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CASE REPORT

An atypical case of isolated immunoglobulin G4-related sclerosing cholangitis with a cholangiogram resembling primary sclerosing cholangitis

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Abstract
An asymptomatic 77-year-old man with intrahepatic bile duct dilation was referred to our hospital. Cholangiography revealed alternations between strictures and dilated segments from the right and left hepatic ducts to the lower bile ducts, with findings of a pruned tree, beaded, shaggy appearance, and diverticulum-like outpouching. Histopathology revealed abundant immunoglobulin G4 (IgG4)-positive plasma cells (> 10 per high-power field) with an IgG4/IgG-positive cell ratio of 40–50%. After 2 weeks of steroid therapy, the cholangiography markedly improved. Because the cholangiographic findings resembled those of primary sclerosing cholangitis, steroid therapy proved useful in differentiating IgG4-related sclerosing cholangitis from primary sclerosing cholangitis.

Keywords Isolated IgG4-related sclerosing cholangitis · Primary sclerosing cholangitis · Endoscopic retrograde cholangiography · Intraductal ultrasonography · Steroid therapy

Introduction

Immunoglobulin G4-related disease (IgG4-RD) is an autoimmune disease of unknown pathogenesis characterized by the infiltration of lymphocytes and IgG4-positive plasma cells in systemic organs. IgG4-related sclerosing cholangitis (IgG4-SC) is a biliary manifestation of IgG4-RD [1]. IgG4-SC has a variety of cholangiographic features that are difficult to distinguish from primary sclerosing cholangitis (PSC) or cholangiocarcinoma (CCA) [2]. IgG4-SC responds well to steroid therapy and has good long-term outcomes, with rare progression to cirrhosis and the development of hepatobiliary malignancies [3]. PSC is a chronic progressive disease with a poor prognosis that leads to cirrhosis and ultimately requires liver transplantation [4]. CCA is treated with a combination of surgery and chemotherapy, but its mortality rate remains high [5]. Accurate diagnosis is important because IgG4-SC and its mimics require different treatment strategies and have different long-term prognoses.

Endoscopic retrograde cholangiography (ERC), intraductal ultrasonography (IDUS), and bile duct biopsy are useful for diagnostic evaluation of bile duct stenosis. Long smooth bile duct stenosis on ERC and concentric bile duct wall thickening with smooth inner and outer layers on IDUS are characteristic findings of IgG4-SC [6]. Pathological tissues showing dense infiltration of lymphocytes/plasma cells and fibrosis, an IgG4/IgG ratio > 40%, > 10 IgG4-positive cells per high-power field (HPF), storiform fibrosis, or obliterative phlebitis are accepted histological diagnostic criteria for IgG4-SC [7]. However, some patients show atypical cholangiograms [8, 9] and the sensitivity of bile duct biopsy for the definitive diagnosis of IgG4-SC is low [10]. A comprehensive diagnosis based on imaging, serological, and histological findings is required. However, in the absence of characteristic findings, the response to a trial of steroid therapy may be useful in diagnosing IgG4-SC.

We report a case of isolated IgG4-SC that presented with an atypical cholangiogram resembling PSC, and the successful use of steroid therapy for differentiation.

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Case report

A 77-year-old man with no remarkable medical history was referred for intrahepatic bile duct dilation incidentally detected on routine abdominal ultrasonography screening during a medical checkup. He was asymptomatic and had never been treated for biliary tract lesions or developed biliary tract infections. Laboratory data were unremarkable other than slightly elevated γ-glutamyl transpeptidase (71 IU/L; normal range, 10–47 IU/L), carcinoembryonic antigen (8.6 ng/mL; normal range, 0–5.0 ng/mL), and serum IgG (1707 mg/dL; normal range, 870–1700 mg/dL) and IgG4 (134 mg/dL; normal range, 4.8–105 mg/dL) levels. Contrast-enhanced computed tomography revealed intrahepatic bile duct dilation and enhanced extrahepatic bile duct wall thickening (Fig. 1a). No obvious extra-bile duct or mass lesions causing bile duct stenosis or pancreatic enlargement were observed (Fig. 1b). Magnetic resonance cholangiopancreatography (MRCP) revealed segmental stenosis and dilatation from the intrahepatic ducts to the extrahepatic bile ducts (Fig. 1c). Main pancreatic duct stenosis or dilation was unclear. Endoscopic ultrasonography showed a circumferential heterogeneous wall thickening of the extrahepatic bile duct. No abnormalities were observed in the pancreatic parenchyma (Fig. 1d). The ERC showed alternations between strictures and dilated segments from the right and left hepatic ducts to the lower bile ducts, with a pruned tree, beaded, shaggy appearance, and diverticulum-like outpouching (Fig. 2a). IDUS revealed asymmetric wall thickness and disappearance of the three-layer structures (Fig. 2b). PSC was most suspected based on these imaging findings. Endoscopic retrograde pancreatography was not performed. Transpapillary forceps biopsy of the stenotic lower bile duct was performed to exclude CCA, and an endoscopic nasobiliary drainage (ENBD) catheter was inserted to obtain bile cytology and prevent post-ERC cholangitis. He had no complications, such as cholangitis or pancreatitis. Cytological examinations were performed three times using the ENBD catheter, and no malignant cells were found. Histopathological analysis revealed almost normal epithelial cells with no inflammatory findings (Fig. 3a), high lymphoplasmacytic infiltration, fibrosis in the stroma (Fig. 3b), and abundant IgG4-positive plasma cells (> 10/HPF), with an IgG4/IgG-positive cell ratio of 40–50% (Fig. 3c, d). There was no evidence of storiform fibrosis, obliterative phlebitis, or neoplastic cells. Colonoscopy showed no abnormality. These findings suggested the possibility of IgG4-SC. Although he was asymptomatic,

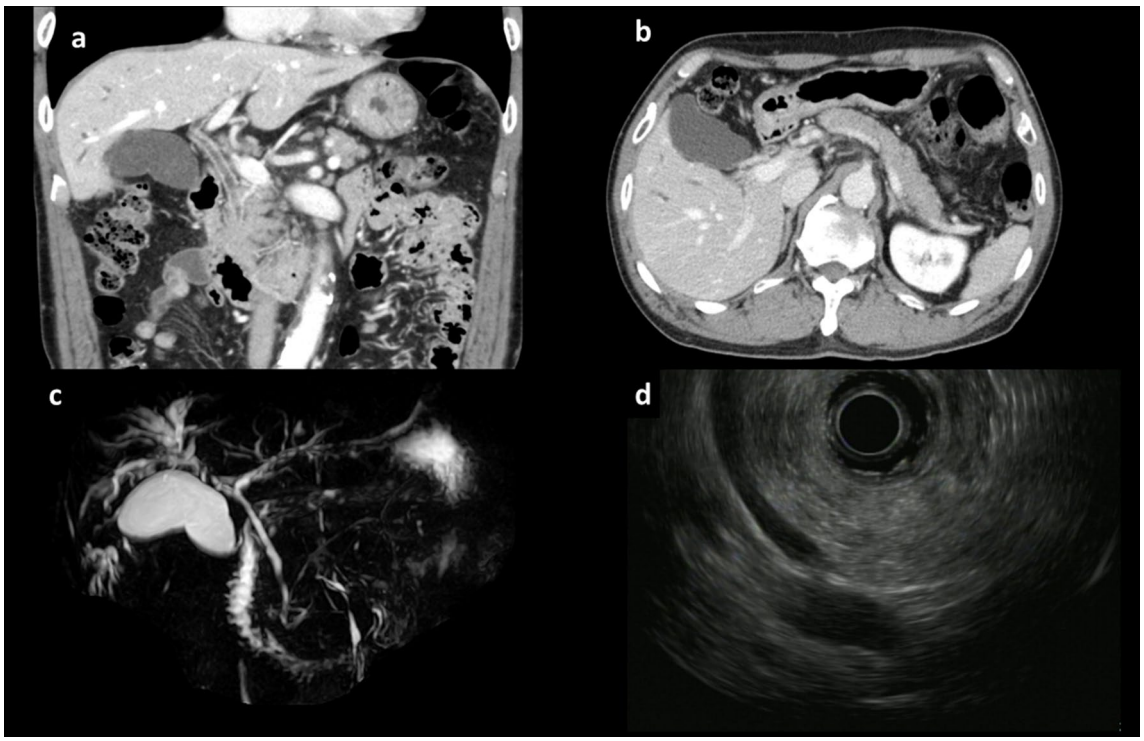

Fig. 1 **a** Intrahepatic bile duct dilation and enhanced wall thickening of the extrahepatic bile duct on contrast-enhanced computed tomography. **b** Contrast-enhanced computed tomography showing no obvious pancreatic enlargement. **c** Discontinuous stenosis and dilatation from the intrahepatic ducts to the common bile ducts on magnetic resonance cholangiopancreatography. **d** Endoscopic ultrasonography showing no evidence of diffuse hypoechogenicity in the pancreatic parenchyma

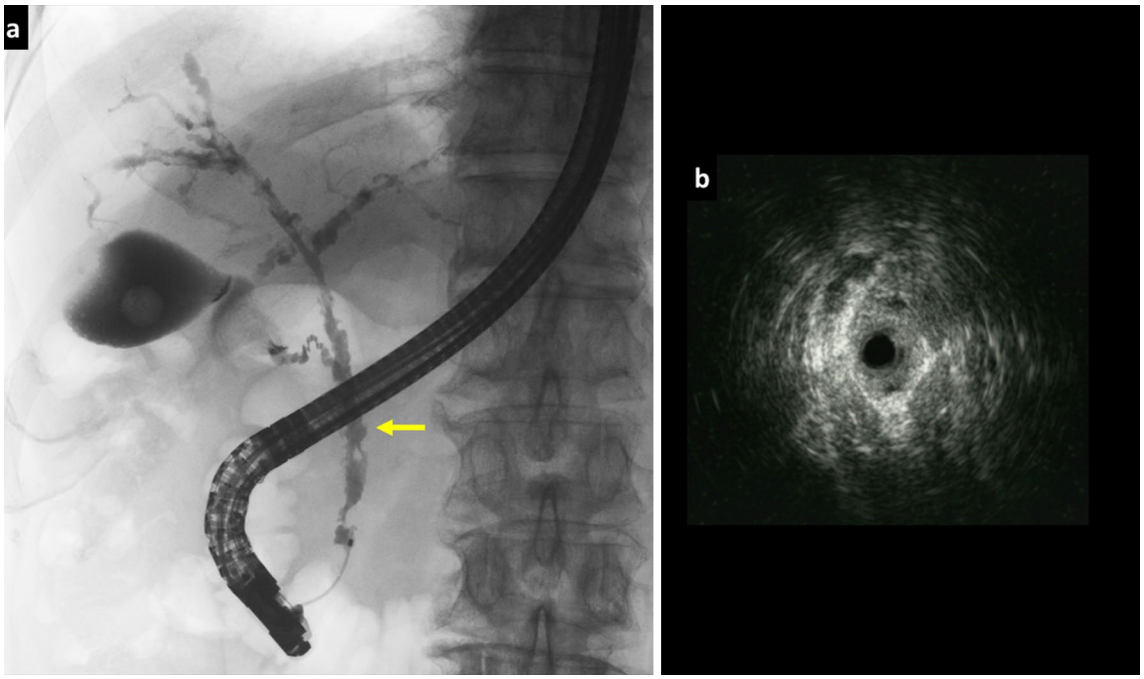

Fig. 2 **a** Alternations between strictures and dilated segments from the right and left hepatic ducts to the lower bile ducts, with pruned tree, beaded, shaggy appearance, and diverticulum-like outpouching findings on endoscopic retrograde cholangiography. **b** Intraductal ultrasonography was inserted up to the yellow arrow portion. Asymmetric wall thickness and disappearance of the three-layer structures of the lower bile duct

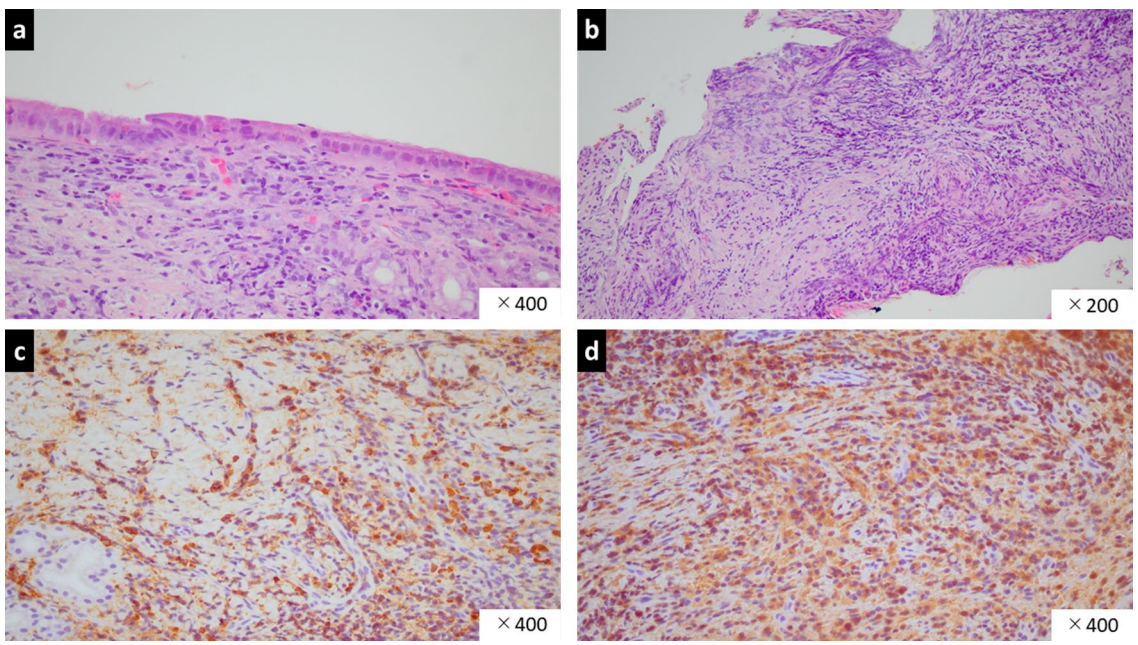

Fig. 3 **a** Hematoxylin and eosin staining (×400) of samples obtained from bile duct biopsies shows normal bile duct epithelial cells with no inflammatory findings. **b** Hematoxylin and eosin staining (×200) of samples obtained from bile duct biopsies shows high lymphoplasmacytic infiltration and fibrosis in the stroma. **c** IgG4 immunostaining (×400) showing many IgG4-positive plasma cells (> 10 per high-power field), with an IgG4/IgG-positive cell ratio of 40–50%. **d** IgG immunostaining (×400) showing many IgG-positive plasma cells, with an IgG4/IgG-positive cell ratio of 40–50%

he was treated with 30 mg/day of oral prednisolone to differentiate between PSC and IgG4-SC. After 2 weeks, cholangiography showed some residual bile duct stricture, but improvement in most areas (Fig. 4a). There was also normalization of γ-glutamyl transpeptidase and IgG4 levels. He responded well to steroids and was finally diagnosed with IgG4-SC rather than PSC. The steroid dose was gradually tapered to 5 mg/day over 6 months and the medication was continued. Six months after starting steroid therapy, the findings of the pancreatic parenchyma on computed tomography remained unchanged from those observed before the steroid induction (Fig. 4b). MRCP performed approximately 6 years after the diagnosis showed worsening dilation of the perihilar and intrahepatic bile ducts (Fig. 5); however, he had no hepatobiliary adverse events (cholangitis, cirrhosis, or CCA). Blood samples were collected once a year for 7 years after steroid therapy, with no evidence of elevated hepatobiliary enzymes or progression to liver dysfunction (Table 1). To date, he has been followed up with maintenance therapy.

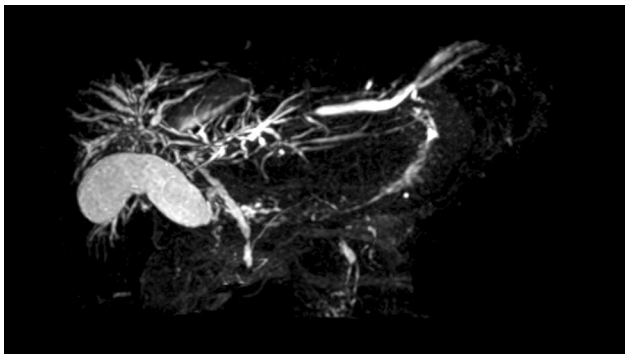

Fig. 5 Approximately 6 years after the initial diagnosis of IgG4-related sclerosing cholangitis, magnetic resonance cholangiopancreatography showed worsening of stenosis of the perihilar and intrahepatic bile ducts

Discussion

IgG4-SC has recently been established as a subgroup of sclerosing cholangitis [11] and is defined as progressive stenosis and destruction of the bile ducts caused by diffuse inflammation and fibrosis. IgG4-SC often presents clinical findings similar to those of CCA and PSC. Correctly distinguishing IgG4-SC is especially important in patients with hilar or intrahepatic bile duct stenosis and dilation because surgery can be avoided in such patients. Cholangiograms of IgG4-SC are classified into four subtypes [12]. The most common cholangiogram pattern is stenosis confined to the intra-pancreatic bile duct (type 1), which may be part of autoimmune pancreatitis and should be distinguished from chronic pancreatitis or pancreatic cancer. Intrahepatic segmental (type 2a) and diffuse (type 2b) stenosis, in addition to intra-pancreatic bile duct stenosis, imitates PSC. Stenosis of both intra-pancreatic and hilar bile ducts (type 3) or only the hilar bile ducts (type 4) must be distinguished from CCA.

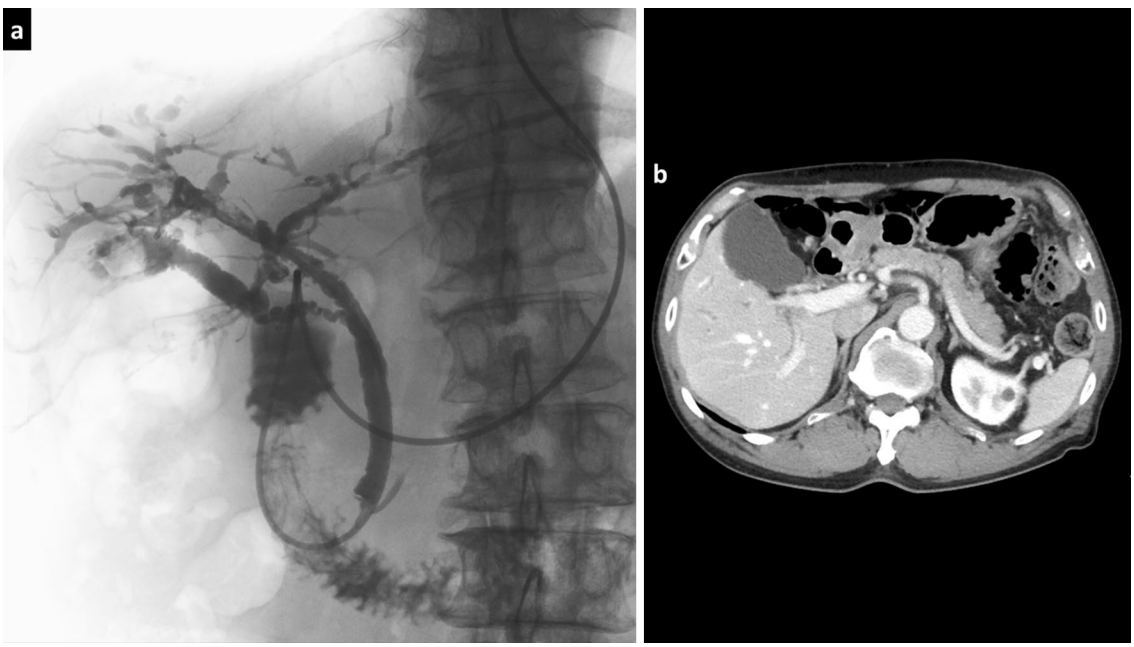

Fig. 4 **a** Improvement of bile duct stricture is observed on a cholangiogram after 2 weeks of steroid therapy. **b** Contrast-enhanced computed tomography performed 6 months after the start of steroid therapy showing no pancreatic enlargement or atrophy

Table 1 Findings of blood sampling before and after steroid treatment

Blood collection items	Before steroid	After initiation of steroid therapy						
		1 Year	2 Year	3 Year	4 Year	5 Year	6 Year	7 Year
WBC (/×10^3)	5.1	5.9	7.9	6.3	6.6	7.0	5.7	8.3
HGB (g/dL)	13.4	13.2	12.8	13.8	13.1	13.0	13.4	12.5
PLT (/×10^3)	176	259	216	199	157	207	177	166
AST (U/L)	17	16	16	17	23	16	17	16
ALT (U/L)	14	12	11	14	23	12	15	17
ALP (U/L)	261	271	234	233	246	224	244	287
γ-GTP (U/L)	71	23	24	40	91	38	37	46
T-B (mg/dL)	1.3	0.9	1.0	1.0	1.1	1.2	1.6	1.4
ALB (g/dL)	4.1	3.9	3.6	4.0	3.9	4.1	4.2	3.9
CEA (ng/mL)	8.6	–	9.7	9.5	9.8	8.6	9.7	9.1
CA19-9 (U/mL)	1	–	1	1	1	1	1	1
IgG (mg/dL)	1707	1416	1401	–	–	1257	1223	1286
IgG4 (mg/dL)	134	97	–	–	–	78.5	77.5	67.1

WBC white blood cells, *HGB* hemoglobin, *PLT* platelet, *AST* aspartate aminotransferase, *ALT* alanine aminotransferase, *ALP* alkaline phosphatase, *γ-GTP* γ-glutamyl transpeptidase, *T-B* total bilirubin, *ALB* albumin, *CEA* carcinoembryonic antigen, *CA19-9* carbohydrate antigen 19–9, *IgG* immunoglobulin G, *IgG4* immunoglobulin G4

IDUS is useful for differentiation, and Naitoh et al. reported that irregular inner margins, diverticulum-like outpouching, and loss of the three-layer structure were specific findings for PSC [13], and that bile duct wall thickness of more than 0.8 mm in IDUS features of non-stricture regions on the cholangiogram was strongly suggestive of IgG4-SC [10]. However, it was difficult to suspect IgG4-SC if no typical findings on ERC or IDUS were observed and no other organs were involved. In this case, the cholangiogram suggested PSC was not classified as any type of IgG4-SC. Assuming that the present case is PSC, the absence of elevated serum alkaline phosphatase levels and no evidence of inflammatory bowel disease on colonoscopy led to a probable diagnosis according to the clinical guidelines for primary sclerosing cholangitis 2017. On the other hand, clinical diagnostic criteria for IgG4-related sclerosing cholangitis 2020 stated that cases with stenosis of the extrahepatic bile ducts, excluding the so-called intra-pancreatic bile ducts, can be diagnosed according to the type 4 IgG4-SC. Therefore, we considered that the diagnostic criteria for probable IgG4-SC were fulfilled with the narrowing of the bile ducts by ERC (Ia), thickening of the bile duct wall by IDUS (IIa), and pathological findings (IVa). Since he met the diagnostic criteria for probable diagnosis in both PSC and IgG4-SC, steroid therapy was performed as diagnostic treatment, and finally diagnosed IgG4-SC.

Some reports have used peroral cholangioscopy (POCS) to differentiate IgG4-SC from PSC, with dilated or tortuous vessels in IgG4-SC, bile duct scarring, and pseudodiverticula as characteristic findings of PSC [14]. Furthermore, POCS-guided biopsies are more sensitive than trans-papillary biopsies [15]. POCS is useful in identifying biliary strictures with an indeterminate diagnosis but is known to have a higher complication rate than endoscopic retrograde cholangiopancreatography (ERCP) [16]. We generally do not perform endoscopic sphincterotomy (EST) in patients with biliary strictures because it increases the risk of retrograde cholangitis. In the present case, EST and subsequent POCS were not performed because ERCP at the time of the initial examination showed a bile duct typical of PSC.

Bile duct biopsies are commonly performed to diagnose biliary strictures. In the diagnosis of IgG4-SC, the sensitivity of bile duct biopsy with IgG4 immunostaining has been reported to be inconsistent (0–88%) [10, 17, 18] because the histopathological features of IgG4-SC are lymphoplasmacytic infiltration and fibrosis in the bile duct stroma, whereas the bile duct epithelium is often normal. It is difficult to obtain large samples containing bile duct stroma endoscopically; therefore, the main role of bile duct biopsy is to exclude CCA. In contrast, the sensitivity of bile duct biopsy in the diagnosis of malignant biliary strictures is 48–64% [19–21]. Hence, if biopsy fails to reveal the characteristic findings of IgG4-SC or CCA, the diagnosis remains indeterminate. For an accurate diagnosis, re-examination is recommended when clinical and histological diagnoses are discordant [7]. In the present case, a bile duct biopsy was performed to collect a sample that fulfilled the diagnostic criteria for IgG4-SC. Yamamoto et al. [22] reported that the sensitivity of biopsy varies depending on the location of the stricture because strictures that do not provide sufficient space for open biopsy forceps make it difficult to perform a proper and accurate targeted biopsy. The cholangiogram

of our patient displayed short stenosis and the bile duct epithelium was asymmetric and irregular, which allowed us to open the biopsy forceps sufficiently to press them against the bile duct wall and obtain a sample that contributed to the diagnosis.

Steroid therapy is the standard treatment for IgG4-SC and helps to rule out PSC and CCA. A nationwide survey in Japan reported remission in 99.5% of IgG4-SC patients treated with steroids [23]. In contrast, steroid therapy for PSC is not recommended because it has a low efficacy rate of 3.7% and significantly reduces bone density [24]. These characteristics are useful for differentiating IgG4-SC from PSC [25], and the 2020 clinical diagnostic criteria for IgG4-SC incorporate them as diagnostic criteria, emphasizing their importance [7]. Responsiveness to steroid therapy should be evaluated using MRCP or ERC within as short a period as possible [26]. Steroid pulse therapy is an effective initial alternative to oral steroid therapy for autoimmune pancreatitis. This surpasses oral steroid therapy in its therapeutic effect on bile duct lesions [27]. In contrast, steroid pulse therapy cannot prevent recurrence. Ikeura et al. [28] reported that the recurrence rate at 3 years after steroid treatment was significantly higher for steroid pulse monotherapy than for oral steroids. Therefore, in Japan, maintenance therapy with oral steroids is recommended for 3 years; however, the discontinuation criteria remain unclear [29]. In the present case, cholangiography 2 weeks after the initiation of oral steroid therapy showed slight residual stenosis of the intrahepatic bile ducts. It has been reported that proximal and intrahepatic bile duct structures are less responsive to steroid therapy than distal bile structures [26, 30], and steroid pulse therapy may be a better option for remission induction therapy.

Interestingly, despite maintenance therapy, the most recent MRCP showed worsening of the biliary stricture, possibly due to IgG4-SC recurrence or PSC complications. Elevated alkaline phosphatase levels in PSC and IgG4 levels in IgG4-SC are considered characteristic blood collection findings; however, even normal levels do not rule out a diagnosis. Considering the long-term course of PSC or IgG4-SC, clinicians should be aware of the development of CCA. PSC is associated with a high risk of CCA, especially when diagnosed within the first year of diagnosis of PSC [31]. In some cases, higher liver biochemical dysfunction [32] and progressive bile duct changes on MRCP [33] during the course of PSC may be important findings that may indicate the potential development of CCA. However, the prognosis of patients with IgG4-SC is good, and the incidence of CCA is comparable to that in the general population [34, 35]; therefore, IgG4-SC is not currently considered a risk factor for cholangiocarcinoma [29]. In the present case, the cause of the worsening bile duct stricture was unknown. Further examination including ERCP and POCS might be recommended for differential diagnosis. However, the patient remained asymptomatic for a long time, and blood samples, including tumor markers, did not change. Therefore, we decided to carefully follow up the patient, considering the disadvantages of performing ERCP and POCS. One year after MRCP imaging, his blood samples remained unchanged, and we continue to follow up regularly.

In conclusion, when diagnosing atypical IgG4-SC with findings of a pruned-tree, beaded, shaggy appearance and diverticulum-like outpouching in ERC, steroid therapy is a useful option to distinguish it from PSC.

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Declarations

Conflict of interest There are no conflicts of interest to declare.

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