Desmoid Tumor Causing Duodenal Obstruction

Haggi Mazeh MD[1], Aviram Nissan MD[1], Natalia Simanovsky MD[2] and Nurith Hiller MD[2]

Departments of [1]Surgery and [2]Radiology, Hadassah University Hospital, Mount Scopus, Jerusalem, Israel

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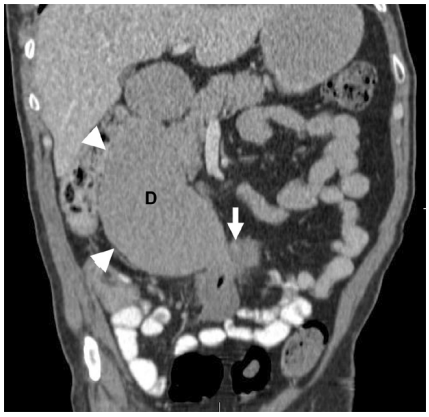
Intraabdominal desmoid tumors are rare soft tissue tumors, usually associated with familial adenomatous polyposis or prior abdominal surgery. The sporadic form of IADT is rare, presenting with a mass effect or abdominal pain. We describe a 55 year old man with a sporadic IADT presenting as chronic duodenal obstruction. The tumor encased the third part of the duodenum encoring the duodenum to the mesenteric mass and causing consequent obstruction. The tumor was not resectable and was surgically bypassed. Duodenal obstruction by desmoid tumor is extremely rare. To the best of our knowledge only one similar case has been published.

Patient Description

A 55 year-old man was admitted to the hospital because of ongoing complaints of early satiety and recurrent vomiting. Past medical history included type 2 diabetes mellitus and spastic hemiparesis due to past cerebral hemorrhage. He had no history of abdominal trauma or surgery. On physical examination a non-tender solid mass about 10 cm in diameter was palpated above the umbilicus.

Gastroscopy with biopsies revealed esophagitis and a severe duodenal dilatation with no evidence of an obstructing mass. Random biopsies were negative for malignancy. An upper gastrointestinal barium series demonstrated severe dilatation of the second part of the duodenum and narrowing of the third part of the duodenum caused by an extra-luminal mass with slow passage of barium distally. Abdominal computed tomography revealed a spiculated, homogenous solid mesenterial mass, encasing the third part of the duodenum with near obstruction. The mass was exophytic, inseparable from an adjacent small bowel loop, and extended into the mesentery while anchoring the duodenum to the root of mesentery [Figure]. Air was evident in the mass, implying duodenal-tumoral fistula. The tumor was elongated in shape, extending from the duodenal level into the pelvis and measured 20.0 x 6.5 x 4.5 cm. CT-guided core biopsy showed fibromatous tissue with no evidence of malignancy.

Surgery confirmed the CT findings. Frozen section biopsy was consistent with fibromatosis. The mass was clearly unresectable, therefore it was bypassed by a gastro-jejunostomy with a small to small bowel anastomosis to avoid future intestinal obstruction. The postoperative course was uneventful. The patient was then treated by tamoxifen and sulindac. Twelve months after the operation the patient is well with no signs of obstruction.



CT of the abdomen with coronal multiplanar reconstruction showing solid mesenteric mass inseparable from the duodenum (arrow). Note the massive dilatation of the second part of the duodenum (arrowheads). The contrast medium in the duodenum is diluted with a large amount of liquid secretions. A tract of gas is seen within the mass, suggesting a duodenal-tumor fistula.

Comment

"Desmoid" is derived from the Greek word *desmos*, meaning band-like. Desmoid tumors are benign slow-growing neoplasms. Although locally invasive, desmoids do not have the capacity to metastasize [1]. The estimated incidence in the United States is 3 per million per year. The etiology of desmoid tumors is unknown. There is an association with some clonal chromosomal changes, e.g., trisomy 8 or 20 which are associated with higher incidence of benign fibrous lesions in soft tissue and bone.

Intraabdominal desmoid tumors most commonly occur in patients with Gardner's syndrome (familial adenomatous polyposis), especially following surgery for carcinoma of the colon [1,2]. There also appears to be a relation between IADT and abdominal trauma, especially surgical trauma [1] and Crohn's disease. Sporadic IADT are rare.

Desmoid tumors occur in various sites of the body but most commonly in the torso and extremities. Intraabdominal desmoids are less frequent. IADT usually arise from the abdominal wall and only rarely originate from intraabdominal structures. Mesenteric fibromatosis commonly arises from the mesentery of the small bowel,

IADT = intraabdominal desmoid tumors

but can also originate from the ileocolic mesentery, gastrocolic ligament and omentum. As desmoid tumors are slow-growing, they cause only subtle clinical symptoms. Most tumors are large at the time of diagnosis. Left untreated, IADT infiltrate neighboring structures and can cause significant morbidity. Most tumors usually present as an asymptomatic or minimally painful mass, but as the disease progresses it can lead to serious complications including small bowel obstruction and mucosal ischemia.

Although the presence of a slow-growing mass at the root of the mesentery is suggestive for IADT, the radiologic appearance is non-specific and cannot reliably distinguish IADT from low grade retroperitoneal sarcoma.

Ultrasonography, CT and magnetic resonance imaging can reveal the intraabdominal solid mass, the exact lesion, and the extent and involvement of adjacent structures. MRI may show characteristic features of prominent low to intermediate signal intensity and bands of low signal intensity representing highly collagenized tissue. However, lesions with less collagen and more cellularity may have non-specific high signal intensity on T2-weighted images [3]. The diagnosis is usually established by needle or surgical biopsy. Incisional biopsy is preferred due to the need to exclude a malignant process with a high degree of confidence.

Surgical treatment of IADT is indicated in symptomatic patients or in case of risk to adjacent structures. Since this tumor has a high recurrence rate, complete surgical resection should be attempted. Because of the close proximity of these tumors to the root of the small bowel mesentery, aggressive resection may cause ischemia of the small bowel with the need for life-time total parenteral nutrition. Therefore, and because of the indolent course of these lesions, major surgical morbidity should be avoided. An aggressive approach should be attempted only in small tumors located away from the superior mesenteric artery. Other treatment options include radiation therapy and chemotherapy. Low dose radiation alone or as adjuvant to resection proved to be very effective for local control of the tumor [1]. Non-cytotoxic medical therapy includes non-steroidal anti-inflammatory agents (sulindac) and hormonal therapy (tamoxifen) alone or in combination [4] Cytotoxic therapy is administered in cases of failure of the non-cytotoxic medical regimen The combination of methotrexate with vinblastine resulted in stabilization of the disease in a substantial number of patients with the aggressive form of the disease.

Our patient demonstrated a rare presentation of sporadic IADT. The radiologic presentation of a soft tissue mass obstructing the third part of the duodenum with megaduodenum is unusual, with only one previous report found in the literature [5]. The differential diagnosis includes malignant or benign primary tumor of the duodenum (carcinoma, lymphoma, mesenchymal tumors, hamartoma, hemangioma etc.), pancreatic or biliary tract carcinoma, complicated pancreatitis and metastasis.

In summary, an unusual case of duodenal obstruction due to IADT is presented. The patient underwent palliative surgery with satisfactory outcome.

References

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Correspondence: Dr. N. Hiller, Dept. of Radiology, Hadassah University Hospital, Mount Scopus, P.O. Box 24035, Jerusalem 19240, Israel.
Phone: (972-50) 787-4980
Fax: (972-2) 643-0337
email: hiller@netvision.net.il