Turk J Gastroenterol 2005; 16 (3): 156-159

Pancreatic mass lesion mimicking carcinoma: Initial presentation of retroperitoneal fibrosis

Retroperitoneal fibrosisin ilk bulgusu olarak kansere benzeyen pankreatik kitle lezyonu

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A 63-year-old woman with upper abdominal pain radiating to her back was admitted to the hospital, and a mass lesion was identified involving pancreatic body and tail, with peripancreatic lymph node enlargement. The patient had normal tumor marker levels, a very high erythrocyte sedimentation rate (104) and negative malignant cells in fine needle aspiration cytology. The patient underwent laparotomy and histological findings of mass were consistent with retroperitoneal fibrosis. She has been in remission after long-term steroid treatment. In the differential diagnosis of cases with pancreatic mass lesion, retroperitoneal fibrosis should also be considered.

Key words: Retroperitoneal fibrosis, pancreatic cancer

Karın ve sırt ağrısıyla müracaat eden, 63 yaşındaki kadın hastanın pankreas gövde ve kuyruk kısmını tamamen infiltre eden, lenf bezi büyümesiyle beraber olan kitle saptandı. Hastaya sedimentasyon yüksekliği (104 mm/h), tümör markerlerinin negatifliği ve ince iğne aspirasyon biyopsisinde malignite saptanmaması nedeniyle laparatomi yapıldı. Laparatomi sonrası retroperitoneal fibrosis tanısı konuldu. Hasta steroid tedavisi sonrası remisyona girdi. Retroperitoneal fibrosis pankreas tümörleriyle karışabilen nadir sebeplerden biridir.

Anahtar kelimeler: Retroperitoneal fibrosis, pankreas kanseri

INTRODUCTION

Idiopathic retroperitoneal fibrosis (IRF), or Ormond's disease, is characterized by the extensive development of inflammatory fibrotic tissue in the retroperitoneum, leading to the compression and obstruction of the ureters and other adjacent organs (1, 2). Aorta, vena cava, mesenteric vessels, biliary ducts (3), duodenum (4) colon (5) and pancreas (2, 6, 7) may rarely be involved. We report a patient with pancreatic mass lesion in the setting of IRF.

CASE REPORT

A 63-year-old woman presented in May 2000 with upper abdominal pain radiating to her back. She had lost 4 kg in weight during the past three months. She denied nausea, vomiting, fever and dark urine. Her physical examination was normal. Her complete blood count, biochemistry, Ca 19-9, carcinoembryonic antigen and α-fetoprotein were within normal limits. Erythrocyte sedimentation rate was 104 mm/hour. Esophagogastroduodenoscopy, colonic barium enema and small bowel enema studies were within normal limits. Abdominal ultrasound showed a pancreatic mass involving the pancreatic body and tail with peripancreatic lymph node enlargement. Abdominal computerized tomography (CT) revealed a mass lesion involving the pancreatic body and tail with peripancreatic lymph node enlargement, and no evidence of biliary dilatation, focal liver lesion or ureteral obstruction. On CT, mass lesion was encasing the vena cava and aorta (Figure 1). There was no evidence of malignancy on cytological examination of the ultrasound-guided fine needle aspiration biopsy specimen of the pancreatic mass.

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Manuscript received: 04.10.2004 **Accepted:** 14.12.2004

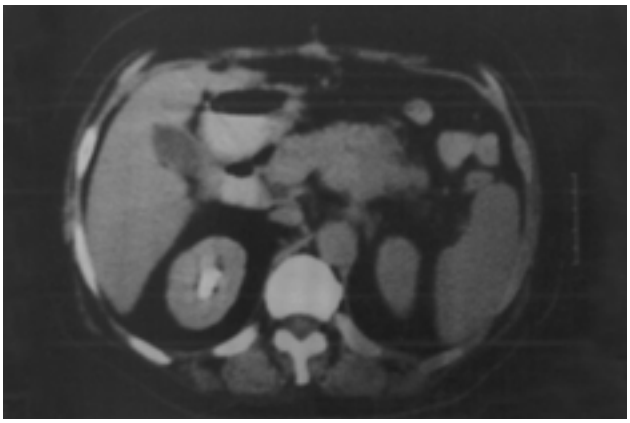

Figure 1. On abdominal CT, mass lesion involving pancreatic body and tail (before prednisone administration; October 2000)

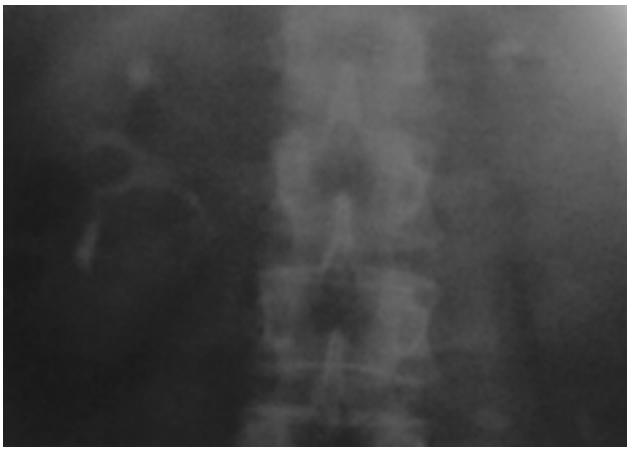

Figure 2. Complete left ureteral obstruction (August 2001; one year after initial prednisone discontinuation)

The patient subsequently underwent exploratory laparotomy with nodal and pancreatic biopsies on 11 September 2000. At laparotomy, a large hard tumor was found invading into the pancreatic body and tail with peripancreatic, paraaortic and paracaval lymph node enlargement. The tumor was attached to, but not infiltrating, the vena cava and aorta. Frozen sections of the pancreas suggested normal pancreatic tissue within adipose tissue. In the final histological examination, the lesion was composed of chronic inflammation consisting of broad collagen fibers, abundance of plasma cells and vascular proliferation. There was no evidence of malignancy. Histologically, it gave a typical picture of IRF.

Oral steroid as an initial dose of 60 mg of prednisone was started. This dose was tapered during the next two months to a maintenance dose of 8 mg per day. Total duration of prednisone use was three months. The patient's clinical symptoms resolved, erythrocyte sedimentation rate decreased to normal level (15 mm/hour) and the pancreatic mass lesion regressed. The patient discontinued maintenance prednisone therapy after three months.

In August 2001, the patient was admitted to the hospital for evaluation of severe left costolumbar pain. The CT and abdominal ultrasound findings included a presacral mass surrounding the aorta, inferior vena cava and the left ureter with left hydronephrosis. There was also progression in the pancreatic mass lesion. Intravenous pyelography showed complete left ureteral obstruction (Figure 2). Erythrocyte sedimentation rate was elevated again (100 mm/hour). The patient was again administered prednisone at 60 mg/day. This dose was tapered during the next three months to a maintenance dose of 4 mg daily. The patient was

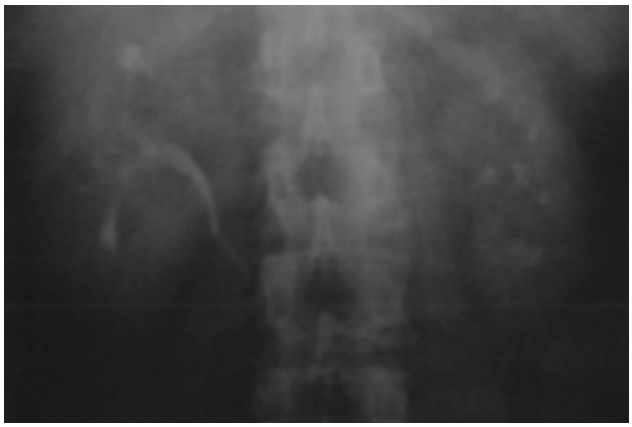

Figure 3. Disappearance of hydronephrosis after prednisone re-administration (September 2001)

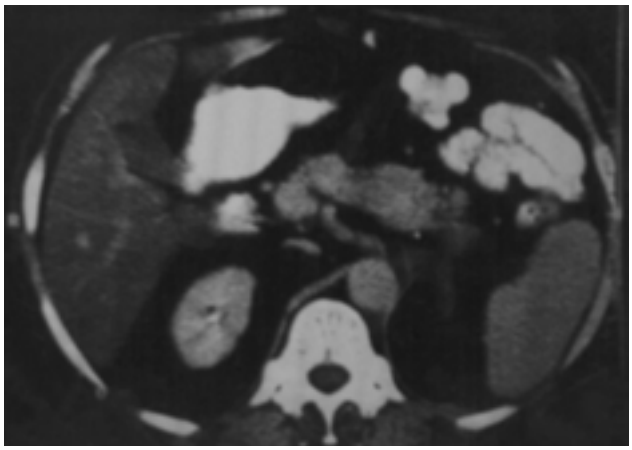

Figure 4. Residue pancreatic mass in pancreatic body and tail during clinical remission (June 2004)

relieved of left costolumbar pain, hydronephrosis disappeared (Figure 3), and the pancreatic mass regressed again (Figure 4). The patient discontinued maintenance prednisone in September 2003, and was still in remission in July 2004.

DISCUSSION

Retroperitoneal fibrosis (RF) is characterized by the extensive development of inflammatory fibrotic tissue in the retroperitoneum. However, clinical presentation and radiological appearances in many cases are variable (4). RF may result in discrete retroperitoneal masses, although it is more often a bilateral disease of the retroperitoneum and may involve the ureters, kidneys, great vessels, and biliary tree, resulting in obstruction of these structures (8). RF locates most commonly as an isolated fibrotic plaque in the lower lumbar region (4) and rarely in peripancreatic, periduodenal, and pelvic regions (4, 8). Diffuse pancreatic involvement (6), pancreatic head (2, 7, 8), complete colonic obstruction (5), and renal vein hypertension from unilateral RF (9) were reported previously. Sign and symptoms of RF are nonspecific. They are mainly related to entrapment and compression of retroperitoneal structures. The commonest symptom is poorly localized back pain. Other clinical findings are anorexia, fatigue, anemia, weight loss, mild fever and malaise. The diagnosis is usually made late and is associated with significant morbidity due to progressive renal failure (4). A difficulty with the diagnosis of RF is that biopsies are often non-diagnostic. Of particular concern are malignant tumors that cause secondary retroperitoneal fibrosis and may be associated with an inflammatory reaction (8).

In our patient, clinical symptoms and radiological findings supported pancreatic malignant tumor at an advanced stage. But in contrast to such an advanced pancreatic mass, normal tumor marker levels, very high erythrocyte sedimentation rate, absence of any imaging findings of metastasis, and negative malignant cells in fine needle aspiration cytology were not consistent with a malignant pancreatic disease. These findings supported an inflammatory process rather than a neoplastic tumor. Therefore, the patient underwent exploratory laparotomy and histological findings were consistent with IRF. Initial clinical presentation of our patient was atypical, but, at recurrence one year later, clinical and imaging findings were typical for RF.

Surgical interventions such as ureterolysis have been the preferred primary mode of treatment because it relieves ureteral obstruction and a specimen can be obtained for histological diagnosis to rule out malignancy (2, 10). However, the effectiveness of steroids (3, 4, 8, 11) immunosuppressive drugs (1), and tamoxifen (12) administration have been reported previously. It has also been reported that steroid therapy is more effective for cases showing high inflammatory activity (11). There is little reported information about the dose and the duration of these medications. Kardar et al. (3) treated 12 patients with RF and their patients were given steroid for two years. Good response in the form of relief of symptoms and regression of the mass in nine cases and treatment failure in three patients were reported. Symptom recurrence after discontinuation of steroid and a further small dose of steroid administration for maintenance was required in one patient. Our patient did not need surgical intervention for the treatment. Exploratory laparotomy was performed for definitive diagnosis. Complete clinical and laboratory remission and significant regression in the pancreatic mass occurred in our patient after long-term steroid administration. We also observed that steroid can relieve ureteral obstruction without a surgical intervention, and that RF may stay in remission for a long term after steroid discontinuation. Steroids may be used as the primary mode of treatment for patients with IRF with minimal complications.

In conclusion, in the differential diagnosis of cases with a pancreatic mass lesion and with normal tumor marker levels, high erythrocyte sedimentation rate and absence of findings of metastasis, RF should be considered.

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