Case Report **OPEN ACCESS**

Gastrointestinal stromal tumor of the pancreas: A case report and review of the literature

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Citation: Trabelsi A, Ben Yacoub-Abid L, Mtimet A, Ben Abdelkrim S, Hammedi F, Ben Ali A, Mokni M. Gastrointestinal stromal tumor of the pancreas: A case report with review of the literature. **North Am J Med Sci** 2009; 1: 324-326.
Availability: www.najms.org
ISSN: 1947 – 2714

Abstract
Context: Primary stromal tumors of the pancreas are extremely rare. Only four cases have been reported in the literature. We describe a new case and analyze, through a review of the literature, the clinical and pathological data, and the outcome of the previously reported cases. **Case report**: We report the case of a 52-year-old-woman who presented with epigastric pain. Preoperatively, abdominal pelvic computed tomography revealed a pancreatic head mass. The surgery confirmed the presence of a large tumor of the pancreatic head. Pathologic results revealed a malignant gastrointestinal stromal tumor of the pancreatic head, completely removed. There is no recurrence or metastases 10 months after surgery. **Conclusion**: The current case consolidates the possibility that this rare tumor can involve the pancreas as a primary site and that GIST of primary pancreatic localization are usually of high risk of malignancy.

Keywords: Stromal tumor, pancreas, histopathology, CD117.

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Introduction
Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract and occur usually in the stomach and the small bowel [1], they are members of a larger family that can also involve other portions of the digestive tract, omentum, mesentery and retroperitoneum [2].GIST contain a spectrum from minute indolent tumors to sarcomas at all sites of occurrence. These tumors are divided into risk categories rather than into "benign" and "malignant" (very low, low, intermediate and high risk of metastases) according to the tumor size and mitotic activity [3]. The diagnosis is made on the basis of histologic and immunophenotypic features.

Primary GIST of the pancreas is distinctly uncommon and according to the previous reports, only four cases have been described in the literature [1, 4-6]. We report a new case of a primary pancreatic GIST with radiological and pathological documentation, diagnosed after surgery, and we analyze the previously reported cases of primary GIST of the pancreas.

Case Report
A 52-year-old-woman without past medical history, presented with epigastric pain for several months. Physical Examination showed an abdominal mass of the epigastria and a good general condition. Abdominal ultrasonography revealed oval hypoechoic solid nodule of the pancreas. Computed tomography scan of the abdomen showed a 10.5 x 8 x 3 cm lesion with low density in the central part that enhanced very strongly in the arterial phase (Fig. 1). The lesion was localized in the pancreatic head, and there were no signs of dilatation of the biliary or pancreatic ducts.

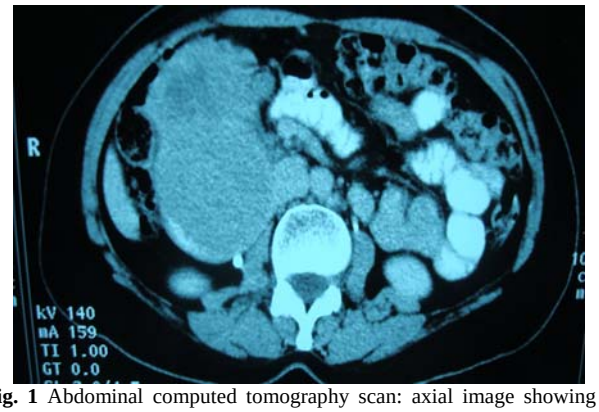


Fig. 1 Abdominal computed tomography scan: axial image showing a solid mass of the pancreatic head.

Hemipancreaticoduodenectomy with antrectomy and partial colectomy were performed.

Macroscopic examination showed a 10.5 x 7 x 3 cm round yellowish mass. It was well-defined but not encapsulated; it was applied to the colon without evidence of intestinal infiltration (Fig. 2). The distance from the resection margin was 5mm.

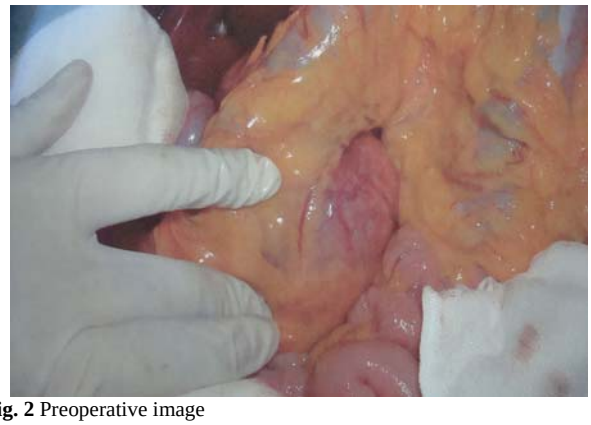
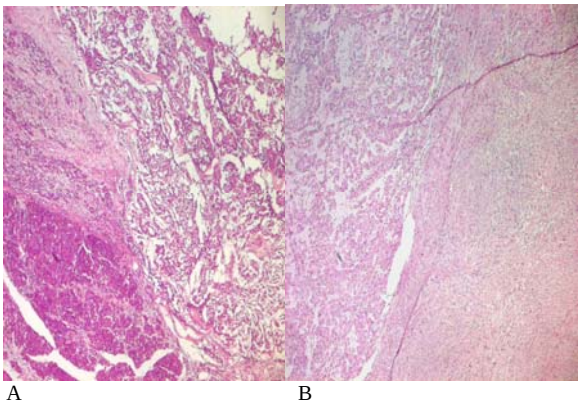


Fig. 2 Preoperative image

Microscopically, the tumor was made of spindle and epithelioid cells (Fig. 3). The mitotic count was 6 mitoses/50 High Power Fields. Immunohistochemical stains showed immunoreactivity for CD117 (c-Kit) (Fig. 4) and CD34, while cells were completely negative for smooth muscle actin, S100 protein, synaptophysin and cytokeratin.



A B

Fig. 3A Tumor surrounded by pancreatic tissue.
Fig. 3B Tumor showing spindle and epithelioid cells.

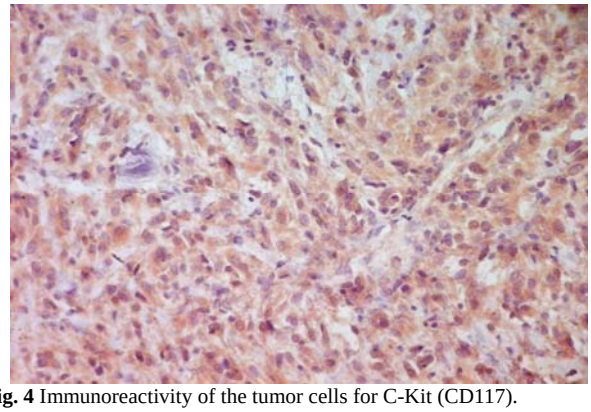


Fig. 4 Immunoreactivity of the tumor cells for C-Kit (CD117).

On the basis of these morphological and immunohistochemical findings, the diagnosis of pancreatic GIST with high risk of malignancy was made. Clinical and radiological findings did not comply with any other primary site disease. Recovery was uneventful and there was no recurrence or metastasis detected during the ten months follow-up examination. The patient didn't receive any adjuvant therapy.

Discussion

Pancreatic GIST is extremely rare. To the best of our knowledge, only four cases have been reported in the literature [1, 4-6]. The clinicopathological features and outcomes of these prior cases are summarized in table I. Patients' age ranged from 38 to 70 years. One patient was male and three were females. The tumor was symptomatic in three cases and asymptomatic in one case. Diagnosis of pancreatic GIST was performed on surgical specimens in three cases and on fine needle biopsy in one case [1, 4]. Tumor was located in the pancreatic head in two cases and in the pancreatic body in the other cases. Pancreatic GIST was of high risk of malignancy in three cases and of low risk of malignancy in one case.

The origin of GIST was at first attributed to Cajal's cells, in mesodermal tissue but it has nowadays been recognized that GIST arise from multipotential mesenchymal stem cells [7]. The clinical presentation is variable but the most usual symptom is the presence of a mass [7]**.** The diagnosis of GIST is based on histological, immunohistochemical and molecular features (oncogenic c-Kit mutation) [5]. Histologically GIST varies from spindle cell tumors to epithelioid and pleomorphic tumors. Most GIST (95%) express c-Kit (CD117), CD34 (70%), and heavy caldesmon (80%), whereas 25% are positive for smooth muscle actin and less than 5% for desmin [8]. The overwhelming majority of GIST are accompanied by a somatic mutation of CD117 (c-kit), a tyrosine kinase receptor normally expressed by the intestinal cells of Cajal, mast cells and germ cells; this mutation occurs independently from anatomic site of the tumor [9].

GISTs contain a spectrum from minute indolent tumors to sarcomas at all sites of occurrence and are divided into risk categories (very low, low, intermediate and high risk of metastases) according to the tumor size and mitotic

activity: tumors larger than 5cm with more than 5 mitoses per 50 HPF as well as tumors larger than 10 cm, regardless of mitotic count, should be considered as lesions with high risk of malignancy [3].

Table 1 Clinicopathological features and outcomes of cases with pancreatic GIST.

Case	Age (yrs)	Sex	Clinical Features	Pancreatic Location	Histological Grading	Outcome (months)
1[4]	68	F	Epigastric pain	Body	High risk for malignancy	Recurrence/peritoneal spread (1)
2[5]	70	F	Asymptomatic	Head	High risk for malignancy	Favorable (7)
3[6]	38	F	Abdominal mass	Body	High risk for malignancy	Favorable (42)
4[1]	47	M	Nausea/bilious vomiting	Head	Low risk for malignancy	?
Our case	52	F	Epigastric pain	Head	High risk for malignancy	Favorable (10)

Standard treatment for primary GIST is complete surgical resection, where feasible, with the aim to obtain negative microscopic margins over the organ of origin. Conventional chemotherapy and radiation therapy have been reported to be ineffective in the treatment of GIST [7]. Until recently no active treatment for advanced or metastatic GIST was available. The advent of target therapy: Imatinib (Gleevec) which is an inhibitor of the tyrosine kinase activity of c-Kit has revolutionized the treatment of this disease and the median overall survival now reaches 5 years [10].

Conclusion

The current case consolidates the possibility that this rare tumor can involve the pancreas as a primary site and that GIST of primary pancreatic localization are usually of high risk of malignancy.

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