A Rare Case of Idiopathic Retroperitoneal Fibrosis Involving Obstruction of the Mesenteric Arteries, Duodenum, Common Bile Duct, and Inferior Vena Cava

Satoru TAMURA*, Yuichi YOKOYAMA**, Kazuo NAKAJO**, Tomoko MORITA**, Kayoko WADA** and Saburo ONISHI*, **

Abstract

Idiopathic retroperitoneal fibrosis (IRF), usually affects the ureter, although the biliary tree, duodenum and vasculature may also be susceptible. This case report describes a 64-year-old man with IRF, who presented painless watery diarrhea, radiological features of obstructive jaundice and duodenal obstruction, and ultimately an obstruction of the inferior vena cava. We employed tamoxifen for his treatment, but the disease progressed and the patient died of multiple organ failure two years after the onset. While the cause of IRF in this patient was obscure, we suspected his painless watery diarrhea indicated chronic ischemia of the small bowel, and the findings of an abdominal CT scan were extremely valuable in indicating IRF.
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Key words: idiopathic retroperitoneal fibrosis, duodenal obstruction, tamoxifen, painless diarrhea

Introduction

Idiopathic retroperitoneal fibrosis (IRF), a rare finding, was first described by Albarran in 1905 (1) as an inflammatory retroperitoneal process compressing the ureters. A mat of fibrous tissue develops in the retroperitoneal wall without obvious cause. The ureter is the most vulnerable organ, although the biliary tree, duodenum and vasculature may also be affected. The ureteral encasement leads to its obstruction and consequent hydronephrosis. The distal part of the common bile duct can also be affected and may lead to cholestasis (2, 3). Duodenal obstruction due to IRF is a rarely reported clinical entity and the symptoms of nausea, vomiting, and body weight loss are usually presented several months after the initial diagnosis (4). IRF association with vascular obstruction also has been reported rarely (5), and chronic small bowel ischemia has primarily manifested itself by abdominal pain, diarrhea and body weight loss occurring in close relation to meals (6).

Surgery and /or drugs have been employed for the treatment of IRF. Corticosteroids have mainly been used and many cases of IRF have been treated (7). Furthermore, some cases of benign fibrotic tumors and IRF were treated successfully with the estrogen receptor antagonist, tamoxifen (8).

Here, we describe a patient with IRF, who presented with painless watery diarrhea, radiological features of obstructive jaundice and duodenal obstruction, with the ultimate obstruction of the inferior vena cava.

Case Report

A 64-year-old man was referred to our hospital with a one-year history of body weight loss (12 kg) and a painless watery diarrhea (8–9 times a day) occurring after meals. He was admitted in January 1999. No previous history of special drug ingestion (e.g. methysergide) was elicited. His mother had died of gastric cancer and his grandfather had died of pancreatic cancer. His abdomen was soft and flat with normal bowel sound. A bruit of the abdominal artery was noted by auscultation. Superficial lymph nodes were not palpable. All other physical examinations were unremarkable. His fecal occult blood test was negative and urinalysis was normal with a specific gravity of 1.017. The complete blood cell count showed normochromic normocytic anemia with a hematocrit 32% (normal=40.5–51.5%). The blood urea nitrogen was 23 mg/dl (normal=8–20 mg/dl), and the serum

From *the Department of Endoscopy and **the First Department of Internal Medicine, Kochi Medical School, Nankoku
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Reprint requests should be addressed to Dr. Satoru Tamura, the Department of Endoscopy, Kochi Medical School, Kohasu, Okoh-cho, Nankoku, Kochi 783-8505

creatinine was 0.8 mg/dl (normal=0.4–1.0 mg/dl). Other biochemical screening tests and electrolytes were all within normal limits. Both the carcinoembryonic antigen (CEA) and CA19-9 level were within normal ranges.

Abdominal ultrasonography, gastrointestinal endoscopy, and colonoscopy showed no remarkable findings. CT scan of the abdomen demonstrated evidence of periarterial (superior mesenteric arteries) soft tissue suspect of retroperitoneal fibrosis, but no sign of hydronephrosis (Fig. 1A). Abdominal angiography was subsequently performed, and revealed a narrowing of the origin of the superior mesenteric arteries and its branches (Fig. 2A), and slight extrinsic compression of the origin of the celiac axis (Fig. 2B). Inferior mesenteric arteries were normal. These changes in the artery were localized at its origin from the aorta, and its peripheral parts were smooth. Collateral flow to the superior mesenteric arteries was not derived from the celiac artery or the inferior mesenteric arteries. We ascribed the symptoms to small bowel ischemia.

The patient started taking tamoxifen, 10 mg twice a day, in March 1999. In July 1999, his diarrhea improved (4–5 times a day) but the periarterial soft tissue was not changed (Fig. 1B). In August 1999, an abdominal sonographic examination detected the dilatation of intrahepatic bile ducts and common bile duct (CBD). Endoscopic retrograde cholangiopancreatography (ERCP) showed a narrowing of the common bile duct with a small stone. A few days after the ERCP, he was readmitted with severe nausea and vomiting. Upper gastrointestinal series showed almost total obstruction of the third portion of the duodenum with proximal dilatation (Fig. 3). To assess the lesion, abdominal angiography was reperformed, which revealed the aggravated narrowing of the origin of the superior mesenteric arteries and its branches (Fig. 2C), as well as the aggravated narrowing of the origin of the celiac axis (Fig. 2D). He discontinued taking tamoxifen.

After eight weeks of hospitalization, laparotomy revealed a diffuse, poorly delineated, focally dense fibrotic retroperitoneum, which encircled the third and fourth portions of the duodenum and abdominal aorta. Gastrojejunostomy was performed in November 1999. Histological assessment of the tissue from three retroperitoneal sites showed fibrous tissue and fat with no evidence of malignancy (Fig. 4). This operative intervention improved his symptoms of nausea and vomiting, and the patient resumed eating.

However, the bile duct dilatation did not improve, along with the following: alkaline phosphatase 698 IU/*l* (normal 70–220); gamma-glutamyl transpeptidase 246 IU/*l* (normal 5–50); aspartate aminotransferase 59 IU/*l* (normal 9–27); alanine aminotransferase 147 IU/*l* (normal 5–37); total bilirubin 0.8 mg/dl (normal 0.3–1.1). ERCP demonstrated a stenosis of the CBD and endoscopic sphincterotomy (Fig. 5A) and subsequent transpapillary biliary drainage was performed in January 2000 (Fig. 5B).

The patient started taking prednisolone (PSL), 60 mg a day, in January 2000. In March 2000, PSL was tapered to 20 mg, but narrowing of CBD was not improved. To make matters worse, a liver abscess concurred (Fig. 1C) and severe edema in both legs appeared. Angiography showed a narrowing of the inferior vena cava with collateral flow (Fig. 6). PSL was tapered off and liver abscess was improved. Unfortunately renal dysfunction, abnormality of electrolytes, arrhythmia and pleural effusion appeared in late June 2000. The patient died of multiple organ failure in July 2000.

Discussion

Retroperitoneal fibrosis is a relatively uncommon disease, with its etiology unable to be identified in two-thirds of patients. Drug-induced retroperitoneal fibrosis occurs in about 12%, and malignant disease has been associated with

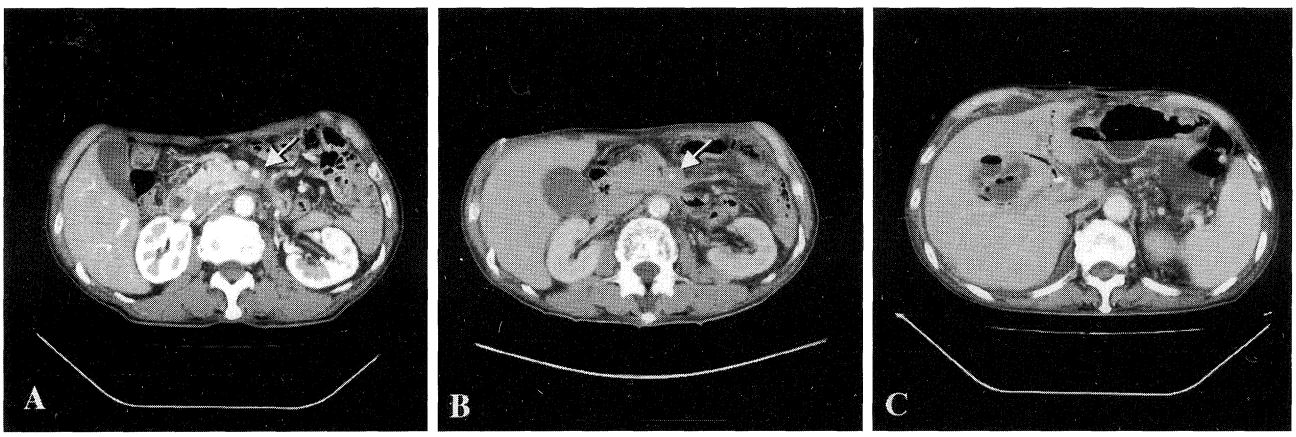


Figure 1. CT scan of the abdomen with intravenous contrast. A. CT scan prior to treatment demonstrate periarterial (superior mesenteric arteries) soft tissue (arrow). B. CT scan performed four months after treatment shows no reduction in the size of the periarterial soft tissue (arrow). C. CT scan shows a low-density lesion of the liver which is compatible with the abscess.

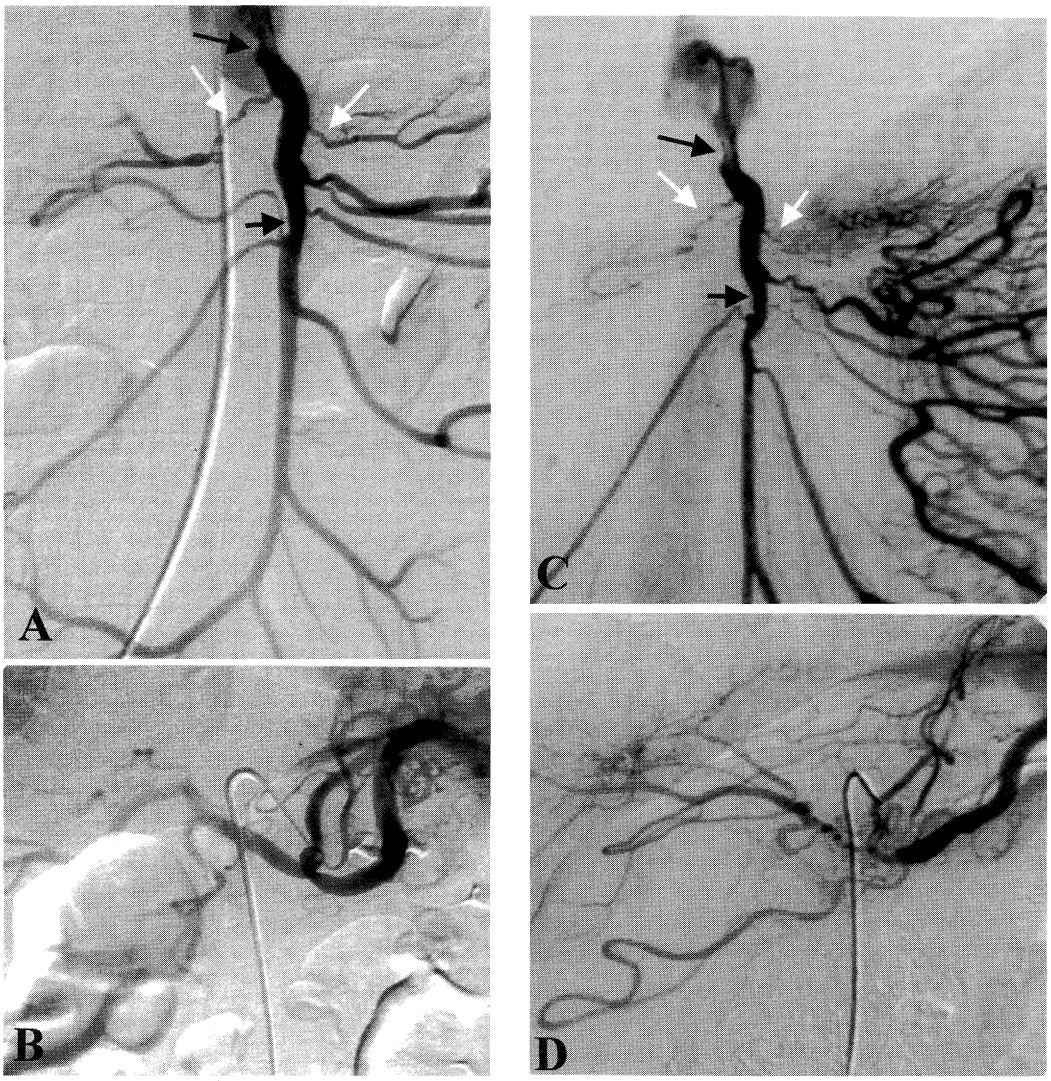


Figure 2. A. Abdominal angiography in January 1999 reveals a narrowing of the origin of the superior mesenteric arteries (black arrows) and its branches (white arrows). B. Abdominal angiography in January 1999 reveals slight extrinsic compression of the origin of the celiac axis. C. Abdominal angiography in October 1999 reveals the aggravated narrowing of the origin of the superior mesenteric arteries (black arrows) and its branches (white arrows). D. Abdominal angiography in October 1999 reveals the aggravated narrowing of the origin of the celiac axis.

retroperitoneal fibrosis in about 8% of patients (2). The clinical features reflect the extent of fibrous tissue proliferation. Usually the fibrous tissue extends from the brim of the pelvis upward to the renal pedicle and ureter; however, occasionally it extends beyond them into the pelvis or through the crura of the diaphragm. The typical patient with IRF is a man in his late 50s (9) and a urinary tract obstruction is the most common clinical manifestation. Review of the literature revealed a few reported cases of retroperitoneal vessel (5, 6, 9–12), duodenal (4) and CBD involvement (3, 13–15). As case reports accumulate, however, it has become more evident that the location and extent of the fibrous tissue may vary widely with the involvement of any retroperitoneal structure. However, a case which coincidently involves vessel, biliary and duodenal obstruction is very rare (16). Furthermore, to the best of our knowledge, no previous case has been reported which coincidently involves the mesenteric artery, biliary, duodenum and inferior vena cava.

The cause of IRF in this patient was obscure, and methysergide, ergotamine and other drugs (analgesics, antihypertensives, etc) were not elicited. Intraabdominal inflammatory conditions or systemic fibrosing disorders or immunological disease were also not implicated. We suspected his painless watery diarrhea to be due to chronic ischemia of the small bowel, and the findings of abdominal CT scanning were extremely valuable in indicating a

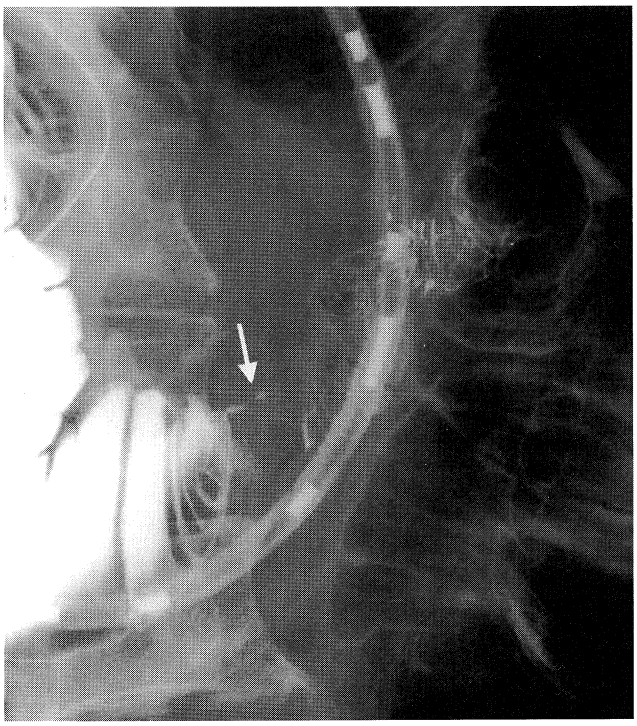


Figure 3. Upper gastrointestinal series shows almost total obstruction (arrow) of the third portion of the duodenum with proximal dilatation.

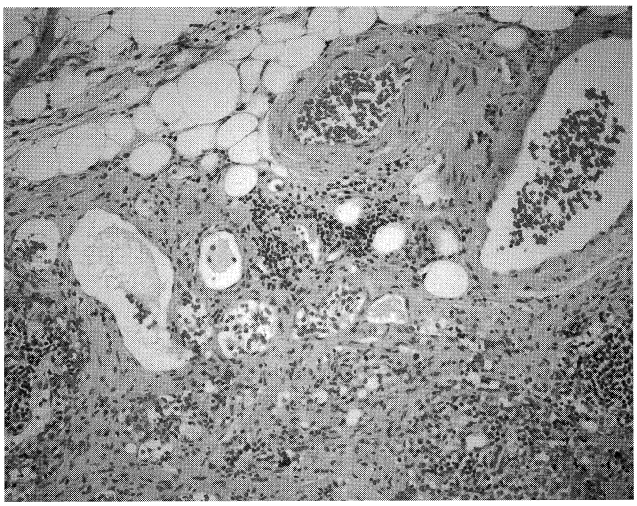


Figure 4. Histology from three retroperitoneal sites reveals fibrous tissue, fat and patchy lymphocytic infiltrates with no evidence of malignancy (HE stain, ×50).

suspicion of IRF. Following a previous report of chronic small bowel ischemia, clinical descriptions of the condition have emphasized the universal presence of food-related epigastric pain of intestinal angina. The case described here, however, strongly suggested that chronic ischemia of the small bowel can occur in the absence of typical pain, and that this condition should be considered in the differential diagnosis of painless water diarrhea in the context of weight loss(5, 6, 10–12). We estimated that the mass surrounding the superior mesenteric artery of CT scan was due to retroperitoneal fibrosis, and that this was the cause of the small bowel ischemia which induced the painless watery diarrhea of our patient. The narrowing of the superior mesenteric arteries of abdominal angiography was compatible with the CT scan findings.

Duodenal obstruction produces vomiting as one of the presenting symptoms, though this is a rare clinical entity (4, 16). The third portion may be the most vulnerable portion of the duodenum in patients with IRF.

Retroperitoneal fibrosis with cholestatic jaundice due to an affection of the distal part of the common bile duct is also a rare condition and difficult to distinguish from cholangiocarcinomas (2, 3, 13, 14), or pancreatic cancer (15). Wetter et al reported that 31% of Klatskin tumors (30/98) were ultimately diagnosed as other than cholangiocarcinomas, and that 3 cases of IRF were included in Klatskin tumors (14).

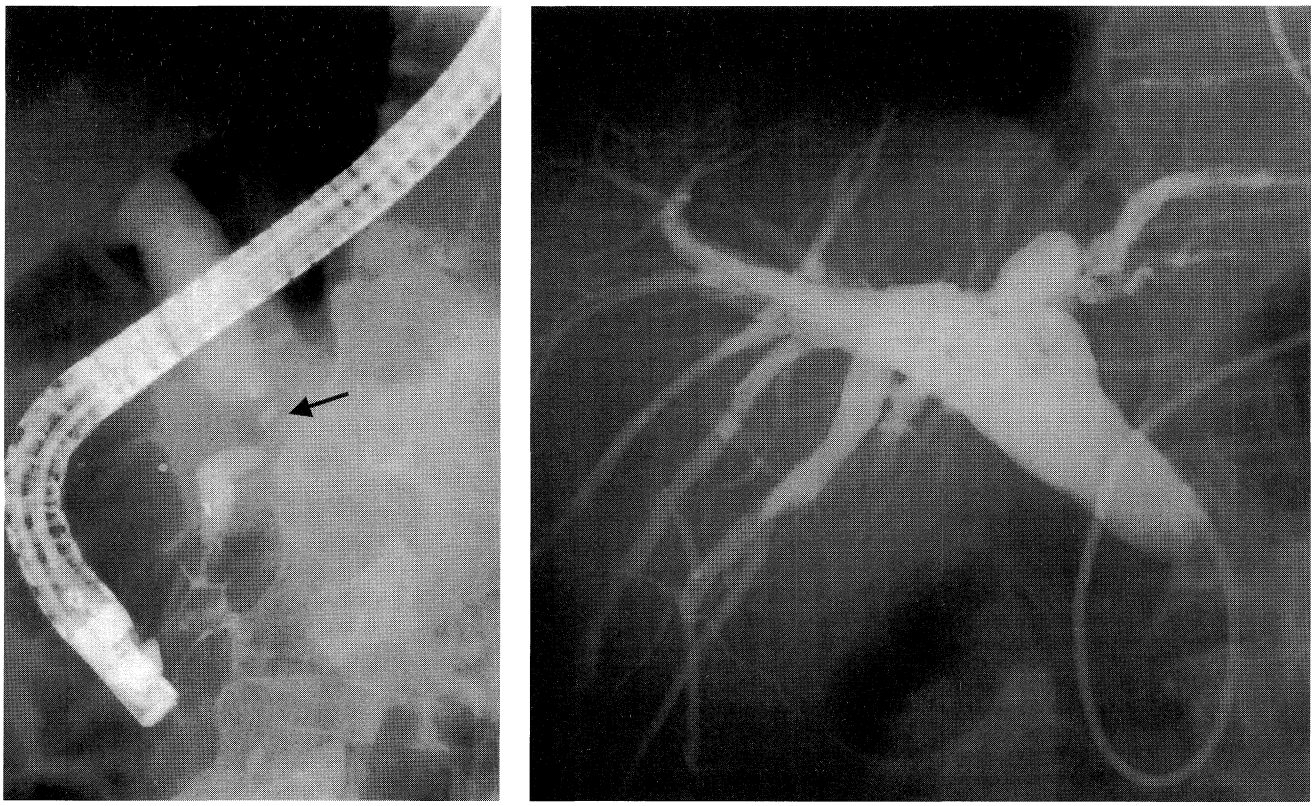
The cholangiogram of this case showed smooth stenotic change, and the cytology of bile from the transpapillary biliary drainage tube showed no malignancy; these findings indicated that the obstructive jaundice of this case originated in the retroperitoneal fibrosis.

Severe edema of both legs due to a narrowing of the inferior vena cava may occur in the late stage of IRF (9), as with our case. Angiography of our case showed a collateral flow, but caput Medusa or other obvious signs of the development of a venous collateral circulation of the surface of body were not noted.

The lack of ureteral involvement may be attributed to the site of the fibrosis in this case because fibrous change usually occurs and extends from the brim of the pelvis upward to the renal pedicle and ureter in almost all cases of IRF.

The treatment of IRF has not been well established. Operative intervention followed by steroid therapy is selected in most cases of ureteral or alimentary tract obstruction (3, 4, 7, 17, 18). However, there is still disagreement concerning the efficacy of corticosteroids (19). Patients with retroperitoneal fibrosis may be divided into two defined groups (7): the first group is those patients who are the most seriously ill and have a poor response both to surgery and corticosteroids; the second group consists of the less seriously ill patients who are presumably in the early stages of the disease. The response to corticosteroids may depend on the pathologic stage of the disease condition. On the other hand, success with the estrogen receptor antagonist, tamoxifen, in the treatment of retroperitoneal fibrosis has been reported (8). Unfortunately, our patient did not respond to either corticosteroids or tamoxifen and died nineteen months following the first admission.

We considered that the response to treatment for IRF mainly depends on the stage of disease, and the patients in the most advanced stage may fail to respond favorably.



A B

Figure 5. A: ERCP demonstrate a tapered stenosis of the common bile duct (arrow). B: Cholangiography using a transpapillary biliary drainage tube shows almost complete obstruction of common bile duct and dilated intrahepatic bile ducts.

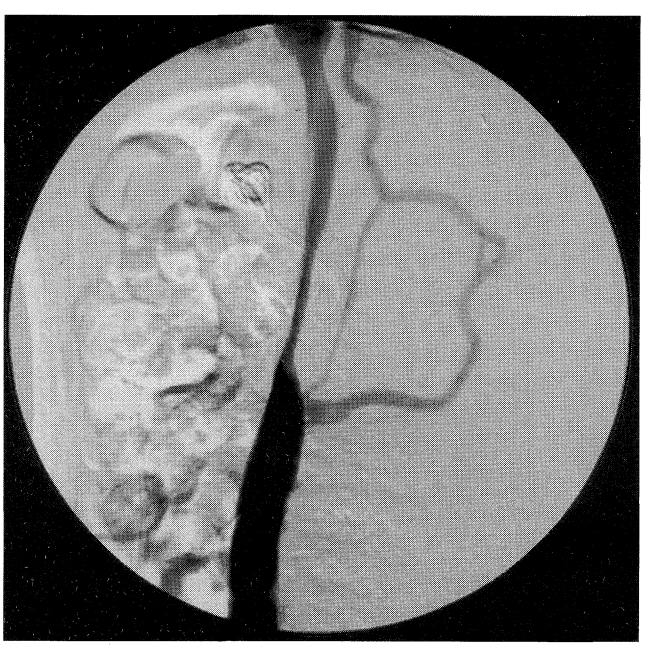


Figure 6. Angiography shows a tapered narrowing of the inferior vena cava with collateral flow.

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