CASE REPORT

Primary Malignant Melanoma of The Small Intestine: A Rare Case Reported

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ABSTRACT

Primary small intestinal melanoma is a rare malignant neoplasm, for which scarce literature offers limited evidence for optimal management. We present a case of a 60-year-old female with primary melanoma of ileum. She presented with left sided abdominal pain, decreased appetite, and history of diarrhoea for 15 days. She developed absolute constipation during her preoperative investigations, in which CT scan revealed ileoileal intussusception. She underwent laparotomy for the resection of intussuscepted bowel segment, which revealed a greyish brown mass attached to bowel, later confirmed by histopathology as melanoma. Postoperative detailed clinical investigation revealed no primary cutaneous, hepatic, pulmonary, ocular, or cranial melanotic lesion. Hence, the lesion was classified as primary small bowel melanoma. She was referred to oncologist who started her on adjuvant chemotherapy. The patient expired of cardiopulmonary arrest three months after her diagnosis due to post Chemotherapy side effects.

Key Words: Gastrointestinal melanoma, gastrointestinal tumour, ileal melanoma, primary melanoma

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INTRODUCTION

Small bowel melanoma is an infrequent tumour, which makes the diagnosis difficult and often presents late[1]. When found, it is usually a metastatic lesion from a primary cutaneous lesion[4]. However, rarely primary melanomas of the small intestines have been reported[1,5].

Primary GI melanomas are most commonly reported in anorectal and oropharyngeal regions[2]. Due to very low incidence, the primary small intestine melanomas have very limited clinical data available[2]. This impedes concluding any specific management as guideline for the disease. Differentiating between primary and secondary melanoma of the small intestine yet remains another concern.

We present a rare case of primary malignant melanoma of the small intestine and discuss relevant literature.

Case

A 60-year-old female came with the history of left sided abdominal pain for two weeks, which increased in intensity two days prior to her presentation in emergency department. The pain was colicky in nature, of moderate to severe intensity, aggravated by taking small meals, and relieved temporarily by antispasmodic medication. Patient also complained of decreased appetite and history of diarrhoea, which were watery in consistency without any foul smell or blood, for the last 15 days. She developed constipation associated with bilious vomiting in the previous two days.

On arrival in emergency department, she was dehydrated with pulse of 102 bpm, blood pressure of 160/60 mmHg, and afebrile temperature. Her abdomen was distended, with generalized deep tenderness and exaggerated gut sounds. On digital rectal examination faecal staining was present with no fresh blood or melena. The rest of the examination was unremarkable. No sign of anaemia, jaundice, cyanosis, or lymphadenopathy was observed. The patient was admitted with diagnosis of subacute intestinal obstruction for further management.

Laboratory investigations revealed Hb 13.1 g/dl, WBCs 7.8 $x10^{12}$/L, platelets 206, and CRP 168 mg/L, in

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addition to normal UCEs and LFTs. Her chest X ray was normal, but abdominal x ray observed air fluid levels in dilated small bowels (fig. 1a and b).

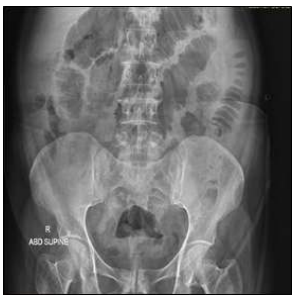

fig. 1a

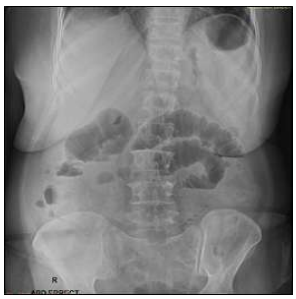

fig. 1b

Fig. 1a: Abdominal X-Ray Supine Showing Dilated Small Bowel
Fig. 1b: Abdominal X-Ray Erect Showing Air Fluid Levels

CT scan depicted ileoileal intussusception with soft tissue thickening and proximal small bowel dilatation. Concurrent hepatomegaly was also observed (fig 2a and b).

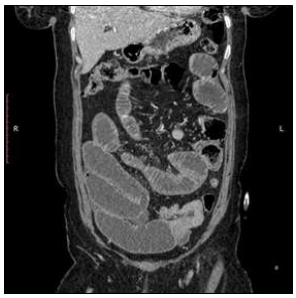

fig. 2a

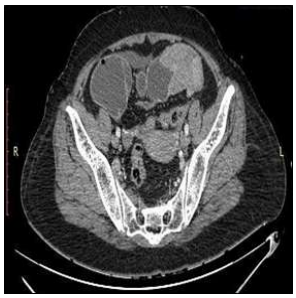

fig. 2b

Fig. 2a and 2b: CT Scan Abdomen Transverse and Coronal Section Showing Ileoileal Intussusception

During preoperative investigations, the patient developed absolute constipation. In view of her worsening condition, exploratory laparotomy was performed and an ileoileal intussusception was found with a dark brown nodule over it. The mass 4x4 cm in size and was covered by serosa extending and involving the mucosa. The involved area was resected and cut ends were exteriorised as double barrel ileostomy (fig. 3a and b).

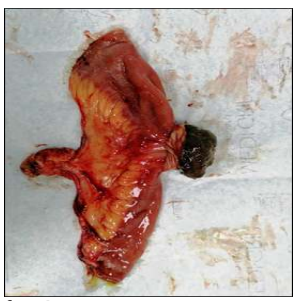

fig. 3a

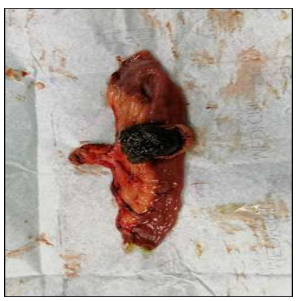

fig. 3b

Fig. 3a and 3b: Surgical Specimen of Resected Ileoileal Intussusception

Histopathology of the surgical specimen reported the mass as malignant melanoma with immunohistochemical stains positive for S-100, Hmb45 and Melan A. It contained five lymph nodes, out of which three were positive for nodal metastasis. The tumor-free ileum extended 7 cm proximally and 8 cm distally from the site of melanoma mass (fig. 4a,b,c and d).

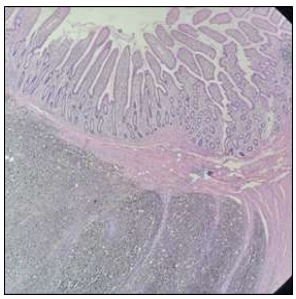

fig. 4a

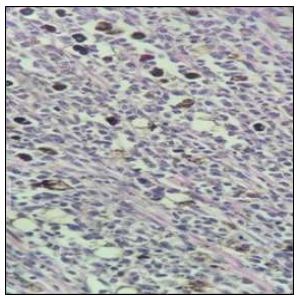

fig. 4b

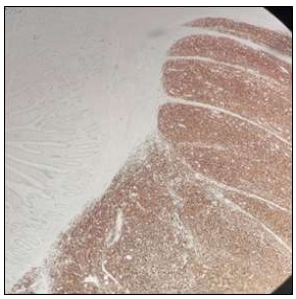

fig. 4c

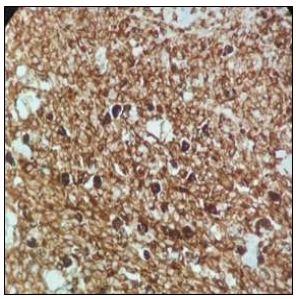

fig. 4d

Fig. 4a and 4b: H&E stain showing wall of intestine showing neoplastic lesion composed of sheets of spindles to epitheloid cells having marked nuclear pleomorphism, hyperchromasia, abundant cytoplasm, and prominent eosinophilic nucleoli. Tumour (hyper pigmented area) is involving dermis but mucosa is uninvolved. Large and small asymmetric tumours with distinct nucleoli and large tumour cells with melanin deposition were observed. Fig. 4c and 4d: Immunohistochemical staining reveals that the tumour is S-100 (+), HMB-45 (+), Melan-A (+).

After the diagnosis of melanoma was established, the patient underwent a clinical and radiological evaluation. The examination of skin, eyes, and anus was negative for any primary lesion. Chest CT scan was performed along with review of abdominopelvic CT scan, which excluded any pleuropulmonary, hepatic, adrenal, or bony lesion, whether primary or metastatic. Therefore, it was concluded that the intestinal mass was primary melanoma of small intestine.

The patient was discharged with ileostomy care counselling and instructions. She was referred to oncologist for adjuvant chemotherapy. She was started on monthly dacarbazine 450mg for five days. After third cycle of dacarbazine patient developed brown pigmented lesions all around the body (fig. 5). Before she could undergo the fourth cycle of her chemotherapy regime, the patient expired due to cardiopulmonary arrest.

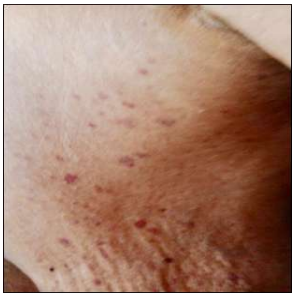

Fig. 5: Brown Pigment Lesions Following the Third Cycle of Chemotherapy

Discussion

Malignant melanoma is a common skin neoplasm which can be rarely observed in other body sites. Primary melanoma of the gastrointestinal tract is uncommonly found in clinical practice; therefore, no definite guidelines exist for its optimal management. The literature has only been able to offer limited case studies as evidence to guide the treatment of the disease.

Primary mucosal melanoma can arise at any site along the GI tract. M.C. Cheung et al. reviewed the available literature and observed that the common sites for presentation include oropharyngeal (32.8%), anal (31.4%), and rectum (22.2%)[1,4]. However, the small intestines account for only 2.3% as the site of primary melanoma[1,4].

Authors have even argued on the possibility of primary melanoma of the small intestine. Although some have defined it as a symptomatic metastatic lesion from a regressed primary cutaneous lesion, few hypotheses have attempted to explain it as a primary disease. Migration of neural crest cells to distal ileum via omphalomesenteric canal offers a justification for distal ileum to be the most common site of the primary melanoma in small intestines. Another rationale can be the origin of tumour from enteric neuroendocrine non-cutaneous tissue in the form of amine precursor uptake decarboxylase cells that have undergone neoplastic transformation1. The latter can also interpret the non-ileal intestinal malignant melanomas[3].

A criterion was proposed by Blecker et al. for the diagnosis of primary intestinal melanoma[1]. It included absence of melanoma or atypical melanocytic lesion of skin, absence of extraintestinal metastatic spread of melanoma, and presence of intramural lesions in overlying or adjacent intestinal epithelium[1,3]. Our patient fulfilled all the conditions of the above recommended criteria in addition to the histopathology, which allowed us to diagnose it as primary melanoma of ileum.

The presentation of primary small intestinal melanoma is dependent on the location and size of the tumour. It can be observed with non-specific signs, like weight loss and anaemia, or intestinal obstruction, which was the primary finding in our case. The primary management for the disease remains surgery in all cases, which may be followed by adjuvant chemoradiotherapy. M.C. Cheung observed no survival benefit of radiotherapy, whereas sufficient data was not available to comment on chemotherapy[2]. Our patient was recommended chemotherapy as the only adjuvant treatment for the disease.

Primary melanoma of the small intestine is shown to have poor prognosis compared to that of skin. M.C. Cheung et al. observed high rates of mesenteric lymph node metastasis, and sixteen months of median survival[4]. Similarly, Hadjinicolau AV also showed only 50% one-year postoperative survival of these patients, and high tumour recurrence[3]. The case presented here by our team will contribute to the knowledge of the disease and assist in decision making for its optimal management.

In conclusion, our case fulfilled the criteria of available literature to be classified as primary small intestinal melanoma. The patient was disease free till four months of postoperative follow up. Reversal of ileostomy was planned after completion of chemotherapy, but the patient expired prior to it.

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

Author's contribution: SS and UHAR conceptualized and supervised the current study; KRN: performed the experimental work and collected the required data; UHAR: wrote the first draft of the article; KRN and MRM: critically revised the manuscript, helped with statistical analysis, and wrote the discussion portion of the article.

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