Unusual presentation of more common disease/injury

Acute small bowel obstruction due to intussusception of malignant *amelanotic* melanoma of the small intestineRajan B Patel,¹ Nitin C Vasava,¹ Minesh B Gandhi²¹Department of Surgery, Smt. NHL Municipal Medical College, Ahmedabad, Gujarat, India²Department of Pathology, Smt. NHL Municipal Medical College, Ahmedabad, Gujarat, India

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Summary

Malignant melanoma is a common malignancy and cutaneous melanoma showed an unusual predilection to metastasise into the small intestine. At autopsy metastatic deposits found in 50–60%, but less than 2–4% of melanoma patients diagnosed to have gastrointestinal (GI) metastasis during the disease course and 4–9% GI melanoma had unknown primary melanoma, which should be considered metastatic because primary lesion may be very small to diagnose or had regressed spontaneously. Few cases reported to have intussusception as a presenting feature. We report a case of *amelanotic* metastatic melanoma presented with intussusception and without known primary melanoma. The patient presented with abdominal pain, vomiting and distension. He was diagnosed as having acute intestinal obstruction as confirmed by erect abdominal x-ray. Ultrasonography showed small intestine intussusception. Emergency laparotomy revealed ileo-ileal-type intussusception with intraluminal mass, which was resected and end-to-end anastomosis of bowel was performed. Histopathological and immunohistochemical examination of the resected specimen showed *amelanotic* melanoma.

BACKGROUND

Malignant melanoma is a common malignancy and its prevalence is increasing. It represents 1–3% of cancers in the USA. Cutaneous malignant melanoma had shown an unusual predilection to metastasise to the small intestine and mesentery. Few cases reported to have intussusception as a presenting feature. Incidence of gastrointestinal (GI) metastasis from unknown primary origin ranges from 4% to 9%. Ileal intussusception secondary to *polypoidal* growth of *amelanotic* malignant melanoma with unknown primary was not reported in the literature.

CASE PRESENTATION

A 60-year-old man presented with complaints of para-umbilical colicky pain, bilious vomiting and inability to pass stool or flatus for 1 day. On examination, he was haemodynamically stable, had generalized abdominal distension without guarding and had absent bowel sounds. The patient had no medical or surgical illness in the past. Erect abdominal x-ray showed centrally located multiple air fluid levels. Ultrasonography of the abdomen revealed intussusceptions, fluid-filled dilated small bowel loops with a maximum diameter of 29 mm and had no free fluid in the abdomen. The patient was operated for emergency laparotomy and had intussusception around 60 cm proximal to ileo-caecal junction (figure 1). Reduction of intussusception had revealed 3 × 3 cm polypoidal mass arising from mesenteric side of the small bowel. No mesenteric lymph node enlargement or other abnormalities detected in peritoneal cavity. Mass along with 10 cm bowel was resected, end-to-end bowel anastomosis was performed. Resected specimen was sent for histopathology examination.

INVESTIGATIONS

Histological examination of the specimen revealed a sub-mucosal tumour composed of nodular pattern separated by fibrous septa. Tumour cells were round to polygonal and focally spindled. Cells showed highly pleomorphic, hyperchromatic and vesicular nuclei with prominent nucleoli. Multinucleated and bizarre tumour giant cells were seen. Numerous mitosis with abnormal mitotic figures were present (figure 2) suggestive of malignant lesion. The tumour was invading the submucosa and sero-muscular wall. Intestinal mucosa was spared by the tumour.

DIFFERENTIAL DIAGNOSIS

Histological impression was undifferentiated malignant tumour with the possibility of gastrointestinal stromal tumour (GIST) or *amelanotic* metastatic malignant

Figure 1 Intussusceptions of the small intestine.

Figure 2 Histopathology examination of the melanoma tumour.

melanoma. Immunohistochemical staining of the specimen revealed S-100 (positive), Vimentin (positive) and negative for CK, CK-7, CK-20, EMA and LCA, confirming the diagnosis of amelanotic malignant melanoma.

OUTCOME AND FOLLOW-UP

Postoperative recovery of the patient was uneventful and was discharged on the 7th day. Postoperatively vigorous search for primary lesion at common sites like skin, anus, eye, oral cavity, nail beds and other locations did not reveal any primary lesion. Postoperatively the patient was followed up in OPD for 6 months and had no complaints.

DISCUSSION

Malignant melanoma is a common malignancy and can occur at various locations where melanocytes or melanoblasts are present. It is presented mostly in the skin, less frequently in the eye (choroids layer), leptomeninges, oral cavity, nasal mucosa, pharynx, oesophagus, bronchus, vaginal or anorectal mucosa and nail beds.¹

Cutaneous melanoma showed an unusual predilection to metastasise to small intestine and mesentery. At autopsy GI metastasis of melanoma reported 50–60%, but only 2–4% of patients with melanoma will be diagnosed with GI metastasis during the course of their disease. The primary melanoma in patients with GI metastasis is typically located in the extremities (15–57%), trunk (13–54%) and less frequently in the head and neck (5–33%). In 10–26% of cases, the primary lesion is occult. The period observed between the diagnosis of primary melanoma and GI metastasis is reported as up to 54 months. The most common sites of metastasis in the GI tract are the jejunum and ileum.² The incidence of metastatic melanoma from unknown primary origin ranges from 4% to 9% and should be considered metastatic because primary lesion may be very small to be identified by clinical and laboratory investigations or regressed spontaneously.³

There appear to be two subsets of primary melanoma, one that occurs among younger patients, which is more aggressive with rapid metastasis leading to early death, and the other that occurs among older patients, which is more indolent and metastasises less rapidly.⁴

Around 70% of the patients of intestinal melanoma presented with at least one of the symptoms related to GI

tract, whereas 30% remained clinically asymptomatic. Common GI presentations included abdominal pain, nausea, vomiting with or without acute symptoms (60%), obstruction/intussusception (27%) and melena/bleeding (26%).⁵ CT scan is the most preferred modality of investigation and can show the involvement in the form of (a) most common is intraluminal polypoidal mass and intussusception, (b) cavitated or ulcerating lesion, (c) diffuse infiltration and (d) implants.^{6–7} The whole-body positron emission tomography (PET) imaging using 2-deoxy-2-fluoro-D-glucose (FDG) has been shown to be highly accurate in assessing patients with metastatic malignant melanoma.⁸

The presence of metastatic lesion was confirmed by surgical exploration in 80%, endoscopic procedure in 11% and percutaneous biopsy in 5% of the patients.⁵

Surgical intervention is often required when bowel metastases diagnosed incidentally or become symptomatic. The patients showed survival of 31 ± 5.2 months who had complete resection of demonstrable disease, 9.6 ± 5.9 months who underwent palliative enteric bypass or debulking procedures and 9.6 ± 3.6 months who received either chemotherapy alone or symptomatic management.^{5–9}

Recently, it is found that functionally active CCR9 on melanoma cells facilitate migration to metastasise in response to the chemo attractant effect of CCL25 (thymus-expressed chemokine). This CCR9–CCL25 axis may explain the high incidence of melanoma metastasis to the small intestine.¹⁰

In conclusion, the cases of acute small bowel obstruction with intussusceptions suspect that the malignant melanoma may be differential diagnosis. Ultrasonography works as a good screening method for the diagnosis of intussusception, but CT scan is diagnostic for intussusception and PET scan is required for preoperative diagnosis of malignant melanoma. Immunohistochemistry is required for confirming the diagnosis. The patients with preoperative diagnosis of small bowel malignant melanoma have better survival.

Learning points

- ▶ The cases of acute small bowel obstruction with intussusceptions suspect that the malignant melanoma may be differential diagnosis.
- ▶ CT scan and positron emission tomography scan are required for the preoperative diagnosis of malignant melanoma.
- ▶ Immunohistochemistry is required for confirming the diagnosis.
- ▶ The patients with preoperative diagnosis of small bowel malignant melanoma have better survival.

Competing interests None.

Patient consent Obtained.

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