

Arteriographic manifestations of the Zollinger-Ellison syndrome

REPORT OF A CASE

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THE roentgenographic manifestations of the Zollinger-Ellison syndrome¹ have been described by various authors.²⁻⁴ In addition, three cases were reported in which arteriography demonstrated the presence of non-beta islet cell adenomas and carcinomas of the pancreas.⁵⁻⁷ Our report concerns a case in which the sites of multiple pancreatic adenomas in the body and tail of the pancreas were demonstrated on arteriograms. A remarkable degree of gastric and intestinal venous opacification was seen during the arterial phase of the study. It is possible that this previously undescribed phenomenon is a physiologic change produced by large quantities of circulating gastrin generated by the adenomas.

Report of a case

A 29-year-old Caucasian woman was admitted to the Cleveland Clinic Hospital on December 21, 1967, because of persistent vomiting and for suspected Zollinger-Ellison syndrome. She had a seven-year history of preprandial epigastric pain. She had been amenorrhic since the age of 16 years, and, although married, had never been pregnant. Six years previously she had undergone operation for a perforated duodenal ulcer. One year later, renal and ureteral calculi were discovered. In 1966, serum calcium values on several occasions were reported to be abnormally high. In March 1967, a parathyroid adenoma was removed; however, serum calcium values remained persistently elevated. She had no history of diarrhea.

Physical examination revealed the pulse rate to be 110 and the blood pressure 140/110 mm Hg. There was a well-healed surgical scar on the anterior surface of the neck. Laboratory examinations demonstrated a blood hemoglobin content of 15.2 g per 100 ml, and serum calcium values of 13.2 mg, 12.6 mg, and 11.8 mg per 100 ml. The serum phosphorus values were 1.9 mg, 1.8 mg, and 1.1 mg per 100 ml. A 12-hr specimen of gastric juice had a volume of 1600 ml, free hydrochloric acid of 110 units, with a total of 128 units; the pH was 1.35.

A selective celiac arteriogram was performed on December 21, 1967, and demonstrated a 2.5-cm diameter area of faintly increased density in the pancreas. The area was located at the junction of the body and tail of the pancreas (*Fig. 1*). In addition to the site of increased density, it was seen that contrast medium returned through the coronary, splenic, and gastro-

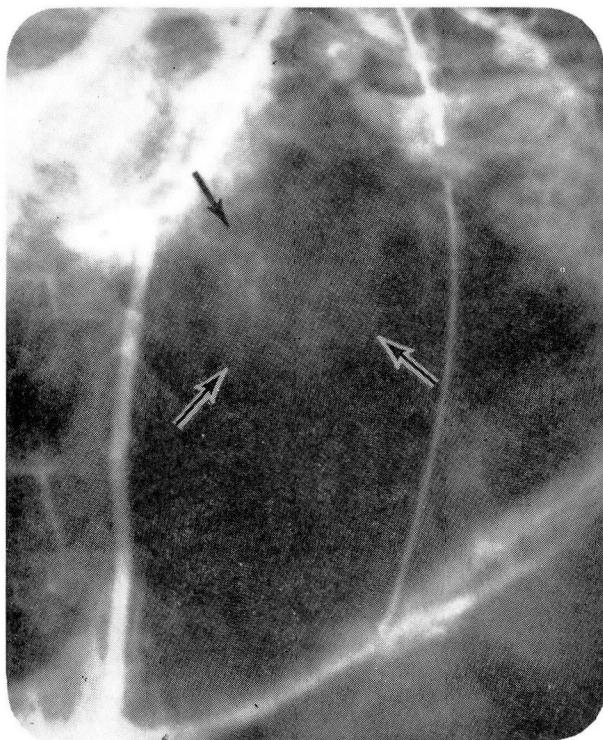


Fig. 1. A faint "blush" 2.5 cm in diameter seen at the junction of the body and tail of the pancreas was the site of multiple pancreatic adenomas removed at surgery. The blush appeared early in the arterial phase and persisted into the venous phase. The stomach was insufflated with gas.

epiploic veins during the arterial phase (*Fig. 2*). In the late arterial phase the portal system was completely opacified. The arteriogram demonstrated dense opacification of the gastric mucosa in the body but not in the antrum of the stomach (*Fig. 3*). A subsequent superior mesenteric arteriogram also demonstrated simultaneous opacifications of the arterial and the venous systems during the latter part of a 4-sec injection of 36 ml of contrast medium (*Fig. 4*). There was no evidence of a macroscopic arteriovenous fistula, and arteriovenous shunting at the capillary level was presumed to be present.

On December 26, 1967, the patient underwent subtotal gastrectomy, subtotal pancreatectomy, splenectomy, and biopsy of the liver. Histologic examination of the pancreas revealed the presence of multiple macroscopic and microscopic adenomas of the islets of Langerhans.

In the postoperative period, serum calcium values gradually decreased from 11.5 mg to 9.0 mg per 100 ml; the serum phosphorus values were 1.7 mg and 1.0 mg per 100 ml. The serum albumin value decreased postoperatively to 1.6 g from a preoperative value of 3 g per 100 ml; this may partially explain the decrease in the serum calcium values. The gonadotropin assay was more than 318 mouse units per 24 hr, which was consistent with a diagnosis of primary ovarian failure. Roentgenograms of the sella turcica showed no evidence of pituitary tumor.

Comment

Conclusions concerning the use of arteriography in the diagnosis of the Zollinger-Ellison syndrome would appear unjustified in the light of the



Fig. 2. During the arterial phase of a 4-sec injection of 40 ml of contrast medium, the portal and gastric veins became opacified. The area of tumor stain is seen to the left and slightly below the origin of the celiac axis.

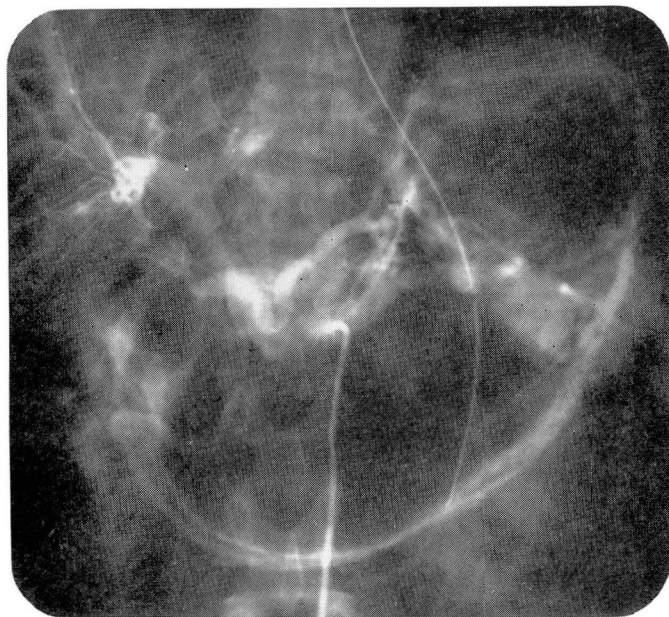


Fig. 3. In the late arterial phase, contrast medium remained in the hepatic arteries and there is dense opacification of the coronary, right gastroepiploic, and portal veins. The splenic vein is opacified, but concentration of contrast medium is greatest in the coronary vein. The stomach is densely opacified on the greater curvature in the region of the body of the stomach; the pyloric antrum and vestibule are normal.

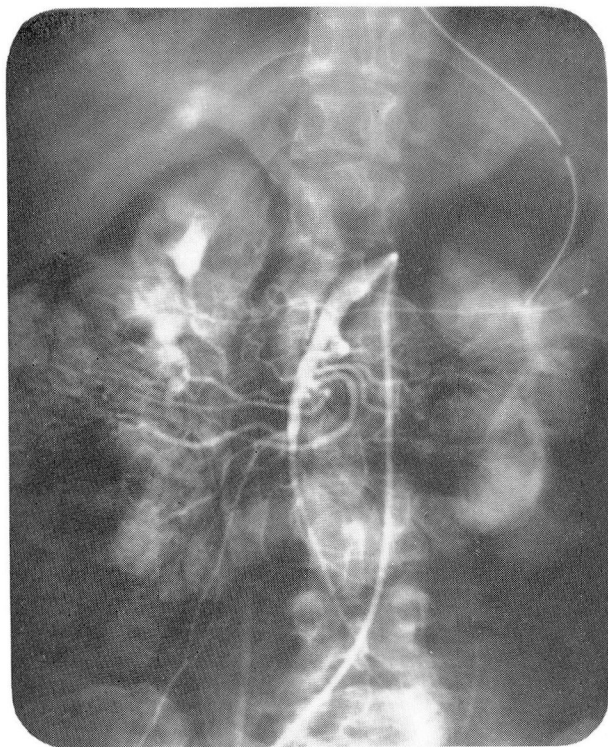


Fig. 4. A superior mesenteric arteriogram showing simultaneous opacification of the superior mesenteric artery and vein without evidence of macroscopic arteriovenous communications.

meager experience accumulated thus far. Nevertheless, arteriography has been successful in localizing the sites of the neoplasms in several reported cases. As previously mentioned, the case we report is believed unique in that it demonstrated an extremely early return of contrast medium through the venous circulation, a phenomenon that may be related to the presence of large amounts of gastrin produced by the adenomas. Swan and Jacobson⁸ showed experimentally that mucosal blood flow in the stomach of conscious dogs was increased as the systemic concentration of gastrin was increased; the total arterial gastric blood flow was not affected. If this observation is extrapolated to the arteriographic findings in the case we report, the presence of a mucosal shunt at the capillary level can be postulated. An increase in mucosal blood flow could also account for the dense opacification of the gastric mucosa as seen on the arteriograms.

In two of the cases previously reported^{6, 7} a rather densely opacified area was seen in the region of the neoplasm; whereas, in the case we report, and in one reported by Clemett and Park⁵ the neoplasms were only faintly opacified. It is doubtful that the latter two neoplasms would have been demonstrated had the stomach not been insufflated.

Summary and conclusion

Arteriography localized the site of the pancreatic tumor in a patient with Zollinger-Ellison syndrome treated by the authors, and in three patients whose cases were reported by other physicians.⁵⁻⁷ The use of arteriography is justified and should be encouraged. The simultaneous opacification of the arterial and the venous systems is an unusual finding, and suggests the presence of arteriovenous shunts at the capillary level, possibly through the mucosa. The findings may be related to the increased amounts of circulating gastrin produced by the pancreatic adenoma.

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