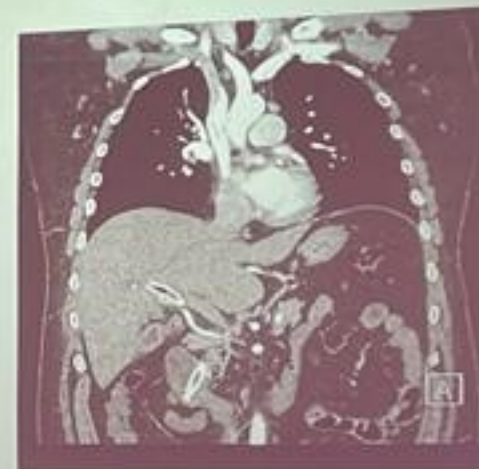
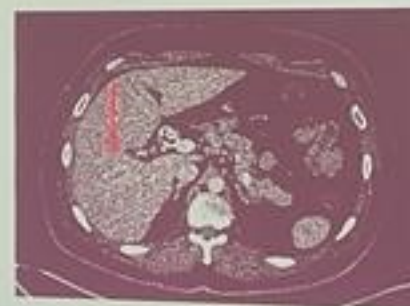
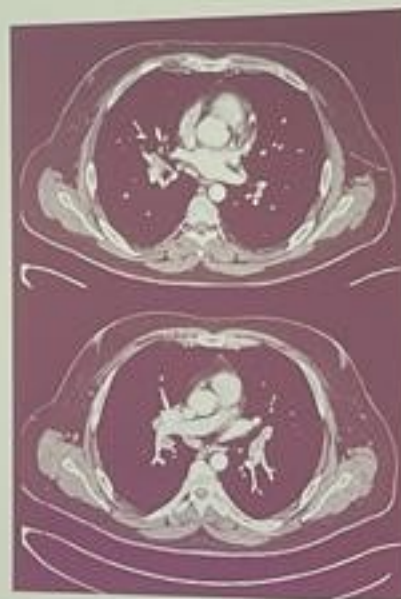


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CT CAP : Multiple enlarged LN : Para aortic, mediastinal, pelvis/groin, 3 biliary stents, subtle 2cm lesion in right liver





Non-Biologic DMARDs in IgG4-RD

- Immunomodulator/immunosuppressant drugs
- Include:
 - Azathioprine
 - Methotrexate
 - Mycophenolate
 - Cyclophosphamide
 - Leflunomide
 - Others (case reports)







