Malignant small bowel tumors are extremely rare, accounting for 0.1% to 0.3% of all malignancies. Fewer than 2,400 new cases of small bowel malignancy are reported in the United States each year.[1] Malignant tumors, which account for about two-thirds of all primary small bowel tumors, consist of four primary subtypes: adenocarcinoma, carcinoid tumor, lymphoma, and sarcoma (or gastrointestinal [GI] stromal tumor). Each malignancy is characterized by unique predisposing factors, anatomy, and biology. The prevalence, pattern, and relevance of both regional lymph node and distant metastases differ. As a result, the study of malignant small bowel tumors, taken as an aggregate, is fraught with difficulty.

Data Limitations
Although retrospective data offer some insight into the natural history of this group of diseases, there are many limitations. Single-center reviews often describe experience spanning many decades, during which the spectrum of the disease must inevitably change (as, for example, refinements in imaging and improvements in treatment and treatment-related morbidity evolve). Large multi-institutional data sets such as the National Cancer Data Base[2] are limited by internal consistency of data entry from multiple sources, details of treatment, and consistent long-term follow-up. These sources of information are the best we have to define the demographics and natural history of these rare diseases. However, they cannot define the impact of any given treatment on outcome. This can only be accomplished by carefully controlled prospective trials. Given the rarity of these tumors and the enormous resources involved in the successful completion of such trials, it is unlikely that they will ever be conducted. Rather, many of our treatment paradigms will evolve as extrapolations from similar, more common tumors in other locations. Alternatively, it is conceivable that we may be able to discern unique biologic susceptibilities of these tumors by genetic fingerprinting of individual tumors. At present, however, that possibility remains within the realm of medical speculation rather than reality.

Therapeutic Options
It is against this background that Kummar and associates present a well-written, concise review of one of the more common small bowel malignancies, small bowel adenocarcinoma. The authors have included in their review adenocarcinoma of the duodenum, a site that accounts for over half of all small bowel adenocarcinomas. These tumors are often included in series reporting peripancreatic malignancy, although it appears clear that their biology is different, ie, consistent with other enteric tumors. Surgical resection—with either segmental duodenectomy or pancreaticoduodenectomy (depending on anatomic considerations)—is appropriate. Local resection is inadequate as it is associated with a high rate of subsequent local recurrence. As there is significant long-term survival in the setting of node-positive disease, regional lymphadenectomy in patients with this type of adenocarcinoma may, in fact, be therapeutic in some patients. As the authors have clearly stated, the value of therapy beyond complete surgical resection, acknowledging the possible therapeutic value of regional lymphadenectomy, remains unknown. Adjuvant radiation therapy to the fixed regional nodal bed of duodenal adenocarcinoma may be
reasonable in selected high-risk patients, though not without some locoregional toxicity. Radiation therapy to the mobile mesentery of the distal small bowel makes less sense intuitively. With regard to systemic chemotherapy, its role in either the neoadjuvant or postoperative adjuvant setting is unproven. Most centers would use data extrapolated from patients with colon cancer to offer systemic adjuvant therapy to those with completely resected node-positive small bowel adenocarcinoma.

**Metastatic Disease**

For patients with established metastatic disease, again, regimens that have been applied to other metastatic GI malignancies have shown modest activity without a clearly demonstrable impact on either length or quality of life. In addition to fluorouracil (5-FU), other agents that may have value in the management of these patients include irinotecan (CPT-11, Camptosar) and oxaliplatin (Eloxatin). Ultimately, however, based on past experience with systemic therapy for advanced metastatic GI adenocarcinoma, major advances with this strategy are unlikely. Meaningful progress is more likely to come from defining a unique genetic vulnerability in these tumors, such as has been seen in the application of imatinib (Gleevec) to patients with advanced metastatic GI stromal tumors.[3]

With regard to palliation, although surgery may be useful in addressing specific symptoms of obstruction in the primary tumor in patients with metastatic disease, the role of surgical palliation of patients with obstruction from recurrent disease should be carefully evaluated on an individual basis. The authors cite several small retrospective studies that suggest a survival advantage seen in patients who receive treatment, as opposed to those who were followed with supportive care. It is possible, indeed likely, that any observed survival difference in these small studies is due to patient selection (ie, that untreated patients had more advanced disease), rather than the impact of treatment per se. We must be cautious about interpreting these results as being due to any treatment-related effect.

**Conclusions**

The authors are to be commended for compiling a very concise review of small bowel adenocarcinoma. They have reminded us that treatment for unusual tumors is much more often guided by general principles of oncology than by specific class 1 data.

**References:**


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