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Introduction

IgG4-related sclerosing cholangitis (IgG4-RSC) has cholangiographic abnormalities that may resemble primary sclerosing cholangitis or cholangiocarcinoma. It is typically seen in males and those in their fifth and sixth decade of life. Diagnosis is achieved with various modalities, which includes serology, histo- and cytopathology, and radiology. It is steroid-responsive, unlike the other differential diagnoses. Delay in treatment may result in infections, fibrosis, and even mortality.

Objectives

To describe the cases of IgG4-RSC in Brunei Darussalam and identify its common presentations, diagnostic methods, and treatment options.

Methods

This is a retrospective study, in which cases between 2019 to 2022 were identified by interview with specialists of the Gastroenterology & Hepatology Unit, RIPAS Hospital.

A template based on the Japanese Clinical Diagnostic Criteria for IgG4-RSC 2020 was created to collect relevant data such as gender, age at diagnosis, symptoms at presentation, serological, histopathological, and radiological findings, and treatment upon diagnosis from the national electronic patient records.

Diagnostic Items	Case 1: 61 F	Case 2: 64 M	Case 3: 44 M	Case 4: 68 M
I. Narrowing of the intrahepatic and/or extrahepatic bile duct:				
a. ERC	X	✓	✓	✓
b. MRCP	✓	X	X	✓
II. Thickening of the bile-duct wall:				
a. EUS/IDUS	✓	✓	✓	X
b. CT/MRI/US	✓	✓	✓	✓
III. Serological findings: Elevated levels of serum IgG4 (≥ 135 mg/dL)	X	✓	X	✓
VII. Pathological findings among i-v) listed below:				
i. Marked lymphoplasmacytic infiltration and fibrosis				
ii. More than 10 IgG4-positive plasma cells per high-power microscopic field				
iii. Storiform fibrosis				
iv. Obliterative phlebitis				
v. No neoplastic cells identified				
a. i), ii), and v) are observed	X	X	X	X
b. v) is observed	✓	✓	✓	X
c. All of i), ii), and v) and either or both of iii) or iv) are observed	X	X	X	X
VI. Other organ involvement (OOI):				
a. Type 1 autoimmune pancreatitis	✓	✓	X	X
b. IgG4-related dacryoadenitis/sialadenitis (Mikulicz disease), IgG4-related retroperitoneal fibrosis, IgG4-related kidney lesion	X	X	X	X
VI. Effectiveness of steroid therapy	✓	✓	X	✓
Diagnosis	Definite	Definite	Definite	Possible

Table 1. showing the characteristics of the IgG4-RSC cases according to the Japanese Clinical Diagnostic Criteria for IgG4-RSC 2020

References:

Nakazawa, T., Kamisawa, T., Okazaki, K., Kawa, S., Tazuma, S., Nishino, T., Inoue, D., Naitoh, I., Watanabe, T., Notohara, K. and Kubota, K., 2021. Clinical diagnostic criteria for IgG4-related sclerosing cholangitis 2020: (Revision of the clinical diagnostic criteria for IgG4-related sclerosing cholangitis 2012). Journal of Hepato-Biliary-Pancreatic Sciences, 28(3).

Results

- Four patients were identified.
 - Three were male.
 - Median age: 64 (44-68) years old.
- All patients had deranged LFTs on presentation.
 - Median bilirubin level = 155.5 umol/L (11.5 - 223.8 umol/L).

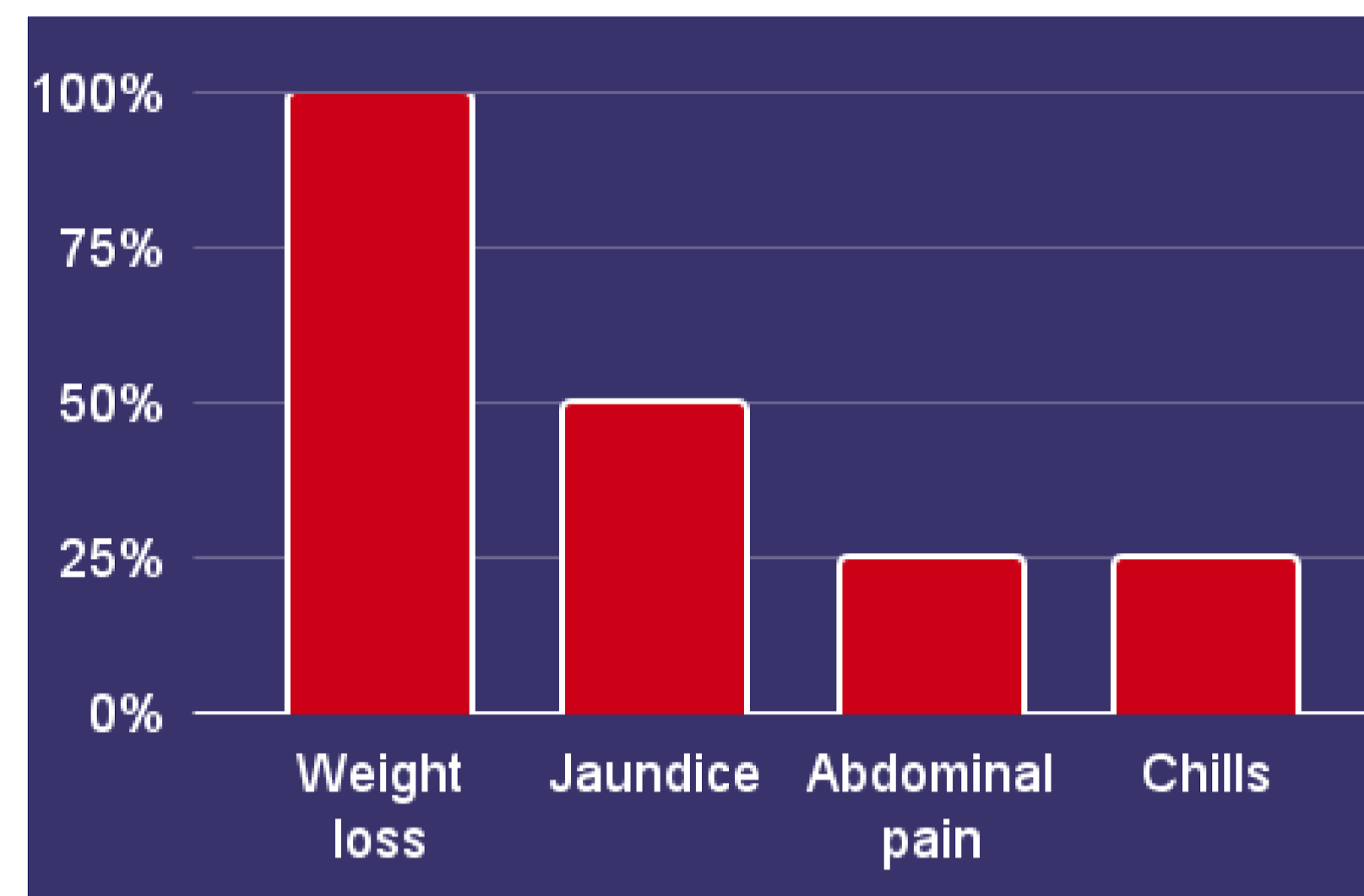


Figure 1. showing the presenting symptoms for the IgG4-RSC cases

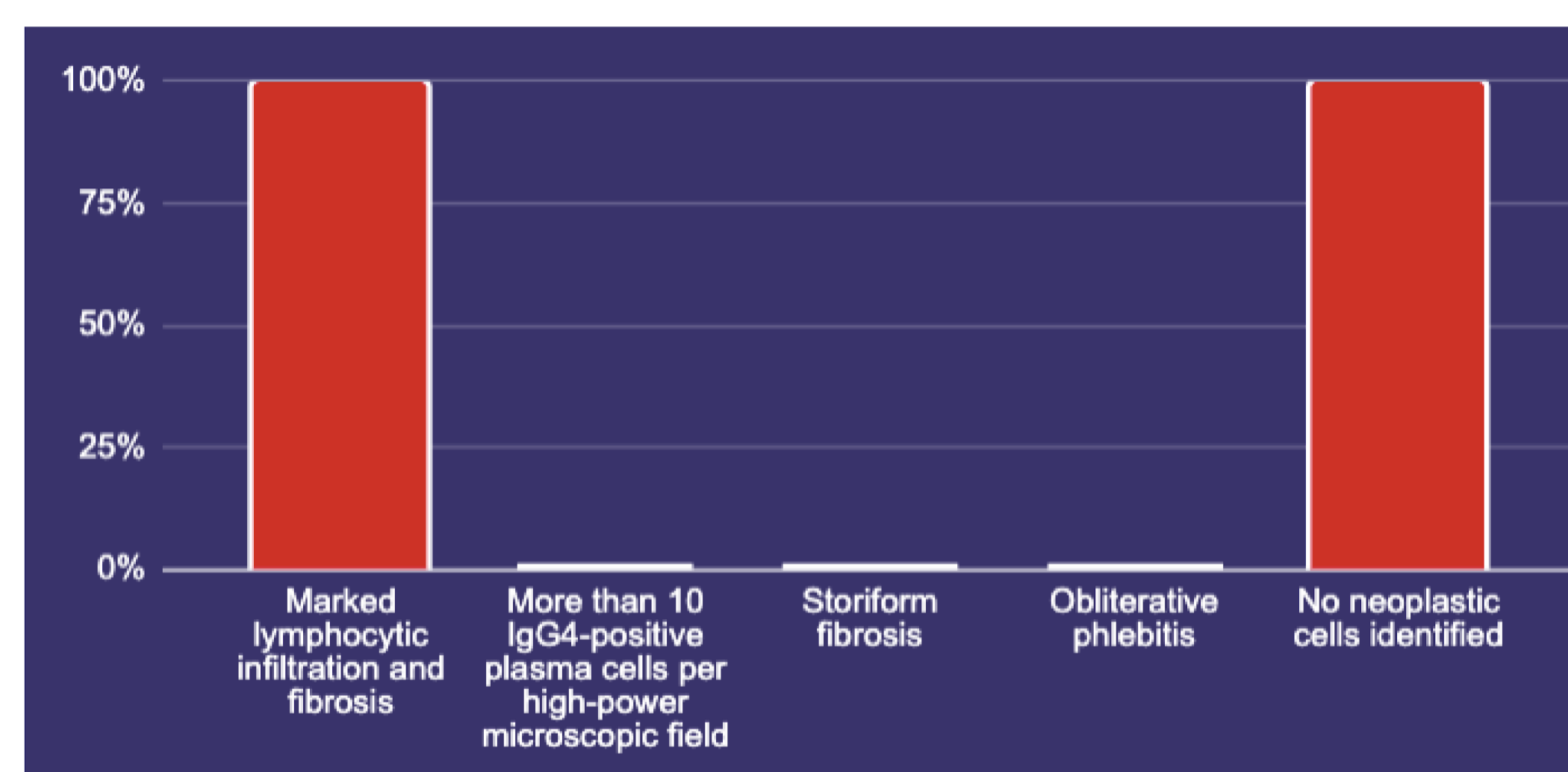


Figure 2. showing the pathological findings of intraductal biopsies based on the Japanese Clinical Diagnostic Criteria for IgG4-RSC 2020 obtained from the IgG4-RSC cases

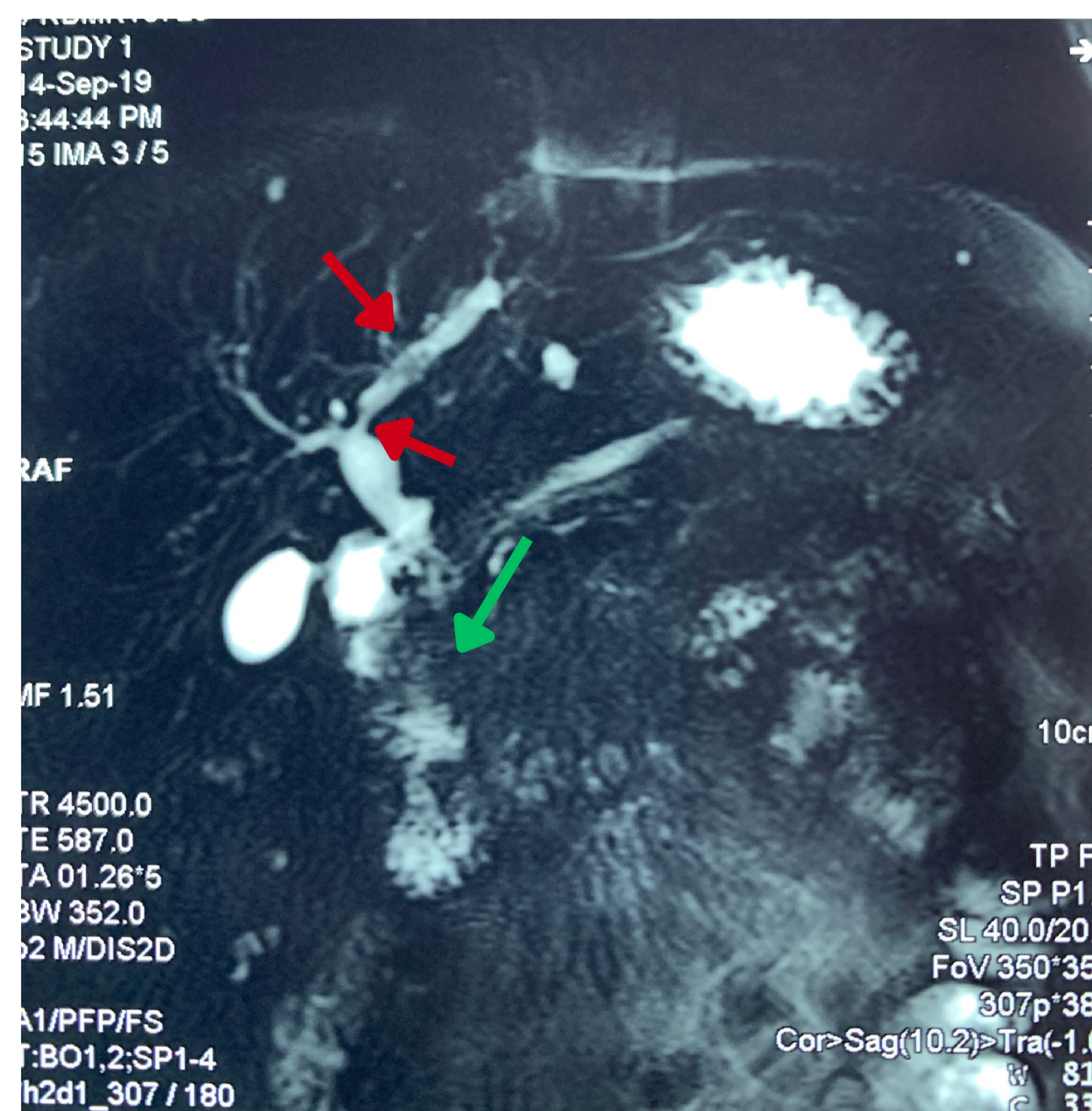


Figure 3. showing MRCP of Case 1: Red arrows pointing to obvious stricture at junction of left intrahepatic duct and common bile duct, and irregularities within the left hepatic duct. The common hepatic duct is also dilated. Green arrow showing CBD stricture.

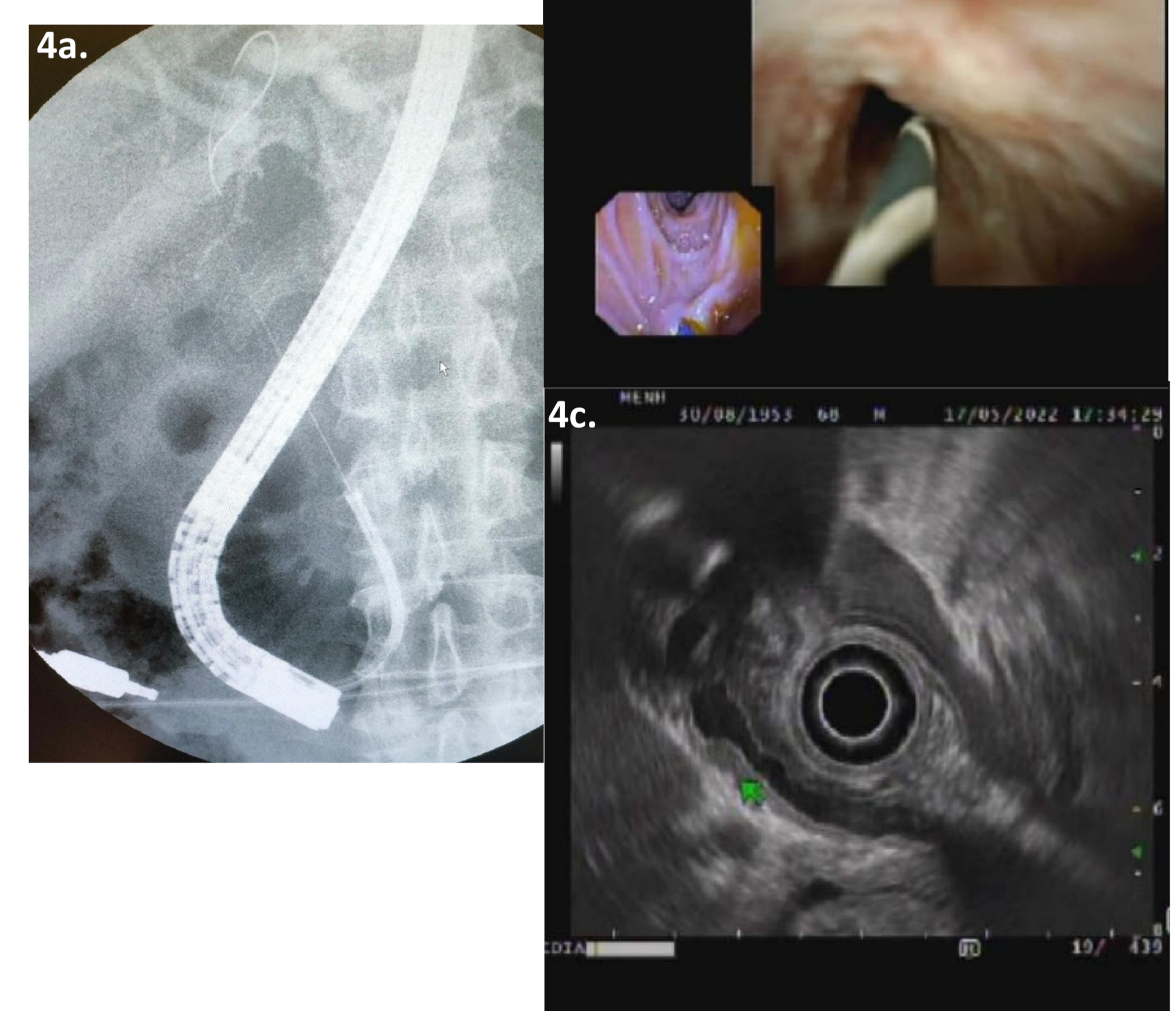


Figure 4. showing (a) ERCP with SpyGlass cholangioscopy. (b) Cholangioscopy showing a distal CBD stricture, absence of neovascularisation, mass lesion and friability. (c) EUS showing diffuse hypoechoic thickening of the bile duct throughout its entire length.

Conclusion

Early recognition of the signs and symptoms is important as IgG4-RSC carries a good prognosis if diagnosed early, with early clinical, biochemical and radiological improvement.

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