Answer

Gastrointestinal Stromal Tumor of the Ampulla of Vater

The endoscopic picture (Figure 1) depicted a bulging ampulla of Vater with an essentially normal overlying mucosa except for hyperemia. At EUS (Figure 2), the mucosa (first layer, hypoechoic) appeared uniform and normal and a localized tumor mass was seen originating from the deeper layers. This ruled out an ampullary carcinoma as well as other periampullary and pancreatic head carcinomas or neuroendocrine tumors.

The differential diagnosis in this patient was therefore between tumors arising from the submucosa (neural tumors, lymphoma, or lipoma) and those arising either from the muscularis mucosa or propria (gastrointestinal stromal tumor [GIST], leiomyomas, or leiomyosarcoma). At EUS, the tumor was hypoechoic and originated from the muscularis propria (the fourth layer, which is hypoechoic). The tumors arising from the submucosa (which is hyperechoic) were thus ruled out. The EUS also provided additional information: tumor size of 3 cm, well-defined margins, and homogenous echo pattern with no demonstrable cystic spaces—characteristics associated with benign tumor.1 With leiomyomas being extremely rare, a clinical diagnosis of benign GIST of the ampulla of Vater with gastrointestinal bleeding was therefore made.

The patient underwent pancreaticoduodenal resection (PDR) and had an uneventful recovery. At histopathologic examination, the tumor was composed of spindle cells that showed cytoplasmic positivity for CD117. The number of mitotic figures was 1 per 50 high-power fields, thereby corroborating clinical diagnosis of probable benign GIST.

The value of the currently available computed tomography or magnetic resonance imaging for characterization of small periampullary tumors is limited. Over the last decade, EUS has emerged as an important modality for this purpose.2 The findings in our case suggest that EUS may impact the treatment planning of uncommon periampullary tumors. In patients with EUS diagnosis of localized benign tumors like lipoma or neural tumors, a local resection rather than a major extirpative procedure like PDR may be planned. The EUS guided fine-needle aspiration can be a valuable adjunct.

The GIST of ampulla of Vater is an extremely rare entity with the few cases reported in the literature managed by PDR.3,4 The appropriate treatment, local resection or PDR, remains undefined. The guiding principal for primary localized GIST is resection with negative margins because this represents the only chance for cure. Molecular therapy in the form of a tyrosine kinase inhibitor, imatinib mesylate, is currently recommended for recurrent or metastatic disease or tumors that are large and require extensive resections. The integration of surgery and molecular therapy for primary GIST is currently being investigated.5

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REFERENCES


Due to the overwhelmingly positive response to the Image of the Month, the Archives of Surgery has temporarily discontinued accepting submissions for this feature. It is anticipated that requests for submissions will resume in mid 2008. Thank you.