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UK IgG4-Related Disease Symposium

2-3 March 2023

Censors' Room
18:30 - 22:00

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THE ROLE OF TRANSIENT ELASTOGRAPHY IN IgG4 RELATED HPB DISEASE

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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, flare 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.

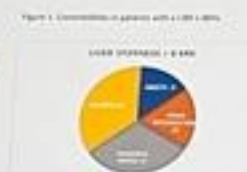
There was no difference in VCTE during an active flare or remission (Table 1). 30.6% of the patients had serial readings over a median of 4 years.

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%) 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 VCTE readings in patients with active disease and remission.

Parameter	Active Disease (n=29)	Remission (n=36)
Median VCTE (kPa)	5.5	5.4
IQR (kPa)	4.2 - 6.5	4.2 - 6.5
Range (kPa)	2.5 - 12.5	2.5 - 12.5
p-value	0.9	0.9



Conclusions

This is the first study to assess liver stiffness measurements with VCTE using Fibroscan 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.

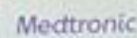
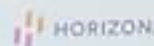
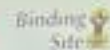
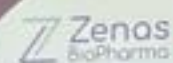
High liver stiffness readings ≥ 8 kPa were seen in majority of concomitant MAFLD patients. This is important as 50% of patients with IgG4-HPB disease will have exocrine diabetes mellitus and steroid-induced weight gain and worsening diabetes may be an additive risk factor for liver fibrosis.



IgG4-RD Diagnostic and Classification Criteria



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Novel Therapies and Clinical Trials



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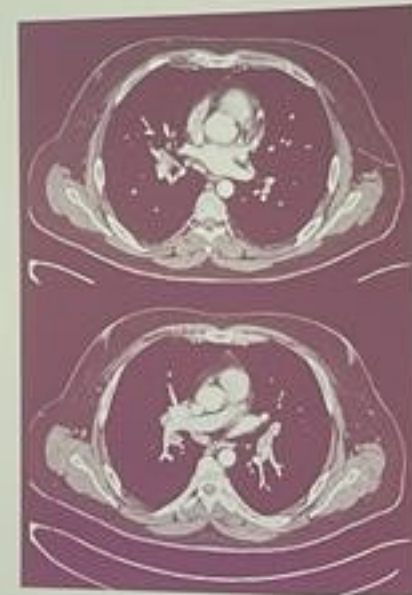

Panel discussion table with two men seated at microphones and water bottles.

Speaker at a podium with a screen displaying the blue logo with a cross and hands.





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
 - Sulfonamide
 - Others (case reports)







