Primary Small-Bowel Melanoma

Color Doppler Ultrasonographic, Computed Tomographic, and Radiologic Findings With Pathologic Correlations

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Malignant melanoma appears in typical sites where melanocytes can usually be found (skin, eyes, meninges, and anal region).[1] In the gastrointestinal (GI) tract, melanoma can be also found in the rectum and sigmoid colon by the local migration of primordial skin melanocytes.[2] Some other rare sites of primary melanomas in the GI tract were described as the gallbladder, stomach, small and large intestine, mouth, tongue, and esophagus.[2–8] Generally, most GI melanomas are metastases from a skin tumor.[9,10] We report the case of a patient with a first presentation of small-bowel melanoma, which was considered a primary site because of the absence of concurrent lesions and no history of removal of a melanoma or atypical melanocytic lesion from the skin or other organs.[11]

Abbreviations
CD-US, color Doppler ultrasonography; CT, computed tomography; GI, gastrointestinal

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Case Report

A 49-year-old woman had a 6-months history of weakness, nausea, mild epigastric pain, chronic anemia, and loss of weight. She denied rectal or other bleeding sites and other previous important diseases in her anamnesis. On admission, clinical examination showed moderate obesity, a normal heart and chest, a nondistended abdomen with tenderness to palpation, and normal bowel sounds in all quadrants. A finger examination excluded hemorrhoids and showed nonbloody feces in a rectal ampulla. Blood tests confirmed sideropenic anemia (blood hemoglobin level, 8.2 g/dL). Liver and renal function test results were normal. All tested tumor markers (carcinoembryonic antigen, cancer antigen 19-9, cancer antigen 125, α-fetoprotein, neuron-specific enolase, and tissue-type plasminogen activator) were within normal ranges. Chest radiographic findings were normal. Twenty-four hours after admission, the patient underwent gastroscopy that showed a normal esophagus, stomach, and duodenum.

Abdominal color Doppler ultrasonography (CD-US), performed on the same day (3.5-MHz probe; Prosound SSD-5500 SV, Aloka Co, Ltd, Tokyo, Japan), showed a hypervascular, hypoechoic mass (diameter, 5 cm) in the left upper abdominal quadrant (Figure 1, A–C) with intralesional arterial and venous vessels at spectral Doppler ultrasonographic examination. Color Doppler ultrasonography also detected a single satellite enlarged mesenteric lymph node (diameter, 19 mm) showing an "atypical hypoechoic, rounded shape" (Figure 2A), absence of hilar echoes, and peripheral irregular atypical vascularization (Figure 2B). The diagnosis of carcinoma of the transverse colon with lymph nodal metastasis was suggested. However, a full colonoscopy failed to show any large-bowel lesion.

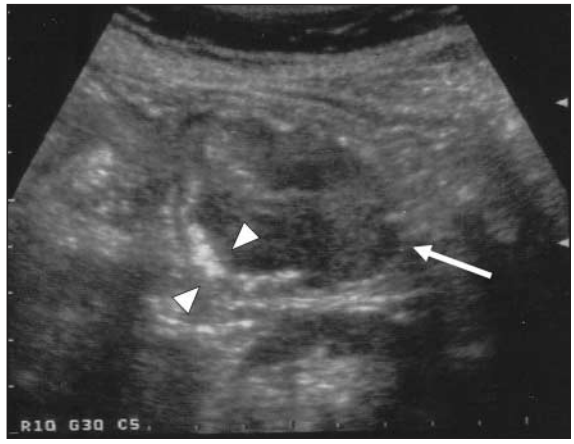
Abdominal enhanced single-slice computed tomography (CT) (Atom Fast Ring, Hitachi Medical Systems, Tokyo, Japan; Gastrografin [sodium amidotrizoate and meglumine amidotrizoate] oral contrast medium, Schering AG, Berlin, Germany; and Iomeron 400 [iomeprol] intravenous contrast medium, Bracco SpA, Milan, Italy) showed an inhomogeneous, well-defined jejunal mass with central contrast enhancement in the venous phase (Figure 3) and failed to detect enlarged lymph nodes. The diagnosis of lymphoma or leiomyoma was suggested.

A single-contrast GI radiologic examination (Prontobario [barium] oral contrast medium, Bracco SpA) with small-bowel follow-through showed stretched and distended loops proximal to a narrowed lumen caused by an intramural mass in the upper jejunum (Figure 4). Although the mass was rather large, obstruction was not present because the barium progression seemed normal. The distal jejunum and ileum were normal.

The patient underwent surgery 1 week after admission. Macroscopically, at laparotomy, a jejunal polypoid mass was found (Figure 5A). The jejunal loop including the tumor and 3 enlarged mesenteric lymph nodes were resected. The open surgical specimen showed a polypoid mass (diameter, 5 cm) 6 cm apart from the resection margin (Figure 5B). Histologic examination of the specimen yielded a finding of epithelioid cells with irregular nuclei and eosinophilic cytoplasm containing dark brown pigment and infiltrating lymphatic vessels (Figure 6A). At immunochemistry, marked positivity for S-100 and focal positivity for HMB-45 antigens[12,13] were present

Figure 1. A, Gray scale sonogram showing a hypoechoic mass (arrow) in the left upper abdominal quadrant almost completely surrounded by a distended bowel wall (arrowheads). **B** and **C**, Color Doppler (**B**) and power Doppler (**C**) sonograms showing numerous chaotic and large intralesional vessels.

A



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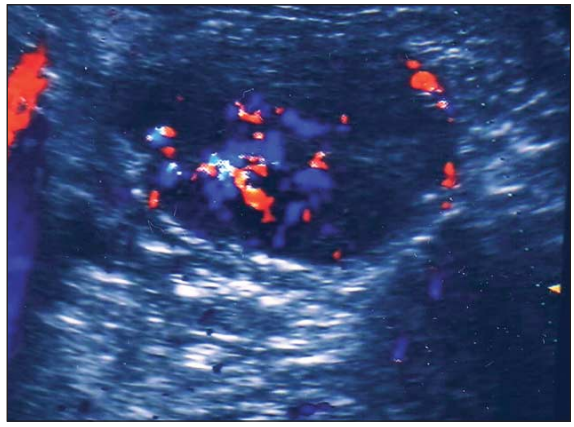

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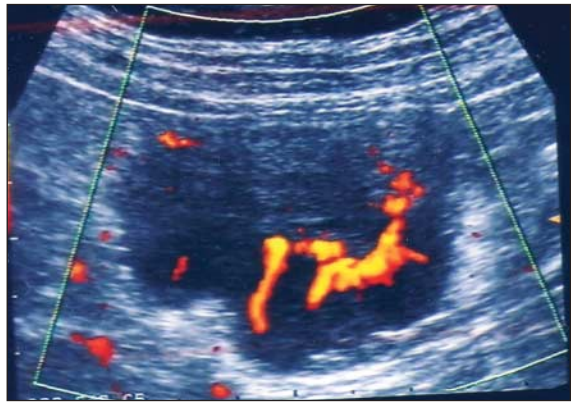
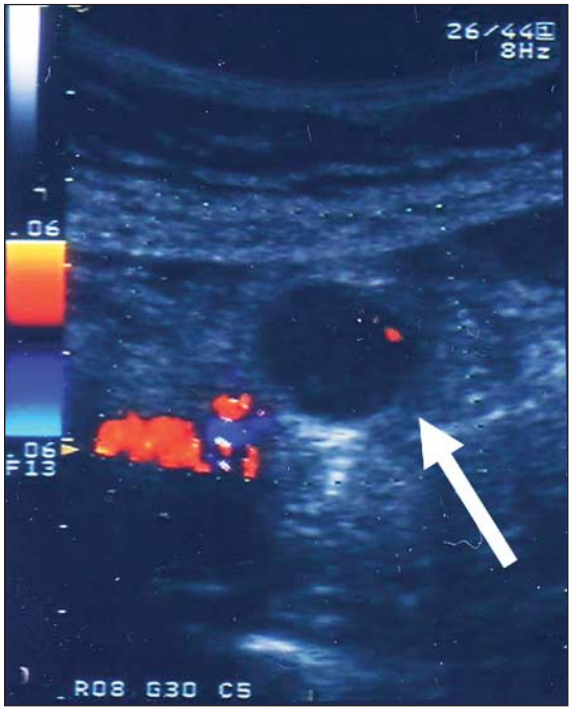


Figure 2. Color Doppler sonograms showing a single enlarged lymph node (arrow) with an atypical hypoechoic, rounded shape (**A**) and peripheral atypical vascularization (**B**). Hyperechoic hilar echoes are absent in both **A** and **B**.

A



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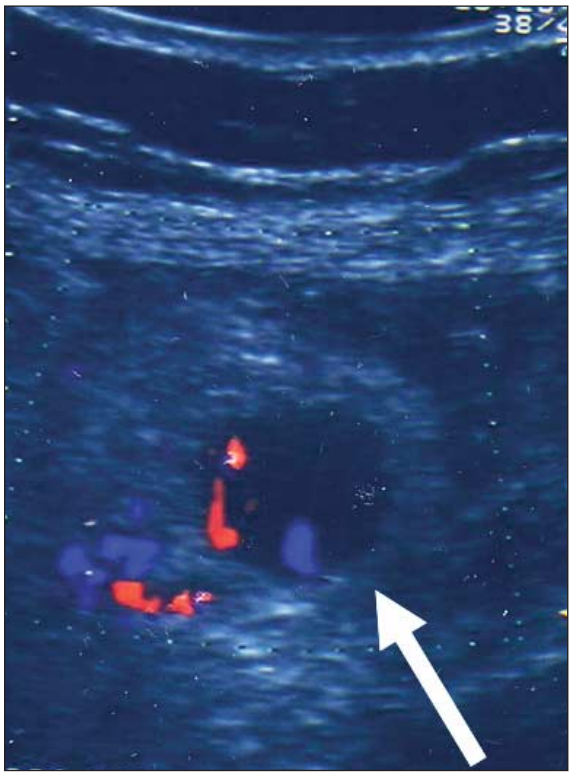


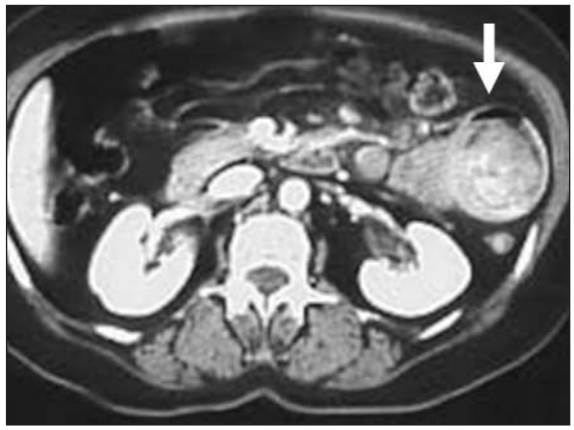
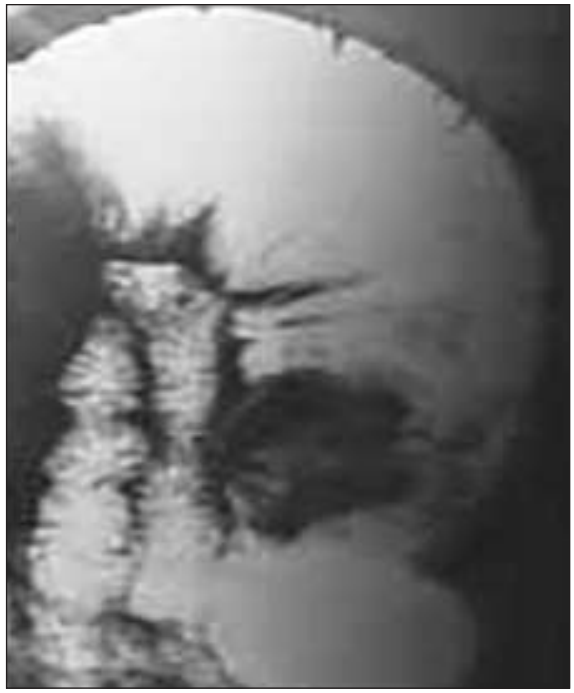


Figure 3. Enhanced CT scan clearly showing the mass (arrow) and correctly defining its origin from the small bowel.

(Figure 6B). Of the 3 resected lymph nodes, 1 was positive for malignancy. The conclusion was melanoma of the small bowel with lymph nodal metastasis. The anamnesis of the patient included no cutaneous or mucosal surgery. Clinical inspection failed to show any suspect cutaneous or mucosal lesions. No suspect skin lesions were detected at 3-, 6-, and 12-month follow-ups. The patient was still alive after a 15-month follow-up without evidence of recurrent disease.

Figure 4. Contrast-enhanced small-bowel radiograph showing a polypoid mass in the upper jejunum.



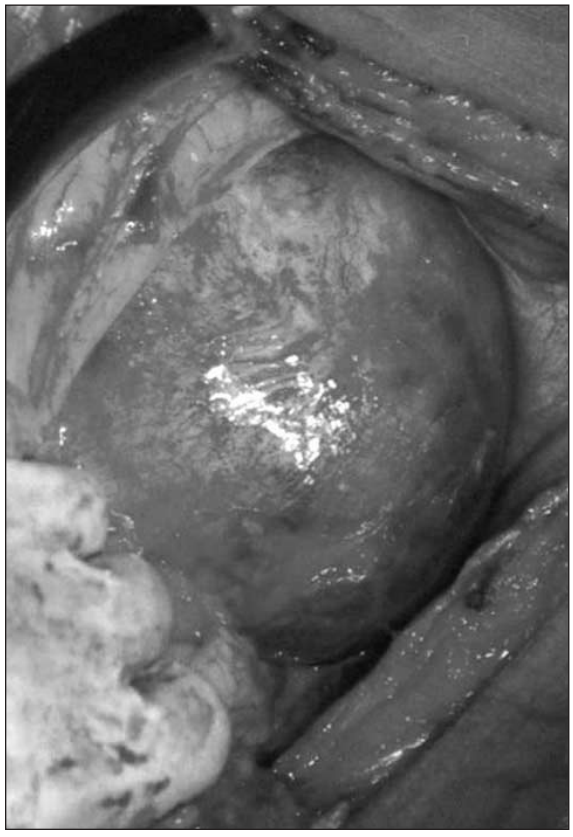
Discussion

Primary malignant tumors of the small bowel account for less than 5% of GI malignancy.[14,15] The most common malignancies involving the small bowel are metastatic.[16,17] Small and large intestines are the most common sites for metastases from cutaneous malignant melanoma. Sixty percent of patients with malignant melanoma had GI metastases in an autopsy series.[18] Primary small-bowel melanomas are exceedingly rare[19] and are associated with a worse prognosis in comparison with cutaneous tumors[10] because of the rich vascular supply of GI mucosa and because detection of the disease is frequently made only at an advanced clinical stage.

The clinical presentation of small-bowel melanoma is similar to those of other small-bowel tumors and includes pain (70% of cases) from intestinal occlusion, anemia (20%–50%) from chronic blood loss, loss of weight (50%), and the presence of an abdominal palpable mass (25%).[2,9,20] Invagination, massive rectorrhagia, and perforation are less frequent findings.[2,20]

Figure 5. A, Tumor appearance at laparotomy. **B**, Resected open specimen showing a polypoid mass (diameter, 5 cm) 6 cm apart from the resection margin.

A



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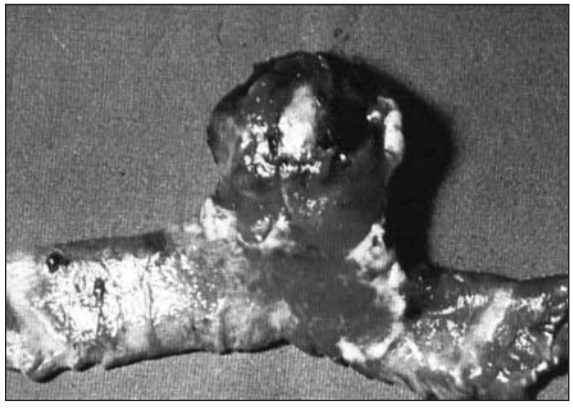
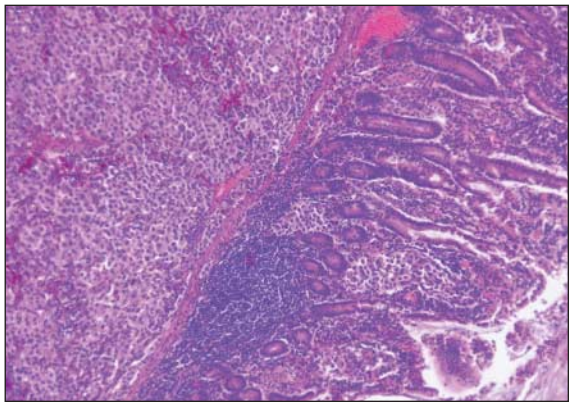


Figure 6. A, Hematoxylin-eosin staining showing wide tumoral infiltration of submucosa with malignant cell clusters in the tunica propria. **B**, Melanoma-specific immunochemical staining showing marked positivity for S-100 antigen.

A



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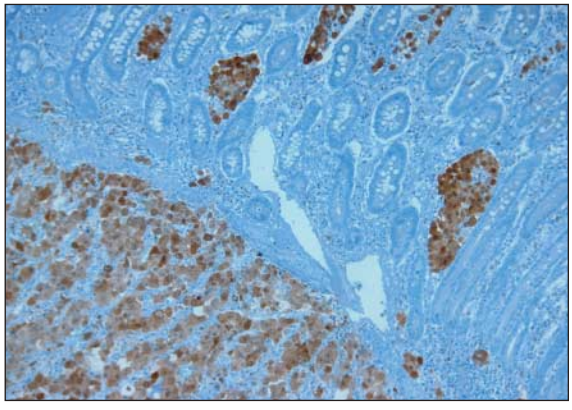

The detection and specific diagnosis of small-bowel tumors is challenging because of limited evaluation of the small bowel with standard endoscopy. Barium examinations (small-bowel follow-through and enteroclysis) and CT are usually the standard techniques for detecting small-bowel tumors.[16–21] Although barium examinations can be very sensitive for intraluminal studies, they do not give any information about extraintestinal findings such as enlarged metastatic lymph nodes.

Bowel wall lesions and extraintestinal alteration can both be studied with CT. The thinner collimation now available with multidetector CT scanners and the 3-dimensional imaging capabilities of CT improved detection of small-bowel tumors and normal and enlarged mesenteric lymph nodes.[21,22] However, the differential diagnosis between metastatic and "reactive" enlarged lymph nodes at CT is difficult, and only lymph node size has been reported as a reliable sign for this purpose.[22,23]

In patients with vague abdominal symptoms, transabdominal ultrasonography is frequently chosen as a first screening method because it is noninvasive, does not require special preparation, and is readily accessible and inexpensive. Fluid collections, lymph node enlargement, and bowel wall thickening are readily diagnosed with high sensitivity by ultrasonography. For these reasons, CD-US could be appropriately considered in the diagnostic workup of small-bowel tumors.[24–27]

Approximately two thirds of small-bowel tumors are malignant. They are located more frequently in the jejunum than in the ileum. More than 95% of these are adenocarcinomas, carcinoids, lymphomas, and gastrointestinal stromal tumors.[2,14,15,20,21]

At ultrasonography, jejunal lymphoma shows a typical pattern in more than 70% of cases.[25,26] In fact, it classically presents transmural circumferential, profoundly hypoechoic wall thickening up to 4 cm in diameter,[25,26] with loss of normal stratification. On the contrary, other neoplasms of the small bowel (eg, adenocarcinoma, gastrointestinal stromal tumors, carcinoids, and also melanoma) may show many different patterns, such as an inhomogeneous hypoechoic mass, irregular and asymmetrical hypoechoic bowel thickening, and an intraluminal vegetant solid hypoechoic mass.[16,25]

Therefore, whereas a typical ultrasonographic pattern favors the diagnosis of a non-Hodgkin lymphoma, ultrasonography seems poorly specific in the differential diagnosis between the other common small-bowel tumors. However, even in an asymptomatic patient, when one of the above-mentioned patterns is detected on ultrasonography in the small bowel, other radiologic studies and surgery are mandatory. In these cases, the presence of atypical mesenteric enlarged lymph nodes strengthens the indication for further diagnostic and therapeutic workup. Color Doppler ultrasonography showed high sensitivity and specificity for the detection and characterization of enlarged lymph nodes. In fact, abdominal CD-US, performed with graded compression and with 5- to 10-MHz probes, allows high definition of the intranodal structure and high sensitivity in depicting nodal vascularization.[28–31] Unlike CT, which considers only lymph node size, CD-US characterization of enlarged malignant lymph nodes is also based on the shape, the presence of an echogenic hilum, and the vascularization. Nodal size alone has been shown to be an unreliable criterion for differentiating reactive from malignant lymph nodes.[21,22] On the basis of the color flow pattern alone, sensitivity and specificity for diagnosis of malignant lymph nodes of 91% and 63%, respectively, have been reported.[30] In our patient, the single malignant lymph node showed a rounded atypical shape, absence of an echogenic hilum, and peripheral and mixed vascularity consistent with aberrant neovessels. The other lymph nodes detected on ultrasonography showed a normal oval shape and absence of intralesional color Doppler signals. Histologic examination confirmed the benign nature of these latter lymph nodes.

Although a cutaneous primary tumor may not be found in up to 46% of patients with melanoma of the GI tract, GI involvement is generally assumed to be metastatic.[32,33] Criteria for the diagnosis of primary melanoma include absence of other primary-site melanoma and no history of removal of melanoma or atypical melanocytic lesions from the skin or other organs.[11,16] Nevertheless, spontaneous primary

tumor regression is described in about 15% of skin melanomas.[17] In our case, there was no history of primary cutaneous or mucosal melanoma, and a 15-month follow-up failed to show any other possible primary site. The tumor was detected by CD-US as a first-approach imaging technique. Color Doppler ultrasonography also showed hypervascularity of the lesion and, moreover, allowed detection and characterization of the only mesenteric lymph node infiltrated from the malignancy. At surgery, the surgeon was unable to reliably establish the number of metastatic lymph nodes. The definitive diagnosis was disclosed by pathologic examination, and in our case, there was agreement with CD-US. However, although very sensitive, CD-US failed to clearly show which intestinal tract was involved. The diagnostic mistake of CD-US entailed a useless colonoscopy showing a normal colon. Both CT and contrast-enhanced small-bowel radiography correctly defined the intestinal site involved from the malignancy. However, both radiography and CT failed to show the single metastatic lymph node.

Percutaneous ultrasonographically guided fine-needle aspiration can be performed in patients not scheduled for emergent surgery.[27] In our case, the diagnosis of small-bowel malignancy was already sufficiently obvious on the basis of the CD-US signs, so there was no need for biopsy before surgery.

In conclusion, primary jejunal melanoma was a reliable diagnosis in our patient. Color Doppler ultrasonography, as first imaging technique, showed the bowel lesion and its hypervascularity and detected the single metastatic mesenteric lymph node. Computed tomography and radiography correctly identified the site where the tumor originated. Inclusion of CD-US among the imaging techniques performed in the pretreatment study of small-bowel masses could be useful for an early noninvasive diagnosis and complete and accurate planning of surgical treatment.

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