Chronic Inflammatory Sclerosis of the Pancreas—An Autonomous Pancreatic Disease?

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On the basis of the work of Comfort et al.¹ and Gambill et al.,² many investigators consider that there is only one sequence of pathologic changes in chronic relapsing pancreatitis—i.e., that it is secondary to recurrent attacks of acute pancreatitis. These attacks may be lethal, as a result of massive necrosis or may be followed by complete recovery without any aftereffects. According to this view, calcifying pancreatitis is the final stage of chronic pancreatitis.

Our own anatomic and statistical investigation has led us to a contrary conclusion. This work, described in part elsewhere,³ is based on the clinical records of 98 patients having unquestionable pancreatitis—recognized by the presence of at least one of the following criteria: radiologic calcifications, diffuse sclerosis on microscopic examination, or necrosis or edematous lesions at operation. The conclusion was reached that there is not merely one pancreatic disease but many diseases having different causes and development.

Among the chronic diseases of the pancreas, one form seemed worth isolating. It is clearly distinguished by lesions of very mutilating and inflammatory fibrosis and by generally rapid progression to cachexia and hypoproteinemia.

The designation “evolutive sclerosis of the pancreas without calcification or Odditis”³,⁴ has been proposed, but this condition can also be called “primary inflammatory sclerosis of the pancreas.” The first investigation was made on 12 patients. In this paper, only the 10 cases in which anatomic examinations were performed are considered. In all cases, systematic sclerosis of the pancreas and the absence of intracanalie—
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ular calcifications were ascertained by biopsy. The possibility that odditis or primary obstruction caused the pancreatitis could be eliminated in every case, either by preoperative or by postoperative cholangiography. Only one cause of error admits of discussion—pancreatitis due to stasis behind a slowly developing small cancer of the duodenum or of the head of the pancreas. The clinical record of 1 of our female patients, not included in this series, convinced us of the difficulty of this diagnosis. At the age of 37, Mrs. B - - -, began having painful attacks in the epigastrium with loss of weight, followed by gradually extending jaundice. Biopsies taken at two operations revealed chronic pancreatitis. Only necropsy revealed a small duodenal cancer, with pancreatitis behind it.

In 3 of our 10 patients, histologic examination of the entire pancreas and of the duodenal mucous membrane unquestionably eliminated the possibility that very small cancers were present.

Two patients lived long enough after the appearance of their pancreatitis to allow one to state almost positively that they did not have cancer. The pancreatic attacks of Mrs. Pour - - -, started when she was 46 years old. Ten years later she underwent resection of the tail of the pancreas. Histologic examination of the operative specimen and detailed surgical exploration did not disclose cancer. The attacks of Mrs. Poi - - -, started when she was 60 years old. Five years later an operation with wide biopsy of the pancreas revealed a sclerous pancreatitis without detectable cancer (Fig. 1).

In 3 patients, the periods during which the disease developed were long enough to indicate the probable absence of cancer (3 years for 1 patient, and 2 years for the others).

Finally, in 2 patients it is possible that cancer is absent, since the periods from the start of the disease to the identification of the lesions at operation have been 6 and 7 months, respectively.

ETIOLOGY

Age

The patients in whom the diagnosis was certain were 58, 60, and 73 years old at the time of the first appearance of the disease; those in whom it was very probable were 46 and 60; those in whom it was probable were 60 and 70; and those in whom it was possible were 58 and 67. The average age is consequently high: 65.6 ± 2.9 years for men, and 57.2 ± 2.9 for women. If the average age of the men is compared with that of our 38 male patients suffering from calcifying pancreatitis (40.0 ± 1.6), a statistically significant difference (P < 0.001) is noted. There is a similar difference in the ages of the women. The fact that “primary inflam-