Abstract
A 37-year-old woman presented to our hospital with jaundice. Clinical results were as follow: serum total bilirubin concentration, 16 mg/dL; serum direct bilirubin, 13 mg/dL; serum alkaline phosphatase, 1,781 IU/L; and \(\gamma\)-glutamyltranspeptidase, 399 IU/L. Abdominal computed tomography (CT) revealed expansion from the common bile duct to the intrahepatic bile ducts of both hepatic lobes and a solid tumor in a multilocular cyst of the left hepatic lobe with neighboring lymphadenopathy. Magnetic resonance cholangiopancreatography and endoscopic retrograde cholangiopancreatography revealed choledocholithiasis in the lower bile duct. The biliary cytology was class I. The multilocular cyst was reduced in size after biliary drainage, but CT revealed a residual solid tumor with a contrasting effect. These findings indicated a intraductal papillary neoplasm of the bile duct (IPNB) or cholangiocarcinoma. Left lobectomy with lymph nodes dissection was performed. Pathological findings indicated no neoplastic lesion in the expanded bile duct; however Xanthogranulomatous inflammation throughout the neighboring fat tissue was observed. These results indicated, Xanthogranulomatous cholangitis (XGC), which is rare. Reports suggest that differentiation between cholangiocarcinoma and bile duct stenosis is needed because of xanthogranulomatous cholecystitis. Reports on case of XGC with a cystic lesion that requires differentiation from IPNB, are extremely rare.

Case report
A 37-year-old woman with several weeks’ history of obstructive jaundice was admitted to our hospital. Physical examination revealed palpebral conjunctival jaundice and mild right higher-quadrant tenderness with a palpable tumor lesion. The results of a complete blood count were as follows: white blood cells, 16,600/\(\mu\)L; hemoglobin, 12.2 g/dL; and platelet count, 442,000/\(\mu\)L. Other laboratory results included the following: serum total bilirubin, 16.0 mg/dL; serum direct bilirubin, 13.0 mg/dL; serum alkaline phosphatase, 1,781 IU/L; \(\gamma\)-glutamyltranspeptidase, 399 IU/L; serum carcinoembryonic antigen and carbohydrate antigen 19–9 levels, within normal limits; antimitochondrial antibody, negative; serum immunoglobulin G (IgG), 1,269 mg/dL; and antinuclear antibody, <40 U. Abdominal computed tomography (CT) revealed dilatation from the common bile duct to the left intrahepatic bile duct (Fig. 1a). A multilocular cystic lesion 9 cm in size that projected from

Key words: xanthogranulomatous cholangitis, IPNB

Introduction
Xanthogranulomatous inflammation (XGI) can occur in the gallbladder, skin, reproductive glands, gastrointestinal tract, and pancreas. However, xanthogranulomatous cholangitis (XGC) is rare. Recent reports suggest that differentiation between cholangiocarcinoma and bile duct stenosis is needed because of xanthogranulomatous cholecystitis. Reports on case of XGC with a cystic lesion that requires differentiation from IPNB, are extremely rare.
the left lobe of the liver was detected via CT and magnetic resonance cholangiopancreatography (Fig. 1b). Wall thickening and a solid tumor in the cyst were observed (Fig. 1c). However, whether a fistula existed between the left intrahepatic bile duct and the cyst was unclear. A choledocholithiasis approximately 15 mm in size was detected in the lower bile duct (Fig. 2) and confirmed via endoscopic retrograde cholangiopancreatography (ERCP). Thus, endoscopic sphincterotomy was performed. Biliary cytological examination results indicated class I diagnosis. No malignancy was diagnosed on the basis of the biliary cytological examination results. The multilocular cystic lesion of the left lobe of the liver was reduced after biliary drainage, but a solid tumor with a contrasting effect was observed on CT (Fig. 3). The dilatations of the intrahepatic and common bile ducts were also improved. These findings indicated cholangiocarcinoma or IPNB in the form of a solid tumor in the multilocular cyst. Left lobectomy with dissection of the lymph nodes was performed. When the choledocholithiasis was detected on cholangiography, a choledocholithotomy was also performed. On inspection, the resected specimen showed a soft yellowish mass; no cystic component was observed (Fig. 4). Microscopic examination revealed that a large number of lymphocytes had infiltrated the area surrounding the bile duct (Fig. 5a). Foamy macrophages, including lipid droplets, were observed (Fig. 5b). No cholangiocar-
cinoma or cystic component was observed. Therefore, XGC was diagnosed. No XGI was observed in the gallbladder. A diagnosis of XGC without xanthogranulomatous cholecystitis was made. The patient recovered well without serious complications.

**Discussion**

XGI can occur in the gallbladder, skin, reproductive glands, gastrointestinal tract and pancreas\(^1\). However, reports on cases of XGC are extremely rare. The pathophysiology of XGI involves a lesion of destructive inflammation that involves foamy macrophages, lymphocytes, and fibrous scars\(^2\).

Only three cases of XGI involving the bile duct have been reported. **Table 1** describes the three previously reported cases in comparison with our case. All of the cases were diagnosed after surgery. The first case involved a 67-year-old man who had had jaundice for 2 weeks and stenosis of the lower bile duct involving a mass lesion that was suspected of malignancy\(^3\). No choledocholithiasis was observed. The preoperative diagnosis was hilar bile duct carcinoma. The second case was of a 34-year-old woman with jaundice and stenosis of the upper to middle bile duct\(^4\). No mass lesion was found in the bile duct, but the biliary cytological examination result led to a diagnosis of class V adenocarcinoma. No choledocholithiasis was observed. The preoperative diagnosis was hilar bile duct carcinoma. The third case was of a 75-year-old woman with a histo-

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Fig. 3 After biliary drainage, the multilocular cystic lesion in the left hepatic lobe reducted. Dilatation of the intrahepatic bile duct and common bile duct also improved. Only a solid tumor is seen.

Fig. 4 The resected specimen shows a soft yellowish mass with no cystic component.

Fig. 5 a: A large number of lymphocytes infiltrate the bile duct area. No cholangiocarcinoma or cystic component is seen (hematoxylin and eosin [HE] stain × 40).

b: Foamy macrophages with lipid droplets are seen (HE × 200).
Xanthogranulomatous cholangitis

Table 1 Characteristics of xanthogranulomatous inflammation of the bile duct

|                | Ma J et al. (2013) | Kawate et al. (2006) | Liron et al. (2004) | Our case (2014) |
|----------------|-------------------|---------------------|---------------------|-----------------|
| Sex/Age        | M/67              | F/34                | F/75                | F/37            |
| Symptoms       | Jaundice          | Jaundice            | Cholangitis         | Jaundice        |
| Primary focus  | Lower bile duct   | Upper to middle bile duct | Left hepatic lobe | Left hepatic lobe |
| Characteristics| Stenosis, Mass lesion | Stenosis, Mass lesion | Dilatation, Cystic lesion | Dilatation, Cystic lesion |
| Pathology      | XGC               | XGC                 | XGC                 | XGC             |
| Xanthogranulomatous cholecystitis | -                | -                   | +                   | -               |

ry of recurrent cholangitis. A mass lesion was identified in the left hepatic lobe and was associated with selective dilatation, irregularity, and stenosis of the intrahepatic bile ducts. Choledocholithiasis was detected and removed from the bile duct by performing ERCP before surgery. The preoperative diagnosis was cholangiocarcinoma. Xanthogranulomatous cholecystitis was observed only in the third case. At this time, the association between XGC and xanthogranulomatous cholecystitis was unclear. In our case, dilatation of the bile duct due to choledocholithiasis and a solid tumor in a multilocular cyst were observed. Cholangiocarcinoma and IPNB are difficult to distinguish from each other. The origin of XGC is unknown, but the bile duct mucous membranes can be damaged when the internal pressure of the bile duct rises sharply due to obstructive jaundice or cholangitis. The origin of xanthogranulomatous cholecystitis has been indicated to be the increase in internal pressure in the gallbladder, such as by an impacted stone. The increase in the internal pressure of the gallbladder may result in damage to the mucous membranes of the gallbladder. Macrophages gourmandize bile elements of the gallbladder wall. Thus, based on the aforementioned considerations, XGI was thought to occur owing to xanthoma cells that consist of lipid and pigment. Speciation of the granulation tissue is expected to occur from 3 weeks to 6 months. As our case was evaluated a few weeks after the onset of jaundice, we can infer that the time for granulation tissue speciation to occur was enough. The cystic lesion identified in the left hepatic duct before surgery was not evident after EST and was not observed in the pathological analysis. Therefore, the dilatation of the left hepatic duct was probably not a cystic lesion but a localized biloma. Inflammation and dilatation of the bile duct that is localized to the left intrahepatic duct could not be clearly identified. The choledocholithiasis was considered to temporarily incarcerate in the left intrahepatic duct. Therefore, the internal pressure of left intrahepatic duct increased. In the case of our patient, the increase in the internal pressure of the left hepatic duct appeared to be due to choledocholithiasis. According to some reports, XGC has been identified as a stenotic lesion of the bile duct. In our case, the dilatation of the bile duct involved choledocholithiasis and was judged to be a cystic lesion in the left intrahepatic duct. Because a solid tumor was detected in a cystic lesion, our case required differentiation between IPNB and cholangiocarcinoma. In previous reports, differentiation between the two conditions was not required.

Conflict of interest: None.

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