

Gastrointestinal Stromal Tumor of the Ampulla of Vater

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Gastrointestinal stromal tumors are the most common mesenchymal derived tumors of the intestinal tract, though they account for only about 1% of all gastrointestinal tract tumors. GISTs show differentiation toward the interstitial cells of Cajal, which are referred to as the GI pacemaker cells [1]. Most GISTs carry a gain of function mutation in the *c-KIT* (CD 117) proto-oncogene, which is the target for the inhibitor imatinib mesylate. About 95% of GISTs are *c-KIT* positive: 60–70% for CD34, 30–40% for smooth muscle actin, 5% for S-100 protein, and 1–2% for desmin or keratin [2].

GISTs frequently metastasize to the liver and the peritoneum but rarely to regional lymph nodes. These tumors can occur throughout the gastrointestinal tract, most commonly in the stomach (50–60%), 25–30% occur in the jejunum/ileum and 10% in the colon or esophagus. Only 5% of GISTs are located in the duodenum and they rarely present with obstructive jaundice [1]. The presence of GIST in the ampulla of Vater is even rarer. A search of the literature revealed only eight such cases. We report here an additional case of GIST in the ampulla of Vater, treated by means of pancreaticoduodenectomy.

PATIENT DESCRIPTION

An 83 year old woman underwent computed tomography of the chest as part of a workup for non-specific chest pain. A mass was noticed in the head of the pancreas. A subsequent abdominal CT scan revealed a solid lesion of 3.2 cm in the pancreatic head. Neither biliary nor pancreatic duct dilatation was observed and no involvement of the nearby vasculature was noted. The mass appeared hypervascular. On the basis of these findings, the diagnosis of pancreatic neuroendocrine tumor was strongly suspected. The patient was asymptomatic, without abdominal pain, weight loss, jaundice or weakness. Her medical history was consistent with type II diabetes and hypertension. At hospital admission, her physical examination was unremarkable. Blood tests were normal, including levels of serum tumor markers: carcinoembryonic antigen and carbohydrate antigen, 1.3 ng/ml and 19-9 6.7 U/ml respectively. An upper endoscopy was normal. The hypervascularity of the tumor precluded a biopsy; the size of the mass prompted surgery.

At surgery a firm lesion was detected in the head of the pancreas. No other gross pathology was observed. A Whipple procedure (pancreaticoduodenectomy) was performed. The patient was discharged 10 days later after an uneventful postoperative course. On gross examination of the duodenum, the ampulla of Vater appeared distorted by a 3.2 cm mass invading the head of the pancreas. Microscopic pathological findings revealed a mass originating from and completely distorting the ampulla of Vater and invading the pan-

creatic tissue [Figure A]. The mass was composed of bundles of spindle-shaped tumor cells [Figure B]. No mitoses were found. The mucosa of the duodenum was intact. Immunohistochemistry for *c-KIT* [Figure C] and *DOG1* [Figure D] was positive. The resection margins were free of tumor cells. Results of a histopathological examination of 10 regional lymph nodes were negative. The pathological examination confirmed that the lesion was a gastrointestinal stromal tumor in the ampulla of Vater.

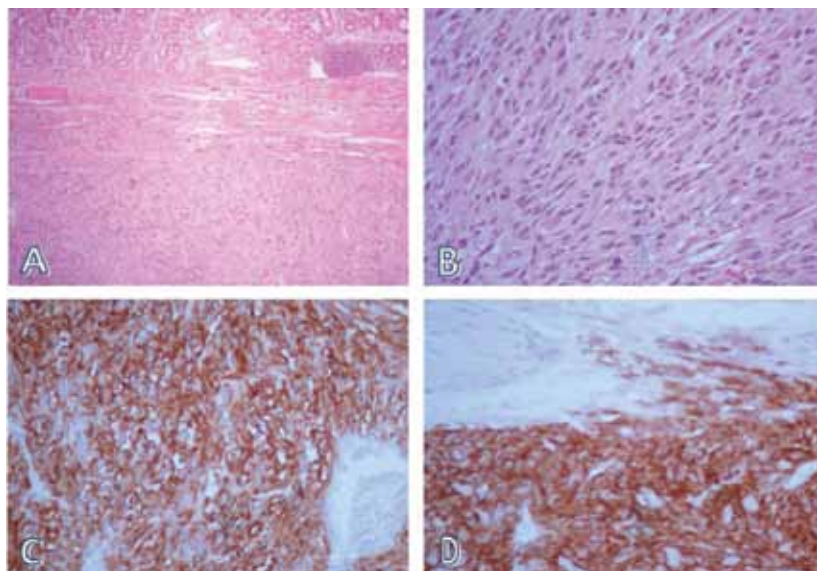
COMMENT

Gastrointestinal stromal tumors are the most common mesenchymal tumors of the gastrointestinal tract. GISTs of the duodenum are rare, and of the ampulla of Vater, especially rare [1]. We present a gastrointestinal stromal tumor in the ampulla of Vater that was discovered incidentally in an asymptomatic 83 year old woman. Review of the eight documented cases of GIST in the ampulla of Vater reveals that half presented in females [3]. The mean age at diagnosis was 56 years.

Similar to most of the published cases, our patient did not present with obstructive jaundice; neither were her bile ducts dilated, though the tumor did originate from the ampulla of Vater itself and distorted its anatomy. Weight loss was the presenting symptom in two cases in the literature, jaundice in two, and melena in three. In six patients a GIST/mesenchymal tumor was diagnosed preoperatively using endoscopy with tissue biopsy. Endoscopic retrograde cholangiopancreatography was conducted in only two of the patients documented. While endoscopic ultra-

GIST = gastrointestinal stromal tumors

[A] Microscopic examination of the sample showed a tumor cell mass adjacent to the duodenal Brunner glands (x 100)
[B] Higher magnification (x 400) shows that the tumor is composed of bundles of spindle cells
[C] Immunohistochemistry for c-KIT was positive (x 400)
[D] Tumor cells showing immunoreactivity for DOG1 (x 400)



sound was not routinely performed, it did suggest the diagnosis of GIST preoperatively in two patients.

The diameter of the mass in our patient would have ruled out local excision. However, since histological diagnosis was not possible due to hypervascularity, the decision for surgery was inevitable. Of the eight patients documented in the literature, five underwent a pancreaticoduodenectomy with an uneventful recovery; two underwent local excision, and one, who was diagnosed with an unresectable lesion due to liver metastasis, died from liver failure [3].

In a recently published analysis of 25 patients with duodenal GIST who

underwent surgery at the National Taiwan University Hospital, Tien et al. [4] concluded that, while 20–86% of duodenal GISTs are treated with PD, limited resections (such as a wedge resection) can effectively treat a duodenal GIST depending on the location and the size of the tumor. Nevertheless, PD is considered the treatment of choice for a duodenal GIST close to (within 2 cm of) the ampulla of Vater. The authors also acknowledged that in some patients PD is used if there is no histopathological diagnosis prior to surgery or imaging findings that suggest a diagnosis other than GIST. For

PD = pancreaticoduodenectomy

our patient, the preoperative tissue diagnosis would not have changed the surgical decision.

In light of the assessment by Rabin et al. [5] that 24% of GISTs are diagnosed as an incidental finding during surgery or other diagnostic procedure, it seems that GIST should be included in the differential diagnosis of tumors appearing in the duodenal ampulla region. New advances in medical treatment strategy using tyrosine kinase inhibitors may change the management algorithm if diagnosis is confirmed prior to surgery. Medical treatment rather than surgery should be considered in high risk patients or in patients whose tumor location might result in extensive surgery.

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The day the child realizes that all adults are imperfect, he becomes an adolescent; the day he forgives them, he becomes an adult; the day he forgives himself, he becomes wise

Alden Nowlan (1933-1983), Canadian poet, novelist and playwright

The best things in life are not things

Albert Einstein (1879-1955), German-born theoretical physicist who developed the general theory of relativity, effecting a revolution in physics