febrile course and its response to symptomatic therapy is usually poor. The rash typically begins on the fourth to sixth day of the illness and consists of small, discrete macules of 1 to 10 mm which are generally noted first on the chest or abdomen, but which can be present later on the extremities. The rash may be transient, as in our patient, or persist for a few days. The response to the tetracycline group of antibiotic drugs is excellent, as in epidemic typhus, and it has been noted recently that even one dose of doxycycline can lead to prompt defervescence.[6] Therefore, therapy with a tetracycline drug should be initiated when classic features are present.

In contrast to epidemic typhus, Brill-Zinsser disease is not generally associated with a positive Weil-Felix test; for example, in our patient, the *Proteus* OX19 titer was only 1:20. The complement fixation test generally attains a maximum level by the 10th to 11th day of illness in contrast to epidemic typhus in which the maximum titers are not usually achieved until two weeks after onset. Therefore, in suspected cases it is important to obtain serum samples during the acute phase of illness for antibody testing to demonstrate a later rise in titer.

It was postulated by Murray and co-workers in 1950 that large numbers of cases of Brill-Zinsser disease might be found in this country due to the large influx of immigrants who had experienced typhus in Nazi concentration camps and elsewhere during World War II.[3] However, very few cases have been reported in North America in the 1960's and 1970's; the last case was reported in Canada in 1974.[5] Possibly, the rarity of recent reports of this disease is due partially to unfamiliarity with its manifestations and the lack, therefore, of attempts to make a specific diagnosis. We hope that this case report will stimulate continued attempts to diagnose Brill-Zinsser disease in the future.

Summary

A case of recrudescent typhus occurring 30 years after the initial bout of epidemic typhus is described. This disease should continue to be considered in the differential diagnosis of fever of undetermined origin in immigrants, particularly those from eastern Europe, from regions where typhus infection may have occurred in the past. The clinical diagnosis should be confirmed by appropriate serological tests.

REFERENCES

1. Brill ND: An acute infectious disease of unknown origin. Am J Med Sci 139:484-502, 1910
2. Zinsser H: Varieties of typhus virus and the epidemiology of the American form of European typhus fever (Brill's disease). Am J Hygiene 20:513-531, 1934
3. Murray ES, Baehr G, Schwartzman G, et al: Brill's disease. JAMA 142:1059-1066, 1950
4. Price WH: Studies on the interepidemic survival of louse-borne epidemic typhus fever. J Bacteriol 69:106-107, 1954
5. Portnoy J, Mendelson J, Clecner B: Brill-Zinsser disease: Report of a case in Canada. Can Med Assoc J 111:166, Jul 20, 1974
6. Perine PL, Krause DW, Awoke S, et al: Single-dose doxycycline treatment of louse-borne relapsing fever and epidemic typhus. Lancet 2:742-744, Sep 28, 1974

Refer to: Hart MJ, White TT: Choledochocele associated with acute hemorrhagic pancreatitis. West J Med 133:340-344, Oct 1980

Choledochocele Associated With Acute Hemorrhagic Pancreatitis

MICHAEL J. HART, MD
THOMAS TAYLOR WHITE, MD
Seattle

CHOLEDOCHAL CYSTS have been reported more than 1,000 times in the medical literature since the first one was described by Douglas in 1852.[1-4] However, the incidence of choledochoceles or type III choledochal cysts as classified by Alonso-Lej[5] is rare, and only 21 cases have been reported.[3,4,6-21] A patient with this type of lesion, which was associated with hemorrhagic pancreatitis and necrosis of the head of the pancreas, was recently treated by us and that case is the basis for this report.

Report of a Case

A 40-year-old man was admitted to Providence Hospital in Anchorage, Alaska, on June 7, 1978, after the sudden occurrence of severe epigastric pain and a temperature that rose to 39°C (102.2°F). The level of serum amylase was 2,700 units (normal less than 200). Past medical history included multiple episodes of abdominal pain.

From the Departments of Surgery, Swedish Hospital and University of Washington School of Medicine, Seattle.

Submitted November 5, 1979.

Reprint requests to: Thomas Taylor White, MD, Department of Surgery, RF-25, University of Washington, Seattle, WA 98195.

Previous upper gastrointestinal series had detected a mass in the second portion of the duodenum, which had been noted to appear and disappear since 1966. The patient's abdominal pain, fever and hyperamylasemia resolved over the next two weeks after nasogastric suction and administration of fluids intravenously. A transfusion of two units of whole blood was also required because the hematocrit had dropped to 30 percent. An attempt to feed the patient at this time, however, resulted in a return of abdominal pain and elevation of serum amylase. Central venous hyperalimentation was begun June 21. A cholangiogram after intravenous injection of contrast medium showed that the common bile duct was of normal caliber. An x-ray study of the upper gastrointestinal tract and a computerized tomographic (CT) scan of the abdomen showed a mass in the medial aspect of the second portion of the duodenum. On gastroduodenoscopy a mass was noted protruding from the medial aspect of the second portion of the duodenum. Biopsy studies of this lesion showed only duodenal mucosa.

The patient was transferred to Swedish Hospital Medical Center in Seattle on July 6. On admission he was noted to have epigastric pain, requiring administration of narcotic analgesic drugs. Serum amylase values were normal as was the remainder of the hemogram and chemistry profile. Endoscopy was carried out on the following day, and a soft, smooth, 2- by 3-cm mass was found on the medial aspect of the second portion of the duodenum. The ampulla of Vater was observed at the upper aspect of this structure and was cannulated. Injection with contrast medium showed a fluid-filled cyst without communication with the bile or pancreatic duct (Figure 1).

On July 11 the patient was operated on. Operative cholecystocholangiography showed a normal common bile duct communicating with a choledochocele and with the duodenum at the ampulla of Vater (Figure 2). Exploration of the lesser sac showed pronounced inflammatory response and necrosis of the entire head of the pancreas to the level of the portal vein. The tail of the pancreas appeared to be intact although it was somewhat indurated. There was some necrosis along the superior surface of the pancreas above the splenic artery. Pancreaticoduodenectomy was carried out to restore pancreatic continuity with the gastrointestinal tract and to remove the choledochocele. Continuity with the gastrointestinal tract was restored by end-to-end pancreaticojejunostomy, end-to-side choledochojejunostomy and end-to-side gastrojejunostomy. A Silastic tube was used to stint the pancreatic duct in the postoperative period. It drained copious amounts of clear pancre-

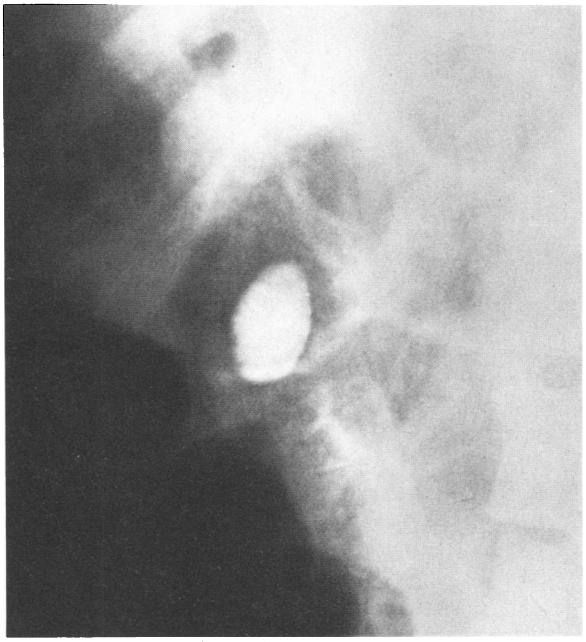

Figure 1.—Choledochal cyst after endoscopic injection with contrast medium. The air-filled shadow of the duodenum can be seen in the background.

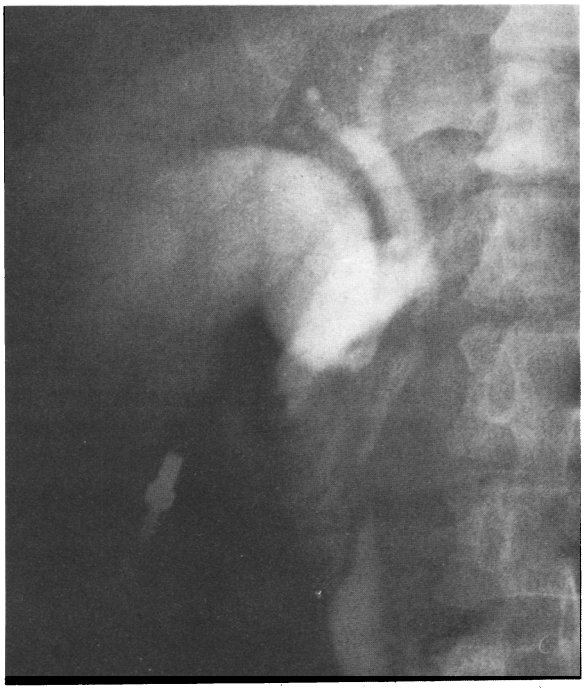

Figure 2.—Operative cholecystocholangiogram illustrating the connection of the common bile duct with the elongated choledochocele in the duodenum.

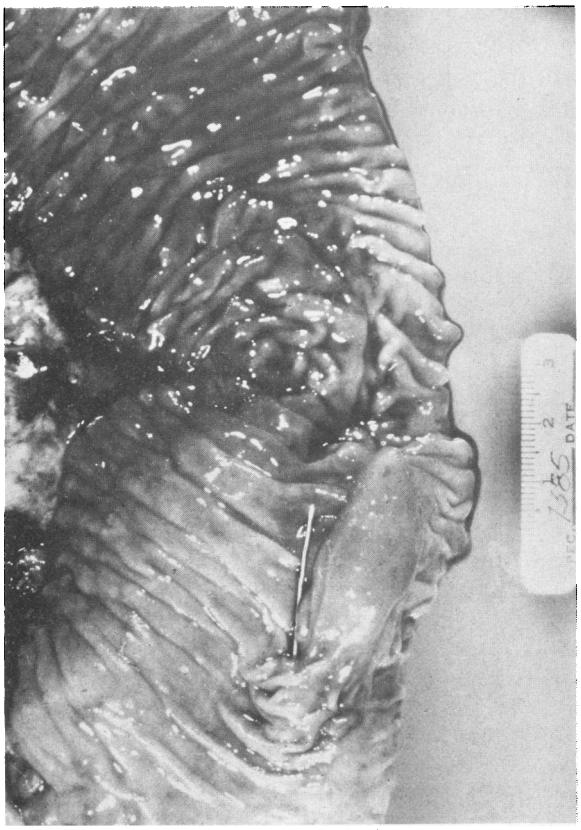

Figure 3.—Photograph of the operative specimen of the duodenum showing the point at which the bile and pancreatic juice entered the duodenum and the adjacent intraduodenal choledochocele.

atic juice, indicating that the remainder of the pancreas was functional. There was an unremarkable postoperative course and the patient was discharged on the 14th postoperative day without evidence of exocrine or endocrine-pancreatic insufficiency. Gross and histological examination of the operative specimen showed that the cyst was 2 by 4 cm in diameter (Figure 3), lined with duodenal mucosa and communicating with the bile duct at the ampulla. The pancreatic duct could not be located. Examination of the pancreas showed necrotic tissue, inflammatory exudate and fat necrosis.

Discussion

Choledochoceles have been described in previous reports as enterogenous cysts of the duodenum[10,15-17] or of the ampulla of Vater,[7,9] and duplication cysts of the duodenum.[14,22] It seems appropriate, however, on the basis of common anatomical and clinical characteristics to refer to these lesions as choledochoceles, as was originally suggested by Wheeler[23] and confirmed by Alonso-Lej and co-workers.[5] A review of the literature uncovered 21 previous case reports that corresponded to this description of a dilatation of the distal bile duct presenting as a mass within the duodenum. Pertinent features of these cases and our case are presented in Table 1.

Incomplete clinical data are available for these earlier cases. Thirteen of the patients were female, eight were male. Their average age was 34 years, with a range from 14 to 73. Of 20 patients in whom symptoms were noted, 18 had abdominal pain, most with an acute episode in addition to a history of intermittent abdominal distress. Pancreatitis was present in 6 of the 20 patients in whom symptoms were reported. A pseudocyst developed in one.[21] In our patient acute hemorrhagic pancreatitis with necrosis of the head of the pancreas developed. Three patients were jaundiced, four presented with anorexia and vomiting and two presented with gastrointestinal bleeding.

The lesion was always identified in the patients in whom upper gastrointestinal series were carried out. It was usually described as a round, filling defect in the second portion of the duodenum. Intravenous cholangiography correctly identified the lesion in four of the nine patients in whom the study was undertaken. In four others, results of the study were interpreted as normal, and in one a dilated duct was seen. The choledochocele was appropriately identified by duodenoscopy or endoscopic retrograde cannulation of the papilla in the four patients in whom it was done. Our patient is the only one in whom the lesion was visualized on a CT scan.

Two anatomical variations of this lesion were observed.[19] The bile duct and pancreatic duct were found to drain directly into the choledochocele in nine cases. The choledochocele then drained through its own orifice into the duodenum. In the nine other cases for which anatomical descriptions are available, the choledochocele presented as a cystic outpouching, draining into the distal bile ducts immediately proximal to the ampulla. Four of the choledochoceles contained gallstones.

Twelve of the choledochoceles were lined with duodenal mucosa, with a layer of muscularis separating the lining from the mucosa of the duodenum itself. In four other patients, the lining consisted of mucosa consistent with the distal bile

duct. In one patient a histological examination showed variable intestinal and bile duct components.

In most cases, surgical therapy consisted of excision of one wall of the choledochocele, essentially marsupializing the cyst into the duodenum, with careful preservation of the bile duct and pancreatic duct sphincters. Sphincteroplasty was used if impingement of the pathological process on these sphincters was suspected. This type of operation achieved excellent results in all of the reported cases. Pancreaticoduodenectomy was used in our patient to remove necrotic pancreatic tissue and restore pancreaticoenteric continuity.

TABLE 1.—*Review of 22 Reported Cases of Choledochocele*

Author/ Date of Report	*Age (yr)/ Sex*	*Symptoms*	*Diagnostic Test*	*Operative Findings*	*Histological Findings*
1. Wheeler[23]/1940	64/M	Abdominal pain, jaundice	..	2×3 cm cyst	None
2. Brooks[7]/1943	30/M	Abdominal pain	UGI	8×15 cm cyst with common bile duct and pancreatic duct entering cyst	Duodenal mucosa
3. Gordimer[10]/1950	30/F	Abdominal pain	..	4×8 cm cyst entering distal bile duct, gallstones in cyst	Duodenal mucosa
4. Polson[17]/1953	20/F	Abdominal pain, vomiting	UGI	3×5 cm cyst with bile duct entering cyst, gallstones in cyst	Duodenal mucosa
5. Alden[6]/1957	18/F	Abdominal pain	UGI	8×15 cm cyst with bile and pancreatic ducts entering cyst	Biliary mucosa
6. Serfas[20]/1957	43/F	Abdominal pain, jaundice	UGI, IVC	3×5 cm cyst entering distal bile duct, gallstones present	Intestinal and biliary mucosa
7. Stephens[21]/1966	18/M	Abdominal pain, pancreatitis	UGI	4×4 cm cyst with bile duct entering cyst	Biliary mucosa
8. Kaftori[13]/1966	18/M	Abdominal pain, vomiting	UGI, IVC	3×4 cm cyst with bile duct entering cyst	Duodenal mucosa
9. Leffall[14]/1967	52/F	Asymptomatic	UGI	8×11 cm cyst entering distal bile duct	Duodenal mucosa
10. Perrin[16]/1969	14/F	Abdominal pain, pancreatitis	UGI, IVC	4×4 cm cyst with bile duct entering cyst	Duodenal mucosa
11. De Oya[9]/1969	19/F	Abdominal pain, pancreatitis	UGI	3×4 cm cyst entering distal bile duct	Biliary mucosa
12. Jansen[12]/1970	15/F	Abdominal pain jaundice, pancreatitis	UGI	3×4 cm cyst entering distal bile duct, gallstones in cyst	Duodenal mucosa
13. Warren[22]/1971	22/M	Abdominal pain	UGI	5×8 cm cyst with bile duct entering cyst	Duodenal mucosa
14. Brunton[8]/1972	33/M	Abdominal pain, vomiting, gastrointestinal bleeding	UGI	Cyst with bile duct entering cyst	Duodenal mucosa
15. Ortiz[15]/1974	14/F	Abdominal pain, pancreatitis	UGI	12-cm cyst entering distal bile duct, gallstones in cyst	Duodenal mucosa
16. Reinus[18]/1976	21/F	Abdominal pain	UGI, duodenoscopy	Cyst entering into distal bile duct	Duodenal mucosa
17. Scholz[19]/1976	59/F	Abdominal pain	UGI	3×6 cm cyst entering distal bile duct	Duodenal mucosa
18. Scholz[19]/1976	73/F	Abdominal pain	UGI, IVC	Choledochojejunostomy, duodenum not opened	None
19. Hadad[11]/1976	50/M	Anemia, gastrointestinal bleeding	UGI, duodenoscopy	Cyst with bile duct entering cyst	Not reported
20. Kimura[3]/1977	..	..	..	..	..
21. Matsumoto[4]/1977	57/F	..	..	..	..
22. Present report/1980	40/M	Abdominal pain, pancreatitis	UGI, CT scan, ERCP	2×3 cm cyst entering distal bile duct	Duodenal mucosa

CT = computerized tomography; ERCP = endoscopic retrograde cholangiopancreatography; IVC = intravenous cholangiography; UGI = upper gastrointestinal (series)

The postoperative convalescence has been without complications (15 months).

The cause of choledochal cysts, particularly that of choledochoceles, remains unclear. The normal development of the gut and bile duct from a solid cord to vacuoles to a tubular structure coupled with epithelial proliferation is a setting in which inequalities of growth might lead to congenital cyst formation.[24] Babbitt suggests that reflux of pancreatic juice into the bile duct causes intermittent low-grade cholangitis with inflammation, dilatation and, finally, cyst formation.[25] Sterling's anatomical studies[26] support an acquired cause. He found evidence of diverticula in the distal common bile duct in 4 of the 70 postmortem examinations. He did not report on the histological anatomy of these cases. His cases were associated, however, with calculi in the common bile duct. For this reason he postulated that impaction of a stone in the papilla could cause increased intraluminal pressure, resulting in a choledochal diverticulum. This theory does not, however, account for the duodenal mucosa that has been found in most choledochoceles.

The fact that 30 percent of the patients reported in the literature had pancreatitis is of particular interest. The presumed cause is obstruction of the pancreatic duct by the choledochocele, which in turn results in acute pancreatitis. Gallstones were found within the choledochocele and may have played a causative role in the pancreatitis in two of the six patients with this disorder. The presence of a choledochocele has not been considered as a cause for pancreatitis in the past. In these cases, however, pancreatitis and choledochocele frequently occurred together. Therefore, a choledochocele should be considered in the diagnosis of young patients with acute pancreatitis but without a history of gallstones, alcoholism or abdominal trauma.

Summary

Our patient represents the 22nd reported case of choledochoceles. The cause of this rare lesion remains obscure. Choledochoceles should be kept in mind as a possible cause of pancreatitis in young patients without a history of gallstones, alcohol abuse or abdominal trauma. Technically, surgical correction is easy and is highly successful in early uncomplicated cases.

REFERENCES

1. Douglas AH: Case of dilatation of common bile duct. Monthly J Med Sci (London) 14:97, 1852

2. Flanigan DP: Biliary cysts. Ann Surg 182:635-643, Nov 1972

3. Kimura K, Ohto M, Ono T, et al: Congenital cystic dilatation of common bile duct—Relationship to anomalous pancreaticobiliary ductal union. Am J Roentgenol 128:571-577, Apr 1977

4. Matsumoto Y, Uchida K, Nakase A, et al: Congenital cystic dilatation of the common bile duct as a cause of primary bile duct stone. Am J Surg 134:346-352, Sep 1977

5. Alonso-Lej F, Rever W, Pessagno D: Collective review—Congenital choledochal cyst, with report of two and analysis of 94 cases. Int Abs Surg 108:1, 1959

6. Alden JF, Sterner ER: Diverticulum of the common bile duct—A case report. Ann Surg 145:269-270, Feb 1957

7. Brooks B, Weinstein A: Cyst of the ampulla of Vater. Ann Surg 117:278-734, May 1943

8. Brunton FJ, Bamforth J: Intraluminal diverticulum and choledochocele. Gut 13:297-210, Mar 1972

9. De Oya JC, Puente JL, Vallanueva A, et al: Enterogenous cyst of the ampulla of Vater. Digestion 2:201-208, 1969

10. Gordimer H, Bluestone L: Enterogenous cyst of the duodenum. Ann Surg 132:1149-1153, Dec 1950

11. Hadad AR, Westbrook KC, Campbell GS, et al: Congenital dilatation of the bile ducts. Am J Surg 132:799-804, Dec 1976

12. Jansen W: Intraduodenal cyst containing bile and stones—"Choledochocele" in an accessory bile duct. Gastroenterology 58: 397-401, Mar 1970

13. Kaftori J, Munk J, Schramek A, et al: Intraluminal diverticulum of the duodenum demonstrated by intravenous cholangiography. Br J Radiol 39:388-390, May 1966

14. Leffall LS Jr, Jackson M, Press H, et al: Duplication cyst of the duodenum. Arch Surg 94:30-34, Jan 1967

15. Ortiz VM, Nealon TF Jr, Mitty WF Jr: Enterogenous cyst of the duodenum. Am J Gastroenterol 61:276-281, Apr 1974

16. Perrin RW: Communicating enterogenous cyst of the duodenum receiving the termination of the common bile duct demonstrated by intravenous cholangiography. Radiology 93:675-676, Sep 1969

17. Polson RA, Isaac JE: Enterogenous cyst of the duodenum. Gastroenterology 25:431-434, Nov 1953

18. Reinus FZ, Weingarten G: Choledochocele of the common bile duct. Am J Surg 132:646-648, Nov 1976

19. Scholz FJ, Carrera GF, Larsen CR: The choledochocele—Correlation of radiological, clinical and pathological findings. Radiology 118:25-28, Jan 1976

20. Serfas L, Lyter C: Choledochal cyst with a report of an intraduodenal choledochal cyst. Am J Surg 93:979-989, 1957

21. Stephens FO, Pauline GJ: Choledochocele—An unusual type of choledochal cyst which presented an acute pancreatitis. Aust N Z J Surg 36:124-127, Nov 1966

22. Warren KW, Mountain JC, Erickson EE: Duodenal duplication cyst—A case report in which the pancreatico-biliary system drained directly into the cyst. L C F Bull 20:114-120, Jul-Sep 1971

23. Wheeler WL: An unusual case of obstruction of the common bile duct (choledochocele?). Br J Surg 27:446, 1940

24. Yotsuyanagi S: Contribution to etiology and pathology of idiopathic cystic dilatation of the common bile duct with report of three cases. Gann 30:601, 1936

25. Babbitt DP, Starshak RJ, Clemett AR: Choledochal cyst—A report of etiology. Am J Roentgen Rad Ther Nuc Med 119:57-62, Sep 1973

26. Sterling J: Diverticula in the terminal portion of the common bile duct. Am J Pathol 25:325, 1949