Original Case Report

Malignant Melanoma of Intestine: Primary or Secondary?
3 Case Reports with Review of Literature

Authors
Ann Mili Kuriakose1*, Sankar Sundaram2, Renu Thambi3, Priya P.V4

1Junior Resident, Department of Pathology, Government Medical College, Kottayam, Kerala, India
2Professor and Head, Department of Pathology, Government Medical College, Kottayam, Kerala, India
3Assistant Professor, Department of Pathology, Government Medical College, Kottayam, Kerala, India
4Additional Professor, Department of Pathology, Government Medical College, Kottayam, Kerala, India

Abstract
Gastrointestinal (GI) melanomas are a rare diagnostic entity. Although there have been cases of primary melanomas in the GI tract, many debate their true origin: the primary versus secondary from an undetected regressed primary lesion (melanoma of unknown primary). We present 3 case reports of Melanoma GIT – primary malignant melanoma involving anorectum and duodenum and a case of malignant melanoma metastasis to ileum.

Keywords: Malignant Melanoma, Primary, Metastasis.

Introduction
Melanoma is one of the most prevalent neoplasms diagnosed, with most cases arising from a cutaneous origin. Though there are many case reports depicting melanoma metastases to GIT, there are only rare descriptions in literature on primary gastrointestinal melanoma. Both diagnosis and management of these tumors is challenging because of rarity of the disease.1 The most common site for primary GIT melanoma is anorectum, which accounts for 0.4-1.6% of all melanomas and 1% of all anorectal malignant tumors.2 Small intestinal melanomas are rare with a few case reports. Rate of GIT metastasis of malignant melanoma detected clinically averages to only 2%, due to the nonspecific symptoms and signs of GIT involvement.3 Within the GIT, small bowel is more frequently involved, followed by stomach, large intestine and esophagus.4

Case Reports
Case 1: Malignant Melanoma as Primary in Anorectum
A 46 year old woman presented with altered bowel habits for a duration of 3 weeks. Clinical examination revealed a polypoidal lesion in the anorectal region. Biopsy was diagnostic of malignant melanoma for which Abdomino-perineal resection was done.
Fig A: Extended right hemicolectomy specimen containing a 5.6 cm exophytic, partially obstructing lesion.

Fig B: Histology showing sheets of large tumour cells with irregular nuclear contours and vesicular nuclei and prominent eosinophilic nuclei (40X, Inset : 100 X)

Case 2: Malignant Melanoma as Primary in Duodenum
A 52 year old woman presented with melena and endoscopy showed a duodenal ulcer. Biopsy showed duodenal tissue infiltrated by pleomorphic cells, some showing intracytoplasmic brownish pigment. Masson Fontana stain and immunohistochemistry with HMB-45 confirmed the diagnosis of Malignant Melanoma.

Fig C: sheets of pleomorphic cells with hyperchromatic nuclei (40 X)

Fig D: IHC with HMB -45 showed cytoplasmic and membranous positivity.

Case 3: Malignant Melanoma as Secondary in Ileum (Metastasis)
57 year old male presented to casualty as acute abdomen. Emergency laparotomy and ileal resection was done. Received specimen of ileum had multiple areas of blackish discolouration and microscopy showed pleomorphic cells with brown pigment. Further, a detailed history revealed past history of malignant melanoma of nasal cavity with lung metastasis.

Fig E & F (10x and 40X): Pleomorphic cells of epithelioid & spindle type involves sub- mucosa, muscle and serosa.

Discussion
GI tract malignant melanoma is an uncommon neoplasm that may be either primary or metastatic. Primary mucosal melanoma can arise at any site within the GI mucosa from oral cavity to anus. It is most common in anorectum (anal canal, 31.4%; rectum, 22.2%) and oropharynx (32.8%), whereas
oesophagus (5.9%), stomach (2.7%), small intestine (2.3%), gallbladder (1.4%), and large intestine (0.9%) are extremely rare sites of origin. Small intestinal melanomas are rare with only a few case reports, mostly observed in jejunum and ileum. Primary melanoma in duodenum is extremely rare and controversial.

Melanomas of GIT that was diagnosed in our institution during a period of 1 year (June 2018-2019) is illustrated in Table 1.

Table 1

| PRIMARY (5) | UPPER LIP—1 | RETROMOLAR TRIGONE—1 | DUODENUM—1 | ANORECTUM—2 | METASTATIC(1) | ILEUM—1 |

There are many postulations regarding the cell of origin of GI melanomas. One hypothesis suggests that melanoma arises from the neural crest cells existing in the oesophagus, stomach and anorectum. Another potential origin for primary melanoma of small intestine is melanoblastic cells of the neural crest that migrate to distal ileum through omphalo-mesenteric canal. Accordingly, ileum, which represents the distal end of the omphalo-mesenteric canal, should be the most common site of primary malignant melanoma within small intestine. Another hypothesis was that these tumours originate from enteric neuroendocrine cells or amine precursor uptake decarboxylase cells (APUD cells) that have undergone neoplastic transformation. This would account for the remaining non-ileal intestinal melanomas.

Although melanomascans arise from GI tract, it is always important to rule out the possibility of a metastatic disease. Many suggest that melanoma of GI tract occurs as a result of spontaneous regression of an oculo-cutaneous lesion, also known as melanoma of unknown primary. In fact, in one study of 437 cutaneous melanomas, 12.3% showed at least partial regression.

Conclusion

Melanoma in GIT is a rare occurrence in which primary lesions are extremely unique. A diagnosis of GI melanoma should warrant surgeon for a thorough physical examination, including eyes, mouth, nasopharynx, genitalia, and anorectum. Major lymph nodes should also be examined that can guide towards a possible primary site. Imaging techniques like positron emission tomography (PET) scan should be advocated. Differentiation between primary and metastatic melanoma of GIT may be challenging but is essential for a treatment plan.

References
