Primary Intestinal Melanoma Presenting as Intussusception - A Rare Case Report

Sandeep Chandrakar¹, Rajendra Ratre², Lizrose Kujur³, T. Narasimha Murty⁴

¹Department of General Surgery, Atal Bihari Vajpayee Memorial Medical College, Rajnandgaon, Chhattisgarh, India. ², ³, ⁴ Department of General Surgery, Pt JNM Medical College and Dr. BR Ambedkar Memorial Hospital, Raipur, Chhattisgarh, India.

INTRODUCTION

Malignant melanomas mostly begin as an oculocutaneous lesion and then metastasise to other sites over time. Gastrointestinal melanomas are rare but primary intestinal melanomas are extremely rare. They account for 1 - 3% of malignant tumours of GI tract.¹ ² Although few cases have been reported, the existence of primary intestinal melanoma is still doubtful. It is difficult to differentiate between primary and secondary intestinal melanomas based on their clinical picture alone. Some authors believe that the metastatic melanoma into the intestine manifest symptoms in the patient after complete regression of some old cutaneous lesion.³ ⁴ But some believe that primary melanoma of small intestine originates from Schwannian neuroblast cells, neural crest melanoblasts, or amine-precursor uptake and decarboxylation (APUD) cells through neoplastic changes.⁵ ⁶ They are unrecognized until late due to absence of symptoms, but if symptoms occur, they include abdominal pain, gastrointestinal bleeding and sometimes with intestinal obstruction and few present acutely with perforation. By any origin, malignant melanoma is an aggressive tumour with very poor prognosis and low survival rates, even after surgery. Here we report a case of primary intestinal melanoma in a young patient who presented as intussusception of small intestine.

PRESENTATION OF CASE

A 30-year-old female presented to our emergency unit with the complaints of abdominal pain, dark coloured stools since 2 months and vomiting since 5 days. It was dull aching pain which was insidious in onset, gradually increasing and not relieved with medication. On examination, abdomen was distended with mild tenderness in right iliac fossa and right lumbar area. On per rectal examination, red current jelly stools were seen. She had regular menstrual cycles, two children with no antenatal, intrapartum, or postnatal complications. Her last child birth was 2 years back. Her past medical or surgical history was insignificant. Patient was admitted in our department for further management.
**Investigations**
Routine laboratory investigations were found to be normal, X-ray abdomen erect shows minimal air fluid level. Ultrasonography (USG) abdomen revealed telescoping of distal ileal segment along with its mesentry into the cecum and ascending colon up to hepatic flexure suggestive of ileo-cecal intussusception. Contrast enhanced computed tomography (CECT) abdomen showed telescoping of part of ileum, cecum along with its mesentry through the ascending colon up to hepatic flexure for a segment of 22 cm with proximal small bowel obstruction. There was ill defined exophytic heterogeneously enhancing mass lesion noted over wall of intussusception measuring 2.7 x 3 cm suggesting neoplastic mass. There were multiple variable sized pleural and parenchymal based enhancing soft tissue density nodules noted diffusely involving bilateral lung largest measuring 2.3 x 2 cm. suggestive of metastatic deposits. CA 125 was slightly raised up to 89.1 U / ml. Other tumour markers including CA 19.9, alpha-fetoprotein (AFP), lactate dehydrogenase (LDH) and human chorionic gonadotropin (HCG) were within normal limits. Uterus and bilateral ovaries appears normal clinically and radiologically.

**Clinical Diagnosis**
Ileo-cecal intussusception with distal ileal neoplastic mass with lung metastasis.

**Differential Diagnosis**
1. Primary small bowel benign or malignant tumour.
2. Small bowel lymphoma.
3. Metastatic small bowel tumour from unknown primary.

**Discussion of Management**
After pre anaesthetic check-up, patient was planned for surgery. Intra-operatively, distal ileum was seen protruding along with cecum and its mesentry into ascending colon up to hepatic flexure. Hence, proceeded with right hemicolecction along with ileostomy.

**Pathological Discussion**
Gastrointestinal (GI) tract melanomas can be either primary or metastatic. Small intestine and colon doesn’t contain melanocytes so the occurrence of melanoma at these places is thought to be due to neural crest cells which migrated to distal ileum through omphalomesentric canal. Although it can arise at any site from oral cavity to anus, small intestine is the most common site in GI tract. There have been many theories to prove a GI melanoma to be primary tumour and not metastatic from an unknown primary lesion. The unknown primary cutaneous sites constitute 26 % of the cases. As proposed by Blecker et al. presence few criterias can diagnose a primary GI melanoma –
1. A single mucosal lesion in intestine.
2. Intramuscosal melanocytic lesions in the overlying or adjacent intestinal epithelium.
3. Absence of any melanocytic lesion on the skin.

Histologically, lymphocytic invasion of dermis with melanophages, blood vessels and fibrosis characterise a metastatic lesion. In our case, there was no primary cutaneous or mucosal lesion at any other site with involvement of intestine as the sole finding. The presence of satellite lesions in adjacent intestinal epithelium, lack of serosal involvement, absence of lymphocytic invasion strengthen our diagnosis of primary intestinal melanoma. Thus, we second the belief of some other authors who claim the existence of primary melanoma of GI tract.

There is very scarce data on primary intestinal melanomas due to very few cases reported till date. A thorough search into literature yielded few cases of primary small bowel melanoma from which we drew conclusions regarding management of intestinal melanoma. Diagnosis of presence of mass can be made using ultrasonography, CT scan, endoscopy or colonoscopy. But in study by Hadjinicolaou et al., video capsule endoscopy helped in detecting a jejunal mass when the above investigations failed. Sinagra E et al. reported a case of primary intestinal melanoma in which they found metastasis from a unknown primary lesion. A case reported by Gabali et al. presented like a gastrointestinal stromal tumour. Two cases were studied by Kwan Mo Yang et al. Radical surgery is considered the best treatment option for patients with GI melanoma. In all the cases, surgery was the only definitive treatment. In most of the cases, the patient either failed to follow up or died within 1 year of surgery. In our patient, no primary lesion in skin, eye or mucosal site could be identified and history was not suggestive of any such lesions in the past or treatment for the same. She died 6 months after surgery. Primary GI melanoma is associated with very poor prognosis and is more aggressive as compared to metastatic melanomas. The five year survival of both primary and secondary melanomas is nearly 10 % and median survival ranges from 4 - 6 months only. The role of systemic therapy after surgery...
for primary GI melanomas is yet to be studied given the paucity of cases in literature.

**FINAL DIAGNOSIS**

Primary intestinal melanoma with ileo-cecal intussusception with lung metastasis

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