Small Bowel Tumors

Shirley C. Paski, MSc, MD, Carol E. Semrad, MD*

KEYWORDS

• Small bowel tumor • Enteroscopy • Capsule endoscopy

Small bowel tumors are rare, accounting for only 2% of all primary gastrointestinal tumors.[1] Several theories to account for the scarcity of small bowel tumors have been hypothesized, including rapid intestinal transit time, dilution of carcinogens in chyme, reduced bacterial load limiting conversion of bile acids to carcinogens, rapid turnover of the small intestinal epithelium, conversion of dietary benzpyrene by benzpyrene hydroxylase into less toxic moieties, and protection by high levels of secretory IgA.[1,2] Benign small bowel tumors are often asymptomatic and cured at the time of resection. In contrast, malignant primary small bowel tumors have a poor prognosis and have often metastasized by the time they are discovered.[3] Videocapsule endoscopy (VCE) and new radiologic imaging techniques have greatly facilitated detection of small bowel tumors. Double-balloon enteroscopy (DBE) and newer overtube-assisted enteroscopes can detect small bowel tumors missed at VCE and allow for tissue sampling, tumor marking, and endoscopic resection when possible.

This article describes the general features of small bowel tumors, clinical presentation, and diagnostic tests followed by a description of the more common tumor types and their management.

EPIDEMIOLOGY

Epidemiologic studies on small bowel tumors are limited, in part, due to their low incidence. Benign small bowel tumors are usually asymptomatic, which makes the precise incidence difficult to determine. Older studies including 22,810 and 2648 autopsies found 35 and 22 benign small bowel tumors, respectively, for an incidence of 0.15% to 0.83%.[4,5] Amongst patients with symptomatic benign small bowel tumors, the most common types are leiomyomas, lipomas, adenomas, and angiomas.[6] In autopsy series, benign tumors are far more common than malignant tumors, accounting for up to 75% of all small bowel tumors.[7] However, the latter are more commonly reported in the medical literature.

The global incidence of primary small bowel cancers is highly variable. Incidence rates are highest in North America and Western Europe, particularly amongst African American men. Incidence rates are lowest in Asia and the Middle East. In most

Section of Gastroenterology, Department of Internal Medicine, University of Chicago Medical Center, 5841 S Maryland Avenue, MC 4076, Chicago, IL 60637, USA
* Corresponding author.
E-mail address: csemrad@medicine.bsd.uchicago.edu (C.E. Semrad).

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countries, men have a slightly higher incidence compared with women.[2] Incidence increases with age, the mid-60s being the median age of diagnosis. Carcinoids (44.3%) and adenocarcinoma (32.6%) are the most common primary cancers of the small bowel, followed by lymphomas (14.8%) and sarcomas (8.3%).[3] Adenocarcinomas most commonly occur in the duodenum or proximal jejunum, carcinoids and lymphomas in the ileum. Recent studies show that the incidence of small bowel cancer is increasing in the United States. Using the National Cancer Institute's Surveillance Epidemiology and End Results (SEER) 9 Incidence database, Bilimoria and colleagues[3] found that the average annual age-adjusted incidence of small bowel cancer nearly doubled from 11.8 cases per million in 1973 to 22.7 cases per million in 2004. The greatest increase was seen in carcinoids, followed by lymphomas and adenocarcinomas. Gastrointestinal stromal tumor incidence has remained steady. Despite these increases, primary small bowel malignancies remain rare with only 6110 new cases and 1110 deaths in the United States in 2008.[8]

Metastatic cancers to the small bowel are more common than primary cancers. They occur by direct invasion from adjacent organs or by distant metastasis.[9]

RISK FACTORS

Several medical and genetic conditions are associated with an increased risk for developing small bowel cancer, including celiac disease, Crohn disease, polyposis syndromes, and hereditary nonpolyposis colorectal cancer. Dietary factors have been associated with small bowel tumors, although study results are inconsistent.

Inflammatory diseases of the small bowel are associated with an increased risk of malignancy. Celiac disease is associated with increased risk for developing small bowel lymphoma and adenocarcinoma. The incidence of malignancy has been reported to be highest within the first 3 to 4 years of diagnosis and in those with poor adherence to a gluten-free diet.[10] Early diagnosis and strict adherence to a gluten-free diet may lessen the risk of small bowel malignancy in celiac disease.[11,12] Crohn disease is associated with an increased risk of developing adenocarcinoma and, to a lesser extent, lymphoma of the small bowel. Increased risk is associated with male gender, extended duration of disease, location in the small bowel, strictures, and fistulas.[13]

Polyposis syndromes and hereditary nonpolyposis colorectal cancer (HNPCC) have been associated with an increased risk for small bowel cancer. New enteroscopy technologies that allow examination of the entire small bowel and therapeutics have the potential for improved surveillance and therapy.[14,15]

Studies exploring the relationship between diet and environmental factors and small bowel cancer have yielded mixed results. Several case-control studies have found a positive association between the development of small bowel cancer and increased consumption of red and processed meat,[16,17] sugar intake,[16] smoking, and alcohol.[16] Others found no relationship between smoking and alcohol consumption.[17,18] Prospective cohort studies found no relationship between red or processed meat and small bowel tumors, but did find a positive correlation between saturated fat and carcinoid,[19] and a negative correlation between whole grain fiber and small bowel cancer.[20] Further studies are needed to clarify the role of diet, if any, in small bowel cancer.

SYMPTOMS AND SIGNS

The diagnosis of small bowel tumors is often delayed because most are asymptomatic or have nonspecific symptoms during the early stages of development. Clinical