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Primary Malignant Melanoma of the Jejunum

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Abstract

Background

Malignant melanomas in the small intestine are commonly encountered as the metastases from primary cutaneous lesions. Even though it's rare, it can also develop as a primary mucosal tumour in the gastrointestinal tract. We present a rare case of primary jejunal melanoma.

Case presentation

A 72-year-old male was investigated for occult gastrointestinal bleeding following three consecutively positive stool occult blood tests and his haematological investigations showed that severe iron deficiency anaemia. Upper gastrointestinal endoscopy and colonoscopy did not revealed any abnormality. Capsular endoscopy was performed and found to have a distal jejunal lesion with ulceration, bleeding and blackish discolouration. It was unsuccessful, even though we attempted single balloon enteroscopy to reach the site to obtain a biopsy and following contrast enhanced CT of the abdomen and pelvis the distal jejunal mass was confirmed with no abnormality found in the rest of the study. Since the CT further revealed that it was approximately 5 cm tumour arising from the jejunal wall and growing in to the lumen of the jejunum, Ultra sound guided Fine needle aspiration cytology attempted and found to have a blackish pigmented aspiration and later it was confirmed melanin pigment rich aspirate during the cytological examination. The mass was confirmed on laparotomy and histologically diagnosed as melanoma. Extensive postoperative clinical examination revealed no cutaneous lesions. Following the curative resection the patient recovered uneventfully.

Conclusions

Primary intestinal malignant melanoma in jejunum is an extremely rare occurrence. Complete surgical resection is the treatment of choice. The lymph node metastasis have the worse prognosis.

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Bullets points of the study highlights

What is already known?
• Primary malignant melanoma of small intestine is a rare entity.
• Diagnosis of a primary malignant melanoma of the small intestine from a metastatic lesion is very challenging.
• Carries a poor prognosis
What is new in this study?
• Primary malignant melanoma in jejunum is reported very few in world literature, since it is an extremely rare entity and this is one of the few.
• As far as we are aware this the first case which the diagnosis was made pre operatively by ultrasound guided FNAC.
What are the future clinical and research implications of the study findings?
• This will help to build up a larger collection of data on small intestine malignant melanoma worldwide.
• Also this will contribute to future studies on treatment modalities and to ascertain the prognostication factors.

Keywords – Melanoma, Jejunum, Bleeding, Primary, Obstruction.

I. Introduction

Primary malignant melanoma of small intestine is a rare entity since the majority are metastases from primary cutaneous lesions [1]. To ascertain a diagnosis of a primary malignant melanoma of the small intestine from a metastatic lesion is very challenging. We present a rare case of primary malignant melanoma of the jejunum.

II. Case presentation

A 72-year-old male was investigated for anaemia and episodic lower left sided abdominal pain. His clinical history and examination did not revealed any significant medical, surgical commodity or malignant diseases. However, investigations he was found to have had three consecutively positive stool occult blood tests. Furthermore his haematological investigations showed a severe iron deficiency anaemia. Initial workup with upper gastrointestinal endoscopy and colonoscopy did not revealed any abnormality. Capsular endoscopy was performed and found to have a distal jejunal lesion with ulceration, bleeding and surrounding blackish discolouration. It was unsuccessful, even though we attempted both ante and retrograde single balloon enteroscopy to reach the site to obtain a biopsy. Since we do not have the facility of double balloon enteroscopy, patient underwent a contrast enhanced CT of the abdomen and pelvis and a distal jejunal mass was confirmed with no abnormality found in the rest of the study. Since the CT further revealed that it was approximately 5 cm tumour arising from the jejunal wall and growing in to the lumen of the jejunum, Ultra sound guided Fine needle aspiration cytology attempted and found to have a blackish pigmented aspiration and later it was confirmed melanin pigment rich aspirate during the cytological examination.

Exploratory laparotomy revealed protruding ulcerated intraluminal mass from the distal Jejunum wall. Careful examination of the abdominal cavity revealed no macroscopic evidence of metastases. Small bowel resection was performed with side-to-side anastomosis. He had an uneventful recovery and was discharged on postoperative day 7. Surgical specimen was 25 cm in length and included a 7.2 × 5.3-cm mass (Fig. 1 and 2). The resection margins and retrieved 7 lymph nodes were free from tumour invasion. Histology revealed diffuse atypical cells infiltration forming solid sheet with cells of medium-to large-sized with irregular nuclear contours, macronucleoli, and numerous mitotic activities. Immunohistochemistry revealed the presence of tumour cells that were positive for the melanoma markers such as Melan-A, HMB45, S100 protein, and SOX10.

After the diagnosis of melanoma was established, the patient underwent examination of skin, eyes, oesophagus, and anus was negative for primary melanoma. The chest CT, brain MRI, and PET-CT scan did not report metastatic disease. Therefore, the resected lesion was confirmed to be a primary melanoma of the small intestine. The patient was scheduled for follow-up at regular intervals every 3 months for the 1st year and every 6 months for up to 5 years. .

III. Discussion

It has been reported that malignant melanoma can occur any site within the gut mucosa. However, primary mucosal malignant melanoma in small intestine is a rare site of origin (2.7%). It is most common in anorectal (anal canal, 31.4%; rectum, 22.2%) and oropharynx (32.8%). According to literature primary melanoma in jejunum is an extremely rare occurrence. [2]

Primary melanoma of the small intestine remains still a mystery since the absence of melanocytes in the small

intestine. Even though it's rare according to the literature most common site of the small intestine to get a primary melanoma is the ileum since embryologically ileum represents the distal end of the omphalomesenteric canal which contains melanoblastic cells of the neural crest. The primary melanoma of the jejunum origins may be due to noncutaneous tissue in the form of amine precursor uptake decarboxylase cells that have undergone neoplastic transformation [3].

It is difficult to differentiate between primary intestinal melanoma and intestinal metastatic deposits based on histopathological features alone. However a primary GI mucosal melanoma is considered in patients with no evidence of concurrent melanoma or atypical melanocytic lesion of the skin, absence of extraintestinal metastatic spread of melanoma, and presence of intramucosal lesions in the overlying or adjacent intestinal epithelium [1]. This important differentiation is related to the prognosis since primary intestinal melanomas carry the worst prognosis [1, 2]. According to the available data this may be due to the late diagnosis of melanoma in the GI tract, aggressive tumour behaviour, and fast tumour growth supported by the rich vascular and lymphatic supply of the intestinal mucosa [4].

It is difficult to assess the prognostic factors in patients with primary malignant melanoma of small intestine because it is rare. According to literature, location of tumour, advanced tumour stage, failure to undertake surgical resection, positive lymph node status, and age have been found to be independent predictors of poorer outcome [2]. However, larger collection of data is needed to ascertain these assumptions [2].

Surgical excision with tumour free resection margins with mesenteric lymph nodes is the main treatment option for primary melanoma of the small intestine. In these patients, systemic adjuvant therapy has a limited role, and chemotherapy regimens have very low response rates [3, 5, 6].

IV. Conclusions

Primary intestinal malignant melanoma in jejunum is an extremely rare occurrence. The definitive diagnosis is established after thorough investigations to exclude the existence of a primary lesion elsewhere. Complete surgical resection is the treatment of choice. The lymph node metastasis have the worse prognosis.

Declarations

Ethics approval and consent to participate

Informed written consent was obtained to write the case report and the approval of Nawaloka Hospital Ethical Review Committee was obtained.

Consent for publication

Informed written consent was obtained for publication.

Availability of data and material

Laboratory reports and necessary clinical data were obtained from Bed Head Ticket.

Competing interests

No conflict of interest

Funding

No funding was received.

Authors' contributions

Study design was carried out by VA. Clinical assessment was carried out by CM and VA. Surgical interventions were carried out by VA. Data collection and manuscript writing was carried out by VA, CDM, SDM, SS, and VA. All authors had equally contributed to the paper and review the manuscript with needful corrections.

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