J Gastrointest Canc (2019) 50:353–356
https://doi.org/10.1007/s12029-017-0031-x

CASE REPORT

# Disappearing Lump—an Unusual Presentation of Large Metastatic Small Bowel Malignant Melanoma

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Published online: 17 November 2017


## Introduction

Malignant melanoma is one of the most common malignancies metastasizing to the gastrointestinal tract. It occurs both at the time of primary diagnosis and many years later after excision of cutaneous melanoma, as the first sign of recurrence [1]. Recent studies have implicated that the expression of functional C-C chemokine receptor type 9 allows malignant melanoma cells to preferentially metastasize to the small bowel [2].

Metastasis when occurs to small bowel are usually small submucosal nodules, a pattern often described as "target lesion" [3]. It rarely presents with large, exophytic metastasis to the small bowel. We report an interesting case of an unusually large malignant melanoma metastasizing to small bowel which disappears intermittently to reappear on examination.

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## Case Report

A 50-year-old gentleman presented to us with 3-months history of pain abdomen, easy fatigability, and black colored stool. He has undergone multiple blood transfusions for symptoms of chronic iron deficiency anemia in the last 3-month duration. On further enquiring, patient gives history of excision of malignant melanoma from the right sole 5 years back. Physical examination revealed pallor with presence of "disappearing lump" in right iliac fossa. The lump was firm, mobile, 7 × 7 cm in size and surprisingly vanished the next day and reappeared later, suggesting the distal ileal origin. On laboratory investigation, his hemoglobin was 6.0 g/dl with normal liver and renal function test. Chest X-ray was normal. The esophagogastroduodenoscopy and colonoscopy revealed no overt bleeding source. Contrast-enhanced CT abdomen showed a thick, homogeneously enhancing mass lesion of size 7 × 8 cm in pelvis arising from the ileum, with multiple enlarged mesenteric and pelvic nodes. Provisional diagnosis of metastatic malignant melanoma was made in view of past history of cutaneous melanoma, and was planned for laparotomy after correction of anemia. At laparotomy, a large, exophytic blackish mass hidden in pelvis was seen arising from the distal ileum with multiple mesenteric nodes (Fig. 1). The proximal jejunal loop showed submucosal metastases at multiple sites (Fig. 2). Moreover, the small bowel mesentery and peritoneum was studded with "melanoma dust." Palliative resection and end to end anastomosis of ileal lesion was done, which was the culprit for small intestinal bleeding. Postoperative patient

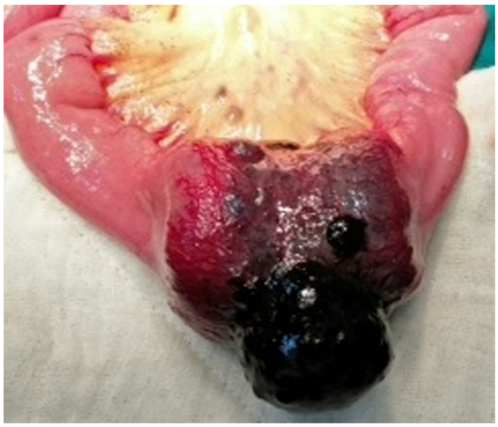

**Fig. 1** Intraoperative photograph showing large blackish exophytic tumor arising from distal ileum

did well and was discharged in a satisfactory condition. Histopathological examination of resected specimen showed extensive proliferation of atypical melanocytes reaching up to the subserosal region (Fig. 3). At 6 months of follow-up, patient is doing well and asymptomatic.

## Discussion

Metastatic tumor involving the small bowel is much more common than primary neoplasm. The most common metastases to small intestine are those arising from other intraabdominal organs. Metastases from extra-abdominal tumor do occur from lung and breast carcinoma or malignant melanoma. Cutaneous melanoma is the most common extra-abdominal source to involve the small intestine, with involvement seen in more than 50% of patients dying from the malignant melanoma [1, 2].

Only 1–5% patients of metastatic small bowel melanoma are diagnosed during their lifetime because the disease is undetectable in its early stages and diagnosis is made only after

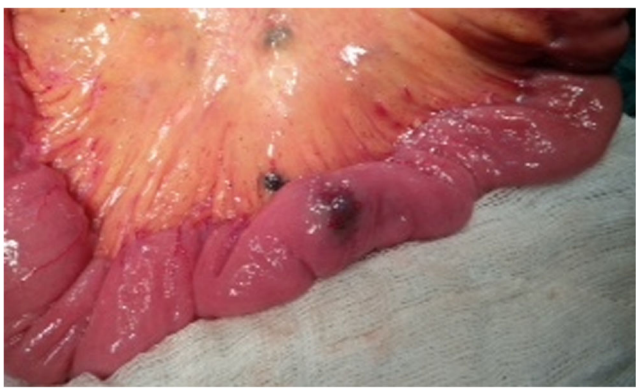

**Fig. 2** Intraoperative photograph showing jejunal submucosal blackish nodules, mesenteric lymph nodes, and melanoma dust

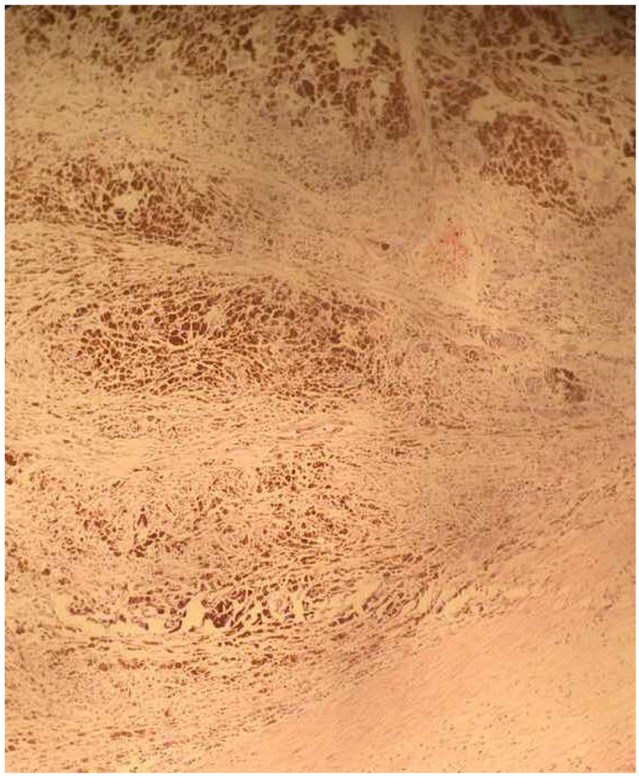

**Fig. 3** (H&E, ×100) Histopathological examination showing proliferation of atypical melanocytes reaching up to the serosa

complications or death [2, 3]. The metastasis typically develops 3 to 6 years after excision of cutaneous melanoma, but they may sometimes present as early as at initial diagnosis or remotely 180 months after detection of primary cutaneous melanoma [4]. The different risk factors for metastasis are superficial spreading melanoma and axial primary tumor site, clark level III or IV, high degree of histologic regression, presence of ulceration, and a high mitotic rate [5].

Most often, the metastatic melanoma to small bowel are multiple with multifocal polypoids distributed to both the jejunum and ileum and are clinically asymptomatic in its early stage. Symptoms if present can vary from abdominal pain, nausea and vomiting, melena, symptoms of chronic anemia, weight loss, and presence of disappearing abdominal mass when the tumor is large and ileal in origin as seen in the present case. Rarely, it can present to emergency with intestinal obstruction due to intussusceptions, enterorrhagia, or bowel perforation [6, 7].

In our patient, we were fortunate to palpate the occult lump present in the pelvis due to bowel peristalsis causing mass to appear in the right iliac fossa. Diagnosis should be considered in any patients with abdominal symptoms presenting with past history of cutaneous melanoma, but most often, the diagnosis is made only at the time of surgery for acute abdomen.

Diagnosis of malignant melanoma of small bowel is difficult by radiological study, because of its presentations as small submucosal nodules in its early stage. Imaging modalities

commonly used to detect metastatic lesions are multidetector CT scan, upper gastrointestinal series, double balloon enteroscopy, capsule endoscopy, and recently PET/CT scan. The typical findings that provoke consideration for metastatic lesion are discrete polypoidal mass with central ulceration, a pattern often described as Bull's eye lesion [1]. Multidetector CT detects enhancing mural and extra-mural lesions only when large in size as seen in present case. Moreover, upper gastrointestinal series and double balloon enteroscopy study might miss these small and distal bowel metastatic lesions. Hence, these are not the preferred diagnostic modality of choice because of its poor sensitivity. PET/CT scan has been shown to be more sensitive (86%) and specific (91%) than contrast-enhanced CT in the assessment of symptomatic patients [8]. More recently, PET/CT scan in combination with capsule endoscopy is an ideal investigation modality for patients with suspected metastatic melanoma presenting with unexplained anemia or gastrointestinal symptoms [8, 9].

Surgical resection with aggressive approach with aim of complete resection with no residual tumor (R0) is the treatment of choice. It includes wide surgical resection of tumor with sufficient bowel margin together with wedge of mesentery to remove regional nodes [10, 11]. Even in setting of diffuse abdominal metastasis, palliative resection of symptomatic lesions improves survival and quality of life. Median and 5-year survival rate after complete surgical resection with curative intent is 49 months and 41%, respectively. However, the overall median and 5-year survival after incomplete resection is only 6–9 months and less than 10%, respectively[12]. Hence, the prognosis of these patients is worse due to aggressive tumor biology, late diagnosis, and fast tumor growth in rich vascular and lymphatic supply of intestinal mucosa and presence of extra-intestinal metastases in 50% of patients at the time of diagnosis [13]. In our patient, the resection was done with palliative intent as the tumor was involving multiple jejunal sites with multiple mesenteric lymph nodes along with diffuse mesenteric deposits in the form of melanoma dust.

The role of adjuvant chemotherapy in metastatic melanoma is limited with very poor response rate [14]. However, recent development of immunotherapy has revolutionized the treatment of metastatic melanoma, preferentially by use of immune checkpoints (CTLA-4 or PD-1) blockade. In this context, ipilimumab was the first check point inhibitor gaining FDA approval for metastatic melanoma in 2011 [15]. Ipilimumab and pembrolizumab, monoclonal antibodies against the cytotoxic T lymphocyte antigen 4 (CTLA-4) and programmed cell death protein 1 (PD-1), respectively, have demonstrated survival benefit (10% to 30%), although modest when used alone or in combination in patients with metastatic melanoma [16]. Therefore, the eligible patients should be considered for immunotherapy for probable survival benefit.

## Conclusion

Metastatic malignant melanoma of small bowel is a rare entity presenting usually with small submucosal nodules. It is very unusual to see large bulky metastasis to ileum presenting as disappearing lump. Diagnosis is often late and made by high index of suspicion in patients with history of cutaneous melanoma who complains of pain abdomen, anemia, or palpable abdominal mass. Surgical resection even if palliative is recommended to improve survival and quality of life.

**Compliance with Ethical Standards**

**Consent** Informed consent was obtained.

**Conflict of Interest** The authors declare that they have no conflict of interest.

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