

Case Report

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Synchronous Primary Small Bowel Melanomas: A Case Report and Literature Review

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Abstract

Background: Malignant melanoma (MM) is an epithelial malignancy originating from melanocytes. Approximately 1%–3% of all gastrointestinal cancers are MM. The majority of these malignancies are metastases or secondary tumors.

Case Report: A 52-year-old male was admitted to our hospital due to abdominal pain, nausea, loss of appetite, weight loss, and chronic constipation. An abdominal CT scan showed a mass of 76×173 mm in the left colon. During surgery, two distinct solid masses were seen in the jejunum, 25 cm and 10 cm in diameter, located about 30 cm and 50 cm after the Treitz ligament, respectively. A segmental enterectomy was performed. Pathology examination of the resected tumor identified MM.

Discussion: Primary intestinal melanomas require follow-up due to their possibility of recurrence, invasive course, and poor prognosis.

Conclusion: Early identification and resection surgery can improve quality of life and prolong the symptom-free survival time of MM. **Keywords:** Small intestine, Malignant melanoma, Primary gastrointestinal melanoma, Case report

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Introduction

Malignant melanoma (MM) is an epithelial malignancy originating from melanocytes, which can be found in a variety of tissues, including the eye, oral cavity, nasopharynx, anus, urinary system, and vagina. However, the skin is where they are mainly located. MM accounts for roughly 1%–3% of all gastrointestinal (GI) malignancies. Most of these tumors are secondary or metastatic lesions from a primary tumor (1,2).

Breast and lung cancers are the next most common cancers to spread to the GI system after MM. MM spreads in the ileum and jejunum, two regions of the small intestine. The most common way melanoma spreads to the GI system is through the limbs, trunk, head, and neck. Anywhere from 2 to 180 months can pass between the diagnosis of the original MM and the discovery of GI metastasis (1).

There have only been a few documented instances of small intestinal primary melanoma. We provide a rare case of synchronous primary small intestinal melanoma, demonstrating the difficulties in diagnosing the disease and the necessity of early surgical treatment, which can improve quality of life and increase survival rates. Our work has been documented in accordance with the CARE guidelines.

Case Presentation

A 52-year-old male was admitted to our hospital due to abdominal pain, nausea, loss of appetite, weight loss, and chronic constipation. Abdominal pain had started four months earlier in the periumbilical and left lower quadrant, was worsened by eating, and was decreased by a bowel movement. He had no history of specific diseases and was not taking any medication. Vital signs were as follows: blood pressure 90/60 mm Hg, pulse rate 86/min, respiratory rate 16/minute, temperature 36.6 °C. He was generally ill and cachectic. On physical examination, there was a generalized abdominal tenderness and a mass-like lesion around his umbilical area. No hepatosplenomegaly was detected. In his extremities, he had pitting edema that was 4+on the feet and 2 in the hands. Initial laboratory test results were white blood cells = $11800/\text{mm}^3$, hemoglobin = 9 g/dl, neutrophil=68%, lymphocyte=24%, platelets=574000/ μ l, albumin = 2.8 g/dL, erythrocyte sedimentation rate = 28 mm/h, and C-reactive protein = 166 mg/L. HBV and HCV serology were negative, and liver and kidney function tests were normal. The liver, biliary ducts, pancreas, and kidneys were normal in abdominal sonography. A 150×95 mm heterogeneous mass was identified close to the colonic splenic flexure, pancreas, and stomach. An abdominal CT



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scan showed a mass of 173×76 mm in the left colon, which narrowed the lumen (Figure 1). The patient underwent a normal colonoscopy.

The patient underwent explorative laparotomy for further evaluation. During surgery, two distinct solid masses with adhesions to the left colon and surrounding tissues were seen in the jejunum, 25 cm and 10 cm in diameter. They were located about 30 and 50 cm after the Treitz ligament, respectively (Figure 2). Two ends of the small intestine were anastomosed together after a lymphadenectomy, and a segmental enterectomy of the jejunal loops with sufficient surgical margins was completed. There was no other pathology in the peritoneal cavity. The patient was admitted to the ICU overnight, and seven days later, he was discharged from the hospital in good general condition.

Pathology examination of the resected tumor identified heavily infiltrated atypical pleomorphic cells with large vesicular nuclei, prominent red nucleoli, ample eosinophilic cytoplasm, multinucleated tumoral cells, and regions of necrosis and ulceration. Although the immunohistochemical stain of the tumor cells was positive for S100 and HMB45, it was negative for CD20, CD138, CD79a, and CK, indicating MM.

The patient underwent an ophthalmological and dermatological examination, which produced normal results. No lesions indicating melanoma were detected. More than a year has passed since the surgery. The patient is under the supervision of an oncologist and has not had any new complications so far.

Discussion

Primary intestinal melanomas are considered to derive from neural crest melanoblast cells that relocate to the distal ileum via the omphalomesenteric channel. Some researchers believe primary melanomas can arise from enteric neuroendocrine tissue of APUD cells that undergo neoplastic transformation or from neuroblastic Schwann cells in the intestinal autonomous nervous system (1,3).

A primary small bowel MM diagnosis could be made based on the following criteria: 1) a single melanoma lesion verified by histology, 2) no other organ involvement except regional lymph nodes, and 3) disease-free survival of at least 12 months after diagnosis (3). The nasopharynx, esophagus, stomach, small bowel, gallbladder, colon, rectum, and anal tract may be affected by the primary MM (2). Primary melanomas that are identified late are more aggressive and have a worse prognosis than secondary melanomas. Differentiating between original melanoma and metastatic MM with an unknown or regressed cutaneous origin might be difficult. Abdominal pain, dyspepsia, weight loss, nausea, vomiting, obstruction, perforation, acute GI bleeding, and chronic iron deficiency anemia are among the symptoms that melanoma lesions of the GI tract can cause (1-3).

CT enteroclysis has improved the rate of intestinal MM



Figure 1. Abdominal CT scan: A 76×173 mm mass in the left colon, which caused narrowing of the lumen



Figure 2. Two distinct masses 25 cm and 10 cm in diameter in the jejunum, about 30 and 50 cm after the Treitz ligament, respectively

identification. However, CT imaging has only a 60–70% sensitivity for detecting intestine MM metastases. Wholebody PET CT has greater sensitivity and specificity for all GI metastatic MM than CT scans. When gastroscopy and colonoscopy fail to discover further melanoma metastases, video capsule endoscopy (VCE) may be utilized as the first endoscopic modality (1).

Badakhshi et al analyzed significant clinicopathological features among melanomas in different GI regions. They identified independent prognostic features for patients with primary GI melanoma (PGIM), creating a prognosisrelated prediction model. Men and women comprise 43.3 % and 56.6 % of PGIM patients. The median age is 71 at the time of diagnosis. The rectum, esophagus, small bowel, and large intestine are the next most common main sites of PGIM, after the anus (50%). The median diameter of the tumor is 3.9 cm. Regarding stage, 33.0% of patients are in the localized stage, 24.0% in the regional stage, and 32.2% have distant metastases at diagnosis. Regarding the type of treatment, 54.2% have surgery alone, 21.4% have surgery with (neo)adjuvant therapy, 5.5% receive nonsurgical therapy, and 18.9% of patients do not receive any type of therapy. Overall survival is affected by age, which is an independent prognostic factor. The survival rates of older patients are lower than those of younger patients. The anatomic site is an independent significant predictor of survival. Despite malignancies arising in the small intestine, large intestine, and upper GI tract, tumors originating from the anus and rectum are often associated with improved survival. The median overall survival periods for primary esophageal and gastric melanoma range from 8.0 months to 18.0 months for anus melanoma. Patients with only surgery have the best survival rates, followed by surgery with (neo)adjuvant and non-surgical treatment (4).

Until a decade ago, individuals with MM who had metastatic GI tract involvement had a poor prognosis, and surgical choices were the only way to improve survival and symptoms. Multiple studies have shown that total surgical resection of GI metastatic MM not only relieves symptoms but also leads to a better prognosis and longer overall survival. Given the result of efficient systemic treatment, chemotherapy is an established treatment with a long history. The conventional treatment used to be dacarbazine, which had a low response rate of 5%–28% (Average 15%). Ipilimumab was the first medication to show a significant improvement in the overall survival of patients with metastatic melanoma. Immunotherapy can be used alone or in combination with other treatments (1).

Crișan et al reported a 39-year-old lady with nausea, abdominal discomfort, bowel changes, and weight loss. A small intestinal mass was discovered on a CT scan. A 6 cm ileal tumor was surgically removed, and regional mesenteric lymphadenectomy was performed. Ileal melanoma metastases were identified histopathologically. A cutaneous lesion on the right forearm was discovered during a subsequent dermatological examination. However, no malignant cells were revealed in the histological analysis. Multiple frontal and parietal lesions were detected in a whole-body PET CT scan. Neurosurgical operations were performed on the patient to remove cerebral metastases. External radiation and chemotherapy were also used as palliative treatments. The patient died after repeated episodes of cerebral hypertension about two years after being diagnosed (3). Our patient was older, and his tumor was in the jejunum. In addition, no skin melanoma was detected in our patient, and there was no brain metastasis.

Ait Idir reported a 75-year-old lady with pelvic pain that had persisted for four months and was unresponsive to analgesics. Melena and asthenia exacerbated the symptoms. A palpable and movable pelvic tumor was identified during an abdominal examination. In the CT scan, the distal small bowel, the bladder dome, and the uterine body were all involved in a massive 119cm pelvic mass. The patient underwent a minor bowel resection with 10 cm margins following an exploratory laparotomy. MM was discovered in the histopathological test. Anoscopy, eye and skin examinations, and several cutaneous biopsies showed no melanoma lesion, and an etiological examination for a primary tumor of the small intestine melanoma came out negative (5). Our patient was younger and presented with an abdominal mass and pain. Similarly, our patient did not have a skin lesion.

Compared to other melanoma subtypes, intestinal melanoma is more aggressive and has a worse prognosis in both its original and metastatic forms. Most of the time, the diagnosis is made after complications such as intussusception, bleeding, obstruction, and perforation. Examinations are not reliable enough to provide a conclusive diagnosis due to non-specific symptoms at the onset of the disease. In most cases, the ultimate diagnosis is determined through surgical intervention. A precise diagnosis of small bowel metastases can be made after a laparotomy or exploratory laparoscopy and resection of the intestinal mass. Palliative tumor resection can be performed in advanced situations (where curative surgery is not possible), resulting in symptom relief and a reduction in the risk of further complications. Chemotherapy and immunotherapy are mainly used to treat metastasis (2).

Conclusion

Primary intestinal melanomas have an invasive course and a poor prognosis, necessitating follow-up because recurrence is possible. Early identification and resection can improve the quality of life and prolong the symptomfree survival time of MM.

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Competing Interests

The authors declare no conflict of interest.

Ethical Approval

The patient's written consent was obtained for the publication of this article.

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