Malignant Extra-Gastrointestinal Stromal Tumor (EGIST) of the Pancreas: A Case Report and Review of Literature.

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ABSTRACT

Context Gastrointestinal stromal tumors (GIST) are CD117 (c-Kit) positive mesenchymal neoplasm considered to originate from the interstitial cell of Cajal. GIST has been described outside the gastrointestinal tract in sites like the mesentery, omentum and retroperitoneum; however, pancreatic extragastrointestinal stromal tumor (EGIST) is extremely rare and there only seven previous reports in literature. Case report We describe a 38-year-old gentleman with a malignant pancreatic GIST. The tumor was located in the head of pancreas, measured 6.5x5.0 cm and was well circumscribed. On histology it showed a mixed spindle and epithelioid cell morphology with presence of sheets and short intersecting fascicles of tumor cells. The mitotic count was 12-15 mitoses per 50 high-power fields. The differential diagnosis included a pancreatic smooth muscle tumor and neuroendocrine tumor. Immunohistochemistry revealed diffuse cytoplasmic positivity for CD117 (c-Kit) and vimentin. Tumor cells were negative for CD34, S100, desmin, smooth muscle actin (SMA), cytokeratin, neuron specific enolase, chromogranin and synaptophysin. The patient developed liver metastasis two years after resection of the primary tumor. The resected metastasis showed a similar tumor. The patient was treated with imatinib mesylate and the post-operative course two years after resection of liver metastasis has been uneventful. Conclusion We report a rare case of pancreatic GIST presenting as a solid neoplasm and review the previously described cases in literature.