

Non-functioning Neuroendocrine Carcinoma Of Ectopic Pancreas in Colon

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BACKGROUND

The ectopic pancreas is pancreatic tissue found outside the confines of the main gland and may be found anywhere in abdominal organs. It may cause inflammation, bleeding and obstruction in the gastrointestinal tract but the risk of its malignant transformation is extremely rare.

REPORT OF CASE

Clinical Findings

A 75 year-old women with senile hypertension was admitted for hematochezia developed 3days ago. She complained mild dizziness but did not abdominal pain, gastrointestinal symptoms. On physical examination, about 6cm sized firm mass with tenderness was palpated on left upper quadrant of abdomen. Complete blood count showed a lower hemoglobin level (7.3g/dl). Ultrasonography revealed colonic mass about 10cm sized in diameters with small lymph node in LUQ (Fig.1). Computed tomography of abdomen-pelvis revealed about 9cm in diameter s mass lesion in the splenic flexure of colon with serosal penetration suggested local invasion and about 5cm sized in diameters Gastrointestinal stromal tumor in the fundus of stomach (Fig. 2). The colonoscopy showed huge mass encircling colonic lumen with bleeding (Fig.3). Pre-operative CEA level was 342.7ng/ml.

Operative Findings

An elective laparotomy was performed. In the peritoneal cavity, there were small amount of ascites but Intraperitoneal seeding and adhesion was not found. There were no distance metastasis in liver. The huge mass over 10cm in diameters was located on splenic flexure of Lt. colon. The mass was penetrated serosal layer of colon and adhered to distal pancreas. The pericolic lymph node was seen in the mesocolon. The Lt. hemicolectomy with distal pancreatectomy was performed(Fig. 4). On opening of the colon, there was a completely luminal encircling, pale yellow to greenish ulceroinfiltrative tumor with necrosed measuring 10X10X7cm. The pathologic report revealed malignant epithelial tumor(carcinoma), poorly-differentiated, involvement of colon and pancreas with 1) infiltration of entire colonic wall from adipose tissue and up to mucosal surface 2) foci of acinar formation 3) tumor metastasis in 11 out of 30 regional lymph nodes suggestive of Poorly-differentiated neuroendocrine carcinoma of pancreas origin (Fig. 5,6,7). The immunohistochemical staining (CK7) suggested the primary pancreatic tumor (F1g. 8)

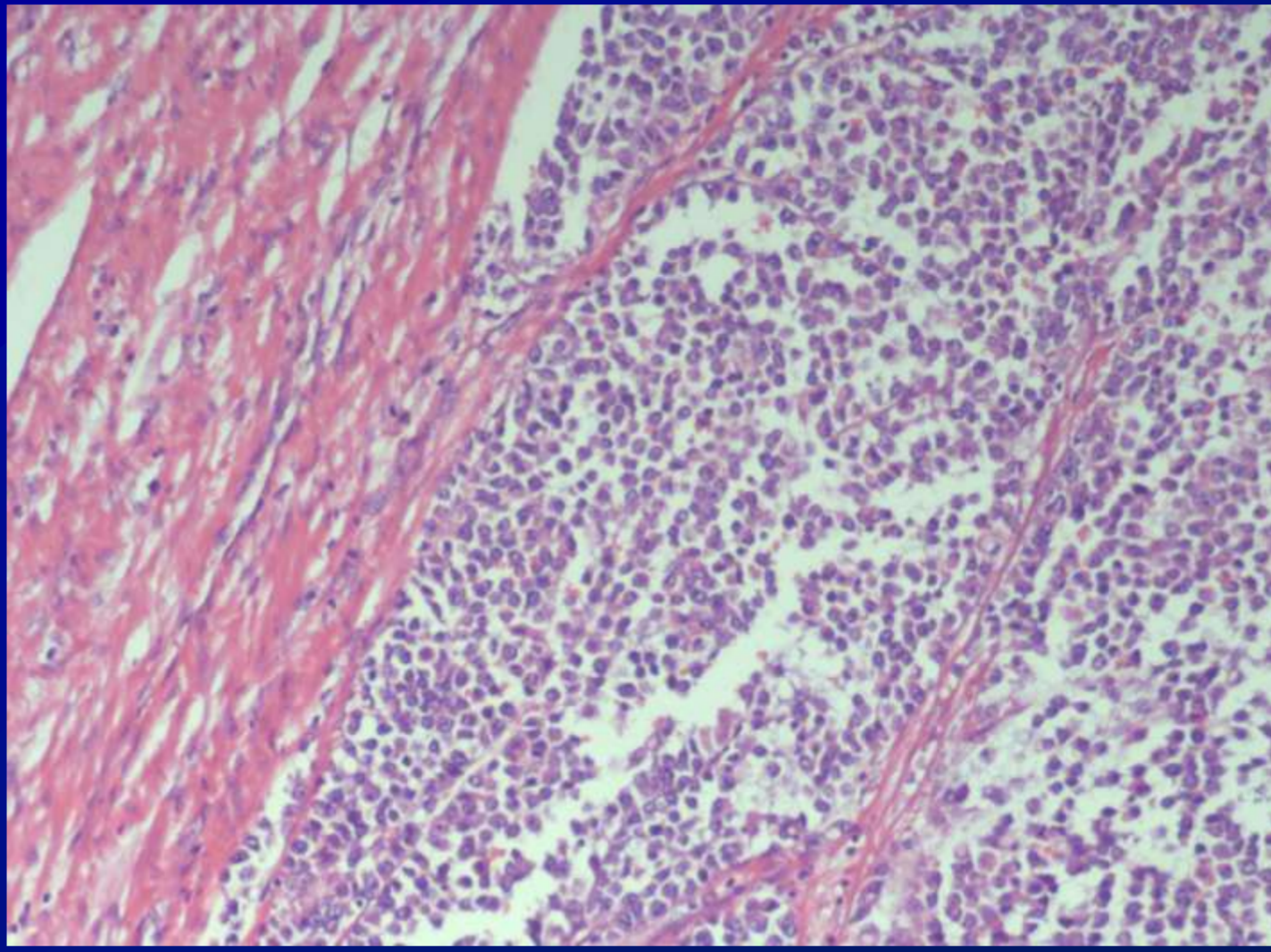


Fig.5. undifferentiated carcinoma infiltrating colonic mucosal layer (X10)

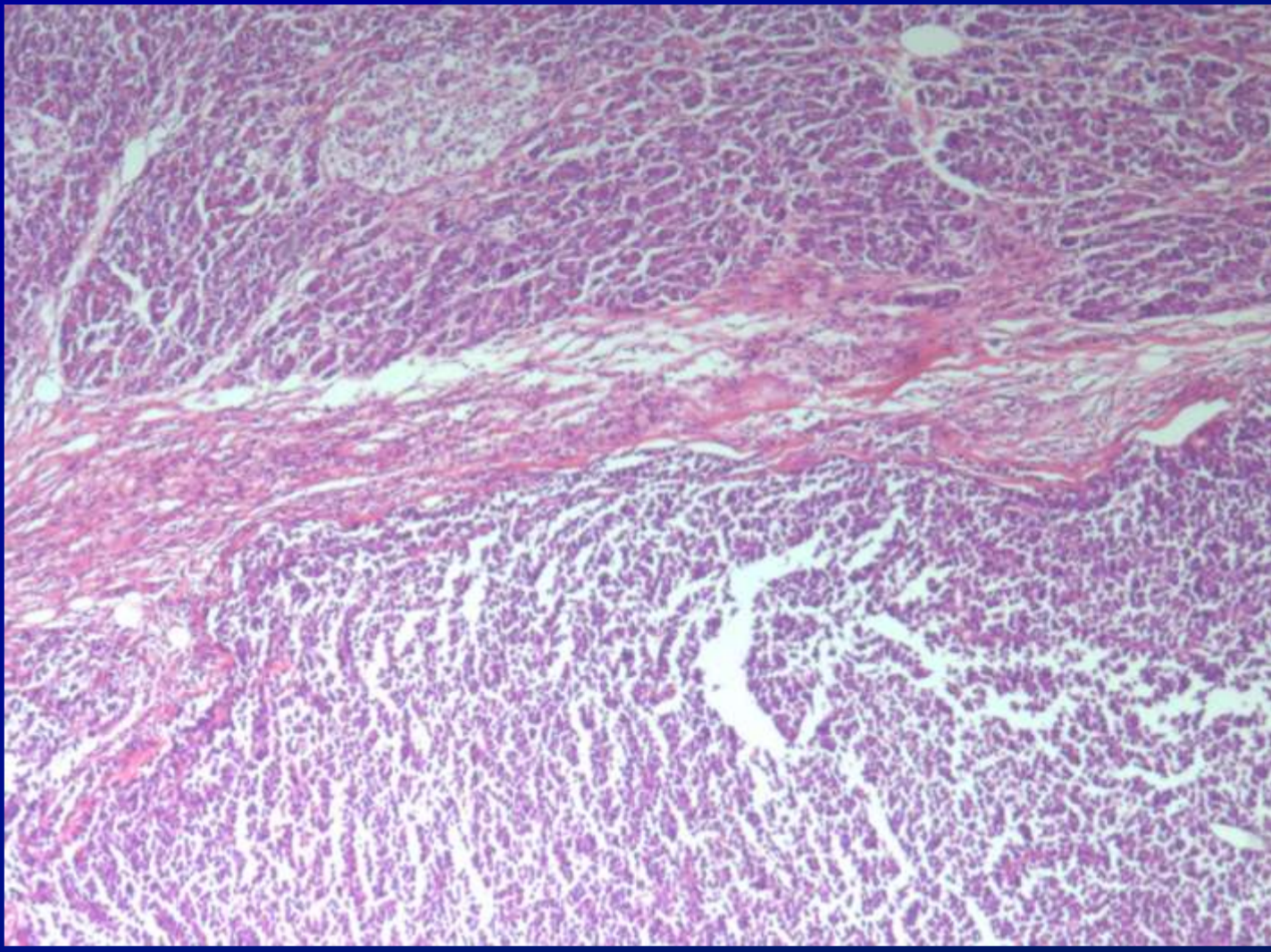


Fig.6. undifferentiated carcinoma with pancreatic involvement. (X10)

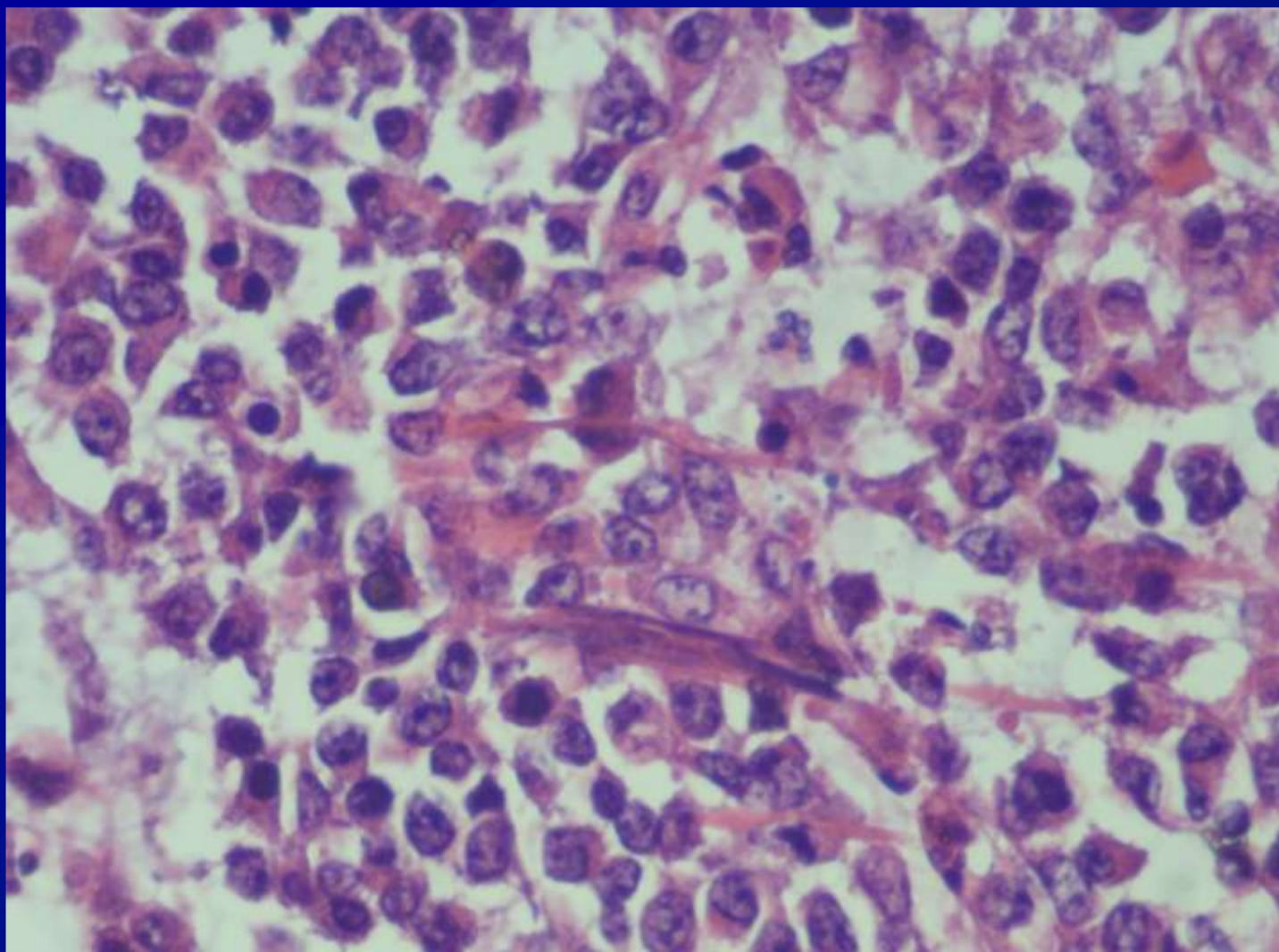


Fig.7. poorly-undifferentiated neuroendocrine carcinoma of pancreas origin

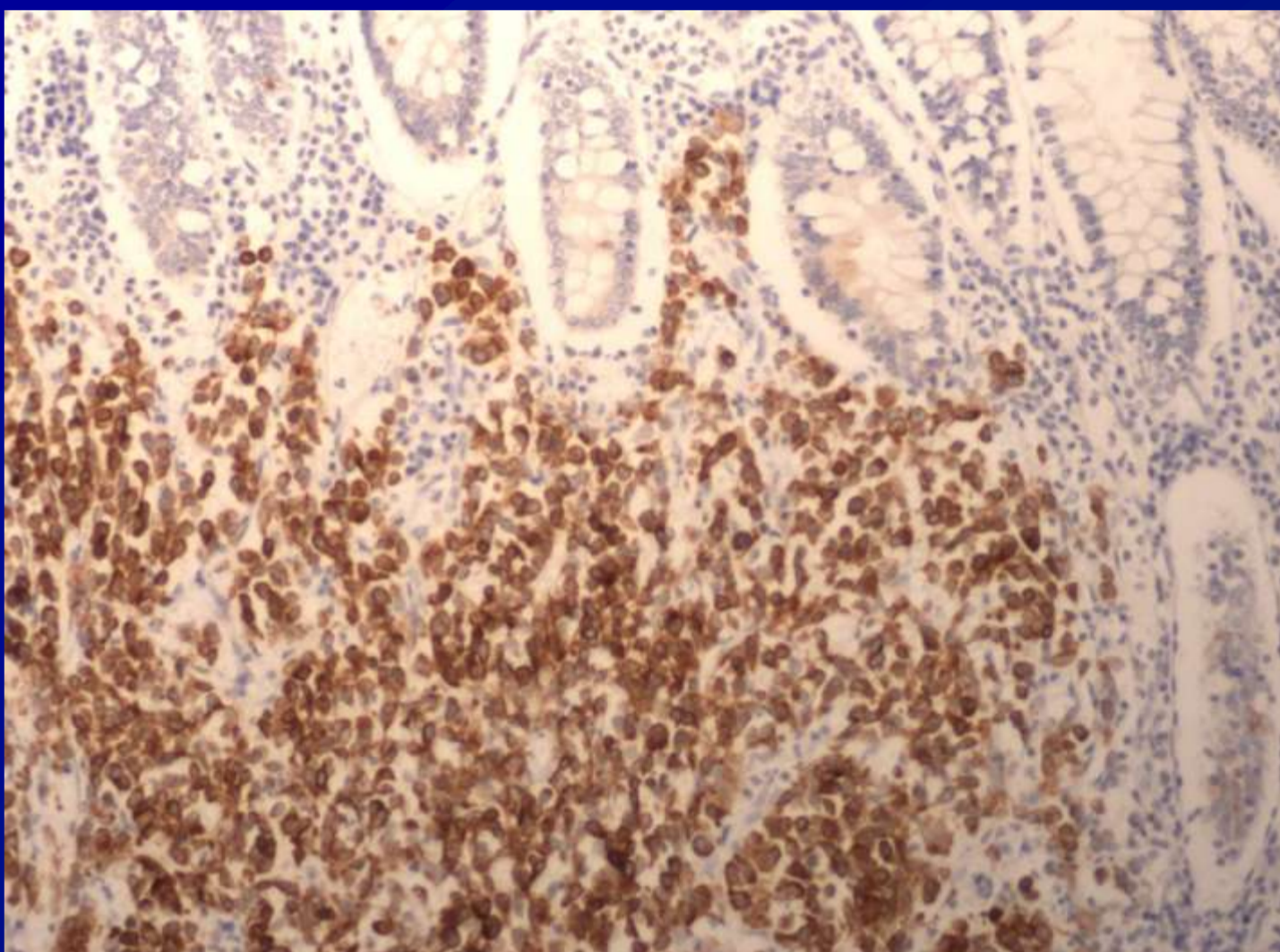


Fig.8. immunohistochemical staining (CK 7) suggested the primary pancreatic tumor.

Clinical Course

At 3th post-operated day, the Levin tube removed. The diet was taken at 6th post-operated day. The CEA levels at 7th post-operated day was decreased 73.4 ng/ml. She discharged at 111th post-operated day without any complication.

DISCUSSION

An ectopic pancreas, also called a heterotropic or aberrant pancreas, was first described by Jean Schultz in 1729 and refers to pancreatic tissue that lacks anatomical and vascular connections with the pancreas¹. It occurs most commonly in the stomach, duodenum, and jejunum and has been reported in other locations, including the ileum, Meckel's diverticulum, colon, gall bladder, umbilicus, fallopian tube, mediastinum, spleen and liver.

An ectopic pancreas is usually discovered incidentally during radiographic or endoscopic examination of the gut, surgery for other abdominal conditions, or upon autopsy. A patient with an ectopic pancreas can be normal or present with symptoms such as bleeding, vomiting, or abdominal pain due to pancreatitis, intestinal obstruction, or intussusception³. Rarely, an ectopic pancreas can be associated with other pancreatic disease, including islet cell tumors, pancreatic carcinomas, and pancreatic cyst. The malignant transformation of ectopic pancreas has been reported in different sites of the gastrointestinal tract⁶. There are few reports of case in gastric and small bowel wall but in colonic wall nearly not.

The diagnosis my be difficult, and is incidentally found by conventional imaging studies or intraoperatively. Fortunately the symptom of hematochezia developed in this case, early diagnosis was performed.

For a carcinoma to be described as arising from ectopic pancreatic tissue, three criteria have been proposed. Firstly, the tumor must be found within or close to the aberrant pancreatic tissue. Secondly, the transitional area between pancreatic structures and carcinoma must be observed (i.e. duct cell dysplasia and/or carcinoma in situ). Thirdly, the non-neoplastic heterotropic pancreatic tissue must comprise of at least fully developed acini and/or ductal structures. The lesion in the present case had findings compatible with all three criteria.

Prognosis of carcinoma from ectopic pancreas is not unknown. In the literature, only ten cases have been reported with a survival time between six month and ten years and all with a life expectancy longer than that in the orthotropic pancreas surgery.

In summary, we report a rare case of malignant transformation of ectopic pancreas of the colonic wall in a patients with hematochezia.



Fig. 1. showed a colonic mass with small lymph node in Lt. upper quadrant of abdomen

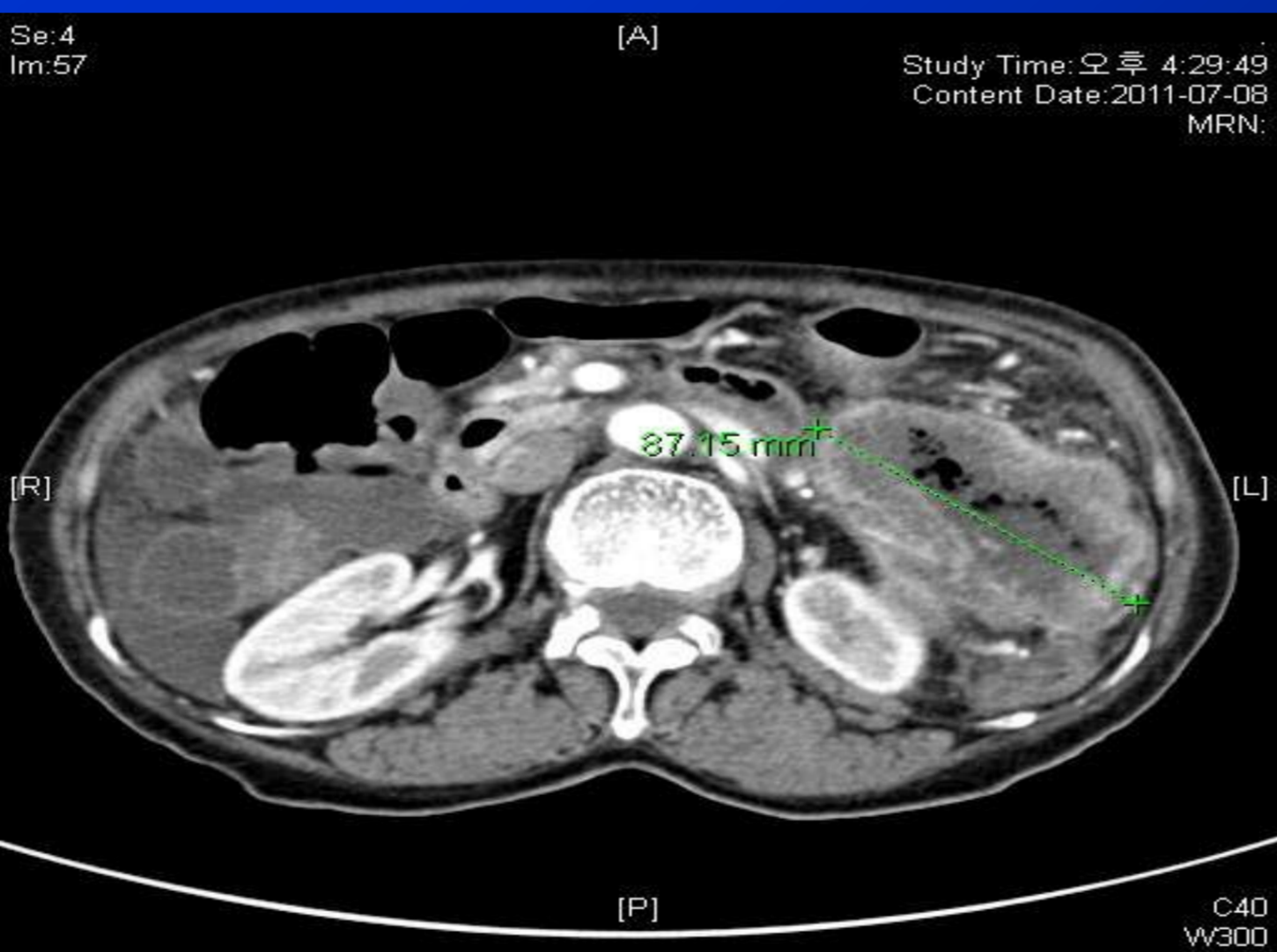


Fig. 2. presented about 9cm sized mass lesion in the splenic flexure of colon with serosal penetration

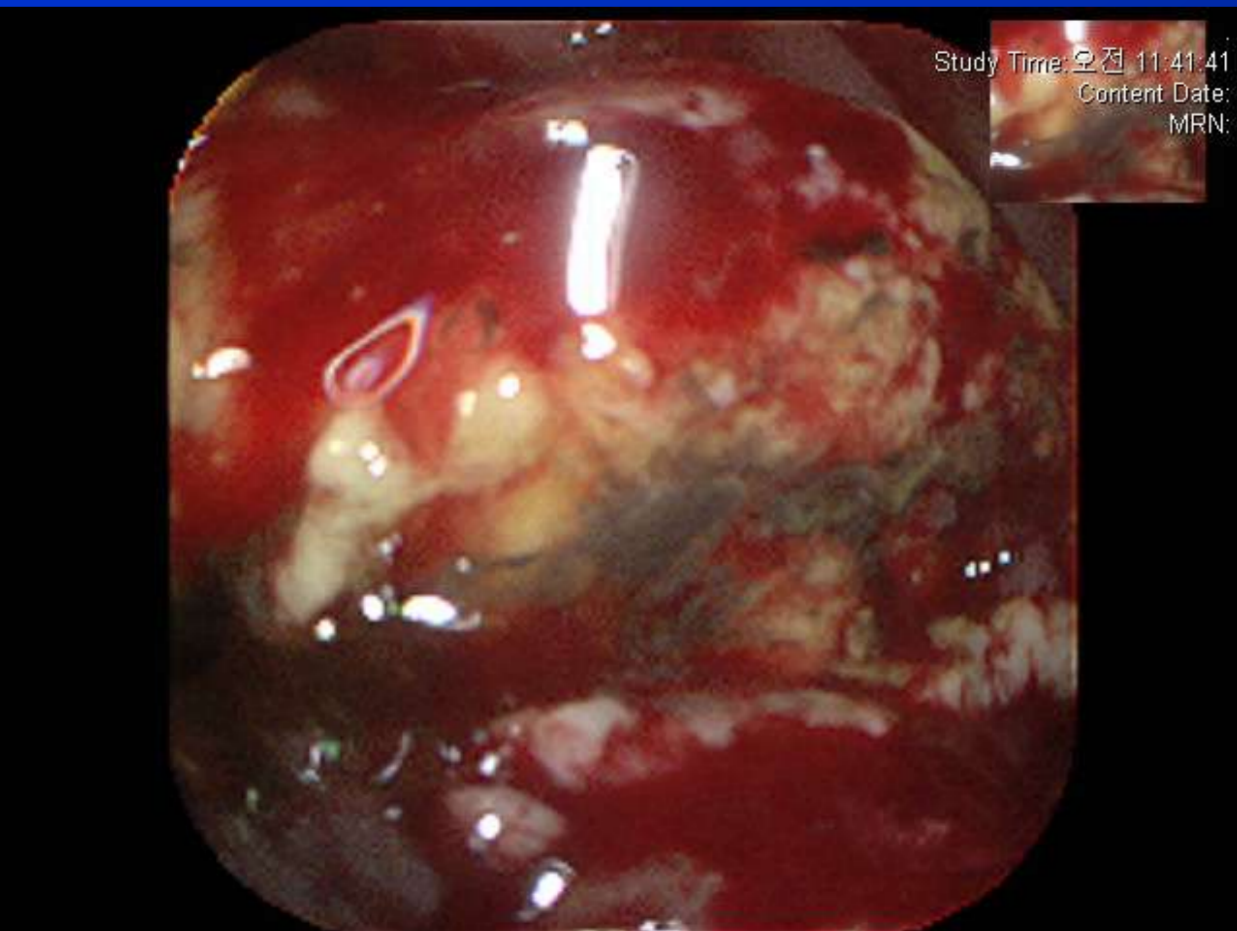


Fig. 3. showed a huge mass encircling the lumen of colon with bleeding



Fig. 4. The specimen showed fibrous adhesion with a central protruding mass formation adhered pancreas tissue