



Immunoglobulin G4- associated cholangitis mimicking cholangiocarcinoma in Elderly

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Background

Sclerosing cholangitis is a diffuse inflammation and fibrosis that progressively leads to stenosis and destruction of the bile ducts. It includes three types: primary sclerosing cholangitis (PSC), secondary cholangitis, and IgG4 sclerosing cholangitis (IgG4-SC). Usually they all present with cholestatic symptoms like pruritus and abdominal pain along with elevated ALP and GGT as well as bilirubin, however IgG4-SC has better outcomes, treatment response to steroids and less recurrence.

Case Description/Methods

- An 80 year-old male with a past medical history of bladder cancer was referred to the gastroenterology for EUS after blood work showed (ALT 320, AST 297, Bilirubin 1.4, GGT 2032), and CT abdomen with IV contrast revealed intra and extrahepatic biliary dilatation with an abrupt cutoff in mid CBD, suspicious for either a primary CBD mass or pancreatic mass (Figure 1).
- EUS showed an irregular mass in the CBD wall, suspicious for cholangiocarcinoma (Figure 1). The ERCP showed malignant appearing stricture of the extrahepatic bile duct consistent with cholangiocarcinoma, Bismuth 2 vs early 3B (Figure 1).
- Exploratory laparotomy was done and portal node biopsy came back benign; for which a repeat ERCP/Spyglass with direct visualization cholangioscopy, was done and showed diffusely dilated main bile duct with sludge and pus, single diffuse stenosis in right hepatic duct sludge with lining malignant appearing abnormal mucosa (nodularity and dilated vessels).
- IgG4 was elevated (931) consistent with IgG4 cholangitis (Figure 1). Biopsy and cytology brush showed no signs of dysplasia or neoplasia. Patient was then started on prednisone for a month and scheduled to repeat ERCP/Spyglass after.

References/Acknowledgements

1. Kahaleh M, Gaidhane M, Shahid HM, et al. Digital single-operator cholangioscopy interobserver study using a new classification: the Mendoza Classification (with video). *Gastrointest Endosc.* 2022;95(2):319-326. doi:10.1016/j.gie.2021.08.015

The authors would like to thank the patient for allowing us to share this case with our colleagues.

Results

Test Name	Reading	Reference Range
IgG4	931	2 - 96
Alkaline Phosphatase	888	34 - 104
AST	108	13 - 39
ALT	97	7 - 52
Protein Total	6.9	6.4 - 8.4
Albumin	3.2	3.5 - 5.7
Total Bilirubin	3.2	.3 - 1.1
Direct Bilirubin	1.7	0.00 - 0.20
Ammonia level	58	16 - 53
T4	1.16	.61 - 1.12
TSH	2.14	.45 - 5.3
CEA	1.9	0.00 - 10.0
CA 19-9	184	0 - 35
PT	15.1	12.2 - 14.9
INR	1.2	Below 1.1

Table 1. Laboratory studies indicating IgG4 cholangitis

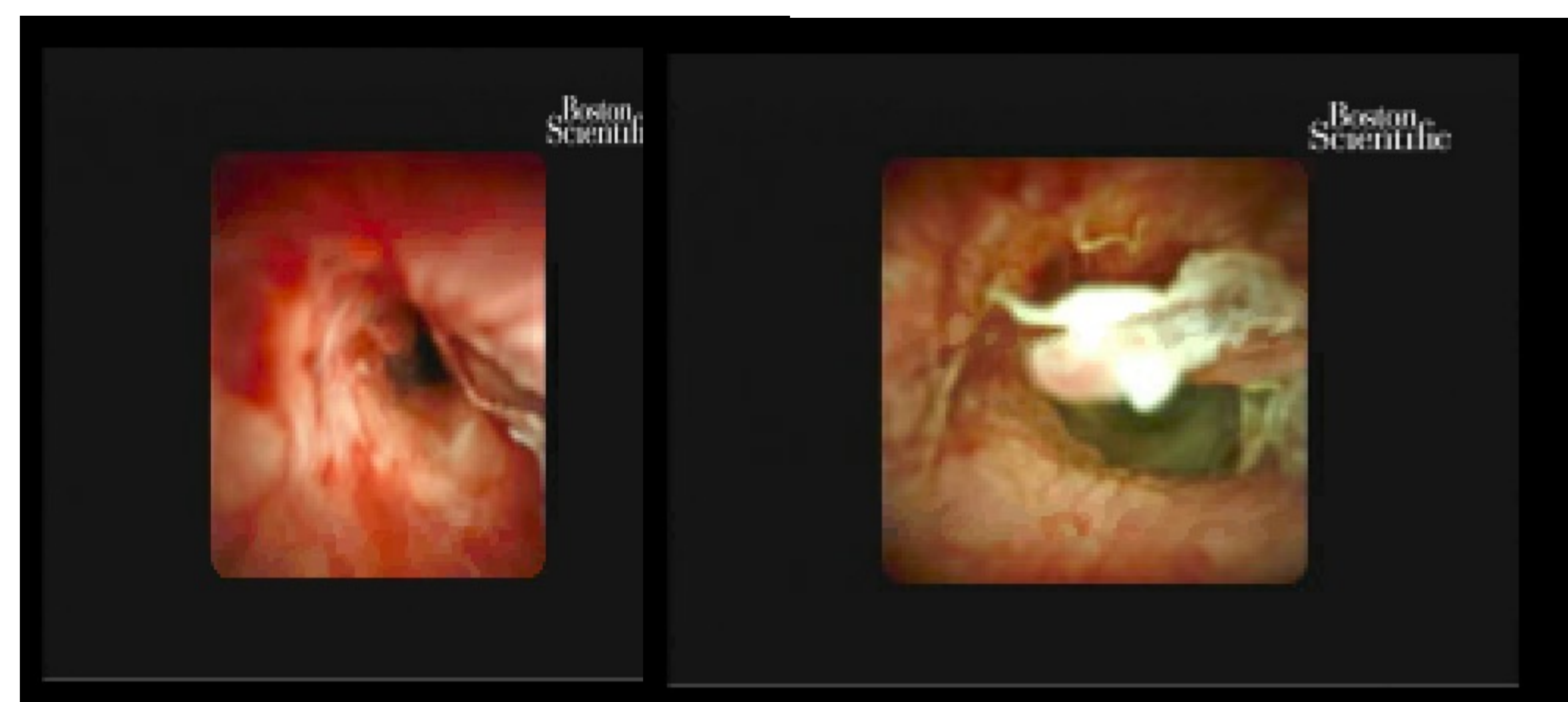


Fig 1. ERCP/Spyglass showing diffuse dilated bile duct with sludge and pus with malignant appearing abnormal mucosa.

Results (continued)

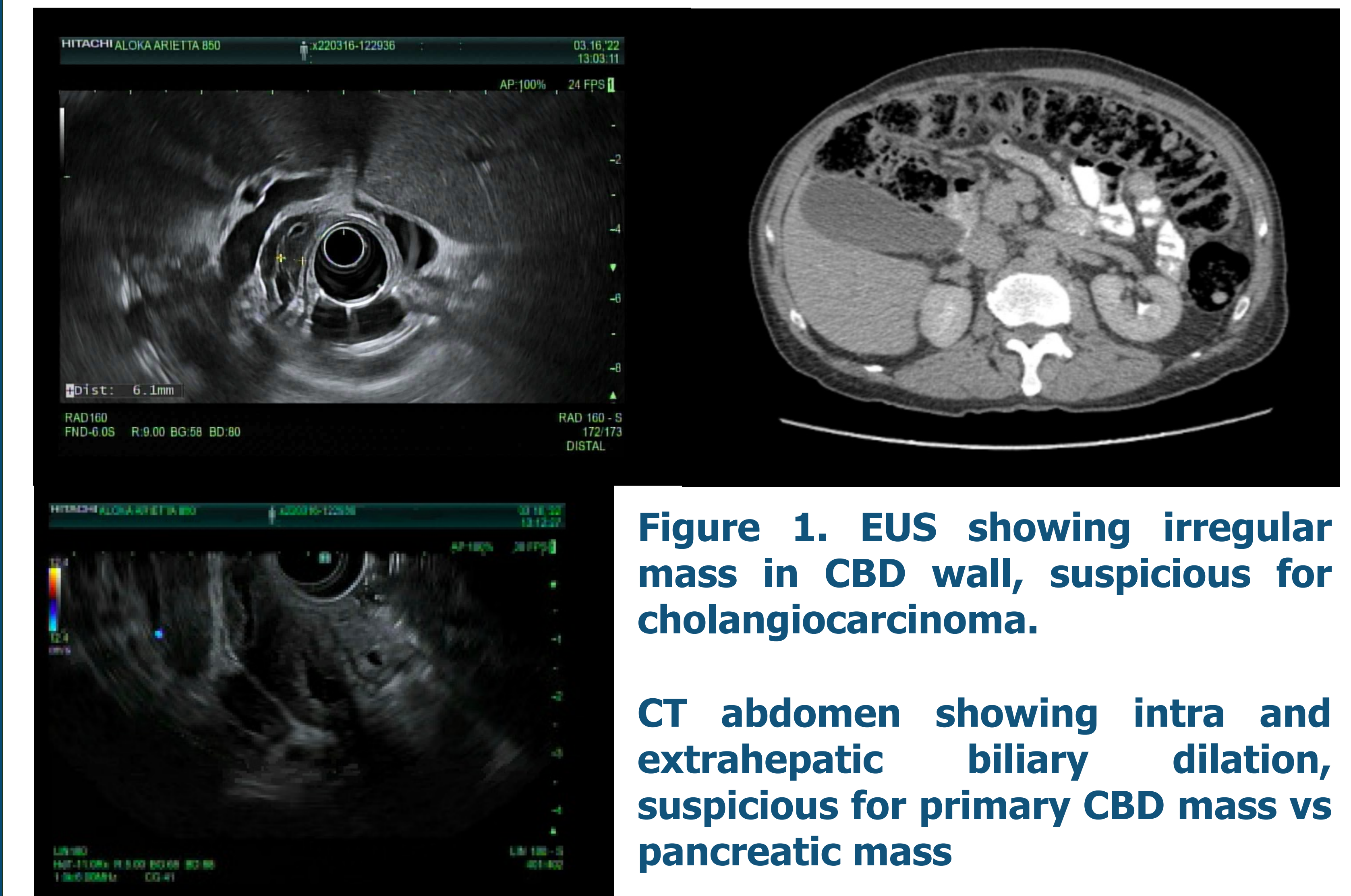


Figure 1. EUS showing irregular mass in CBD wall, suspicious for cholangiocarcinoma.

CT abdomen showing intra and extrahepatic biliary dilatation, suspicious for primary CBD mass vs pancreatic mass

Discussion

- IgG4-RD is a fibrous-inflammatory process related to immunomodulation, therefore IgG4-SC often co-presents with auto-immune pancreatitis, dacryoadenitis and sialadenitis.
- IgG4-SC is extremely challenging, as it's frequently misdiagnosed on imaging as a pseudotumor, and can progress to liver cirrhosis if left untreated, however it comes with great response to steroids.
- In 2021, Mendoza criteria (tortuous vessels, irregular nodulations, raised intraductal lesions, irregular surface, and friability) was published and found to be accurate to differentiate malignant and benign causes of biliary stricture, with overall diagnostic accuracy of 77%. Our case didn't meet the 5 criterias of Mendoza, which is consistent with IgG4-SC.