INTERMITTENT JAUNDICE DUE TO NEUROMA OF CYSTIC AND COMMON BILE DUCTS*

By Mandred W. Comfort, M.D.
of the division of medicine
and
Waltman Walters, M.D.
of the division of surgery
of the Mayo Clinic, Rochester, Minnesota

Intermittent obstructive jaundice due to the formation of tumor of the bile ducts is comparatively rare. Marshall, in 1930, found that only four cases of benign tumor and forty-nine cases of primary carcinoma of the extrahepatic bile ducts had been seen in The Mayo Clinic in the last twenty years. Rolleston and McNee, in 1929, recorded 112 cases of carcinoma of the ducts exclusive of the growths in the ampulla, and of only ten cases of benign tumors. Of these tumors the benign, although much more rare than the malignant, lend themselves to surgical cure and afford the patient assurance that recurrence or metastasis will not occur. A twofold interest is found in these cases of intermittent obstructive jaundice due to a benign tumor of the extrahepatic bile ducts, of which the following is an example.

Report of Case.—A woman, aged fifty-five years, registered at the clinic September 30, 1930, complaining of pruritus and jaundice of two and a half weeks' duration. At the age of thirty years a series of typical attacks of colic of the gall-bladder began; they were not associated with jaundice. Ten years before admission cholecystectomy and appendectomy had been performed elsewhere. A biliary fistula persisted. Two temporary closures of the fistula were followed by jaundice; the final closure occurred eighteen weeks after operation and the patient remained well for five years. Five years afterward deep jaundice developed, which was essentially painless and was associated with anorexia and pruritus. The jaundice disappeared after five weeks, to reappear one and a half years later and to last five or six weeks. This second attack of jaundice as well as the third, which occurred a year before admission, was accompanied by severe prostration, loss of strength and weight, chilly sensations, and slight fever. The last attack began at night in the right upper quadrant two and a half weeks before admission with discomfort which awakened the patient from sleep. Vomiting relieved the distress. Jaundice and pruritus appeared two days later. The appetite remained fair and the digestion good. The stools became clay-colored and the urine dark.

The patient was 5 feet, 2 inches tall and weighed 126 pounds. She had lost 20 pounds in the last five years, but appeared to be well nourished. She was markedly jaundiced and her skin had been diffusedly excoriated by scratching. Nodules were not found along the course of the nerves suggestive of von Recklinghausen's disease. The liver was slightly enlarged and firm. The specific gravity of the urine was 1.021; it was acid in reaction, did not contain sugar, but a moderate amount of bile, and an occasional erythrocyte and leucocyte in the high-power field. The test of urobilin was positive. The percentage of hæmoglobin was 80; erythrocytes numbered 3,950,000 and the leucocytes 7,100. The coagulation time by the Lee and White method was 6

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minutes and 30 seconds. The serum bilirubin was 11.5 milligrams and the van den Bergh reaction was direct. The stools were negative for bile with the Schmidt test. Clot retraction was slight at the end of one hour and complete at the end of two hours. The fragility test gave normal resistance. The blood urea was 22 milligrams for each 100 cubic centimetres. Repeated duodenal drainage showed a trace of bile in the contents.

During the patient's stay in the hospital before operation normal temperature and pulse persisted. The concentration of serum bilirubin fell gradually and October 8, the reading was 7.1 milligrams. The coagulation time increased to ten minutes, but following transfusion October 9, and intravenous administration of 10 per cent. calcium chloride in amounts of 5 cubic centimetres, it dropped to seven minutes. A diagnosis was made of intermittent and partial obstruction of the common bile duct. The history of cholecystectomy for cholecystitis with stones and the intermittent appearance of jaundice over a five-year period indicated stones as the cause of the obstruction. On the contrary, the initial appearance of jaundice after operation and its painlessness indicated stricture. The unusual feature was the long interval between attacks.

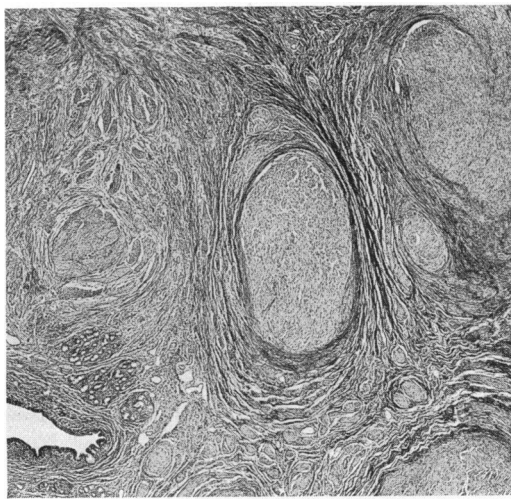

FIG. 1.—Sheath neuroma of the common and cystic ducts. Interlacing cellular strands covered by fibrous sheaths in a groundwork of connective tissue surrounding the cystic duct (van Gieson's stain).

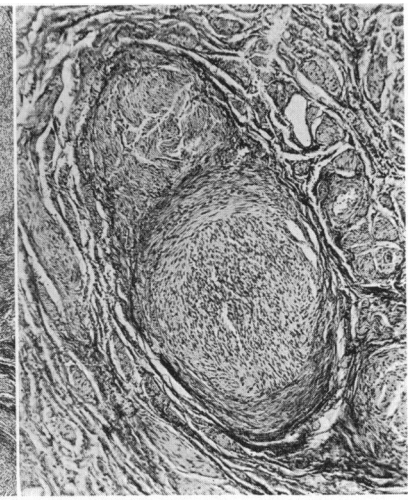

FIG. 2.—Sheath neuroma of the common and cystic ducts. Cross section of a cellular strand ensheathed by well-developed fibrous capsules (van Gieson's stain).

October 10, 1930, operation was performed. There was a solid lesion in the middle portion of the common bile duct approximately 1 centimetre in diameter, and obstructing the duct. This portion of the duct was excised. A common-duct probe and scoop could be passed through the lower end of the duct into the duodenum. The two ends of the stumps of the hepatic duct were sutured to the lower end of the common bile duct over two small T-tubes, interrupted sutures of silk being used in the anastomosis. Satisfactory anastomosis between the ducts was accomplished.

The patient's post-operative convalescence was uneventful, and she was allowed to return home, November 3. The incision healed except for the sinus the T-tubes occupied. She was in excellent condition. The T-tubes have remained closed, allowing bile to pass into the external limb of the duodenum. She continues to be in excellent condition (January 8, 1931) and will return to the clinic within a few weeks for the removal of the T-tubes.

The specimen removed at operation measured approximately 1.5 by 1 by 0.75 centimetres. It was firm, and on cross section a central canal whose walls were continuous

with the attached portion of the common bile duct could be traced three-fourths of the way to the free end of the mass. Sections were stained with hæmatoxylin and eosin, van Gieson's stain, and by silver impregnation methods for axis cylinders. The central canal, observed grossly and microscopically, had the appearance of the cystic duct. Chronic inflammatory changes (lymphocytes and fibroblastic reaction) were present in the mucosa. Mucous glands were seen. Two types of tissue composed the remainder of the tumor: Connective tissue, and large and small bundles of tissue which ran in all directions. The many interlacing strands gave an unusual plexiform arrangement. The connective tissue had an adult appearance and did not contain evidence of inflammatory reaction seen in the mucosa. The interlacing bundles of tissue were composed of numerous elongated cells lying parallel to the long axes of the bundles, often assuming the wavy arrangement characteristic of nerve tissue; they stained yellow with the picric acid of van Gieson's stain. The nuclei stained brown. Palisade arrangement of the nuclei, interlacing fibrils, or foam cells were not present. Usually the bundles had heavy sheaths of a mixture of young and old connective-tissue cells. Red- or pink-staining connective fibrils were not present inside the bundles. Silver impregnation showed swollen fragments, which were difficult to recognize as axis cylinders, although they gave fairly characteristic staining reactions. The tumor had an unusual structure. Its classification was difficult because of lack of data on a similar tumor. It did not correspond to the acoustic or von Recklinghausen's neurofibromas; moreover, it was solitary. Rosettes such as were described by Stout in his case of tumor of the ulnar nerve were not observed nor was the peculiar cellular arrangement of the sheath neuroma of the Gasserian ganglion described by Learmonth and Kernohan present. There seemed to be an excessive overgrowth of the sheath cells which are normally present in the sympathetic nervous system, accentuating the numerous small nerve bundles which normally lie in the connective tissue around the cystic duct. The term sheath neuroma has been suggested by Kernohan for the tumor in spite of the fact that it is dissimilar to one previously observed by him. He explained the difference on the basis that in this case the tumor is of the sympathetic nerve trunk and not of the sensory ganglion.

The neuroma in this case is not only rare but its situation is unusual. Cysts of the common bile duct,[5] adenofibromas of the cystic ducts,[8] and papillomas of the cystic and common bile ducts have been reported.[5, 7, 9] Fibromas,[1, 4] adenomyofibromas,[11] lipomas,[3, 12] and hydatid cysts[2] are other types of benign tumors. The types seem to be almost as numerous as the tumors themselves. The explanation of the obstruction by external pressure of the tumor is of interest. W. J. Mayo found the same explanation feasible for his two cases of adenofibroma of the stump of the cystic duct which similarly produced intermittent obstructive jaundice with symptoms of cholangitis. The symptoms of obstruction of the common bile duct due to benign tumors are indistinguishable from those due to stone, especially if the stone is silent, to partial stricture secondary to surgical accidents, or to the formation of scar from pressure of the stones. The progressive course and at times the blood in the duodenal contents will serve to distinguish benign from malignant stricture. Pain and signs and symptoms of coëxisting infection may or may not be present. The benign nature of the lesion may be suspected in similar cases. Its rarity will not encourage one to venture a definite diagnosis.

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