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Primary Intestinal Malignant Melanoma

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The patient, a 58-year-old woman underwent an exploratory laparotomy for an obstructing acute intussusception in July 1992. She was found upon exploration to have a 3. 0 by 2. 0 cm tumor in the distal jejunum causing partial obstruction of the proximal small bowel and resection of the jejunum was done with endto-end anastomosis. On the fixed specimen, the tumor was 3.5 cm in diameter with a 3.0 cm proximal and 6. 0 cm distal margin. Frozen and permanent sections showed malignant melanoma (Fig. 1,2). The mesenteric lymph nodes revealed reactive lymphadenitis without evidence of metastases. No primary lesion was found on repeat thorough physical examination. Postoperatively, the patient underwent fundoscopic examination, colposcopy, and flexible sigmoidoscopy to 65 cm, but no other primary site could be found. The patient's recovery was uneventful, with no evidence of infection. She was discharged 10 days postoperatively. Her final diagnosis was primary malignant melanoma of the small intestine. After discussing all of the possible standard and investigational systemic adjuvant therapies for malignant melanoma, the patient declined further treatment. Repeated clinical examinations, serum immunoglobulins, chest radiograph, bone scan, and computed tomography (CT) scans of the abdomen and brain were all normal. A laparatomy was performed in May 2000 because of an incisional hernia. There was no sign of the disease in her abdomen. She is still without any evidence of the illness 9 years after diagnosis.

Most cases of malignant melanoma of the GI tract are secondary to metastases (1). According to Reemst,

less than 5% of the patients present with symptoms that are often nonspecific and late. The most common symptoms are acute gastrointestinal bleeding and anemia (2). The areas of the GI tract that can be involved and their frequencies are as follows: small bowel (58-71%), stomach (20-27%), colon (22%), esophagus (5%), and rectum (2%) (3). There have been very rare case reports of GI tract malignant melanomas without any other known primary site. In Elsayed's retrospective study, he reviewed 103 cases of malignant melanoma in the small intestine (77 surgical resections, 26 autopsies) accessioned at the Pathology Department between 1945 and 1991. In 46% of cases (47 patient), there was no record of a primary lesion preceding the intestinal tumor and 23% of these were autopsies (4). Ollila reported that GI tract involvement was diagnosed premortem in only 4% of patients (5). In approximately 60% of patients who die from melanoma, the autopsy reveals metastases in the GI tract (6). Because the small and large intestines normally contain no melanoblasts, some authors believe that primary malignant melanomas of the GI tract are unlikely (7). However, melanocytes have been found in the alimentary tract as well as the respiratory tract even in lymph nodes. Another hypothesis supporting a primary etiology proposes that malignant melanomas can originate from neural crest cells (8). These multipotential cells become amine precursor uptake and decarboxylation (APUD) cells. APUD cells can undergo neoplastic transformation and produce tumors such as carcinoid, gastrinoma, and medullary thyroid carcinoma. Melanomas may originate from APUD cells, although



Figure 1. The focal mucosal infiltration of the tumor (x100, H&E).

unlike the usual 'APUDomas', malignant melanomas are not hormone producers.

Systemic adjuvant therapy for metastatic malignant melanoma is limited. There is no standard, effective chemotherapy after either palliative or curative resection of malignant melanoma of the GI tract. Investigational adjuvant therapy protocols include alpha interferon, interleukin, various chemotherapeutic drug combinations, active immunotherapy by vaccination with modified melanoma cells, and high-dose chemotherapy with



Figure 2. Intrastoplasmic melanin pigmentation of the tumor. Epitheloid and spindle cell formation is also seen (x400, H&E).

autologous bone marrow transplants (9). At present, the prognosis for patients with untreated metastatic malignant melanoma remains poor. However, as demonstrated by our case and other reports, primary malignant melanoma of the GI tract may exist, and longterm survival may be accomplished with close follow-up and prompt surgical treatment.

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