



Case Report

A case report of primary gastroesophageal melanoma: Presentation, diagnosis, and surgical approach

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ABSTRACT

Introduction & importance: Gastrointestinal tract is an uncommon site for primary melanoma and its annual incidence is reported 0.47 cases in million. Thus, limited information is available about its medical or surgical treatment, long-term complications of melanoma, and survival rates of each therapeutic method.

Case presentation: A 47-year-old male was admitted to the emergency department with massive rectorrhagia. With not notable medical history except recent episodes of dyspepsia, melena, malaise and weight loss. Melena and weight loss in a 47-year-old patient is considered as suspicious signs for malignancy and should be investigated. The patient was finally diagnosed with primary gastrointestinal melanoma (PGIM). He underwent *trans*-hiatal total esophagectomy and proximal gastrectomy with gastric pull-up and lymph node dissection. Immunotherapy with Interferon- α was chosen as adjuvant therapy for this patient. After 10 months, CT scan of abdomen with intravenous and oral contrast revealed multiple foci in liver and spleen consistent with metastasis without any evidence of recurrence at primary tumor excision site.

Clinical discussion: In this article, we presented a rare case of PGIM with later metastasis in liver and spleen. Gastric pull up was preferred to colon interposition for conduit reconstruction after esophagectomy in this case. However due to the rarity of this category of tumor more information must be gathered on the amount of margin to be resected and long-term outcome of different surgical approaches.

Conclusion: Based on the poor prognosis of PGIM, less invasive surgical procedure which provides the radical resection and adequate onco-surgical dissection should be considered.

1. Introduction

Even though cutaneous melanoma is the most prevalent form of melanoma, gastrointestinal tract may be a very rare origin for this category of malignant tumors. As reported in a study published in 2005, 659 cases were diagnosed with primary gastrointestinal melanoma (PGIM) from 1973 to 2004. Epidemiological studies have shown that 4% of patients diagnosed with cutaneous melanoma have symptomatic GI involvement. GI involvement is also found in 60% of patients after autopsy [1]. Thus when found, GI melanoma is considered metastatic unless no cutaneous source is found despite thorough physical examination [2]. Because of rarity of this subgroup of melanoma, almost all information is based on case reports and there is no consensus on

therapeutic plan of choice. Surgical resection of tumor with lymph node dissection has been identified as the most beneficial treatment modality especially when lymph node involvement is negative; nevertheless, average survival rate is only 19 months [1].

In this article we present a middle-aged man diagnosed with primary gastrointestinal (GI) melanoma located in gastroesophageal junction (GEJ) that primarily presented with massive GI bleeding. Then we discuss about challenges in choosing the most advantageous surgical approach and a brief follow up.

This case report has been reported in line with the SCARE 2020 Criteria [3].

Abbreviations: primary gastrointestinal melanoma, PGIM; computed tomography scan, CT scan; positron emission tomography scan, PET scan.

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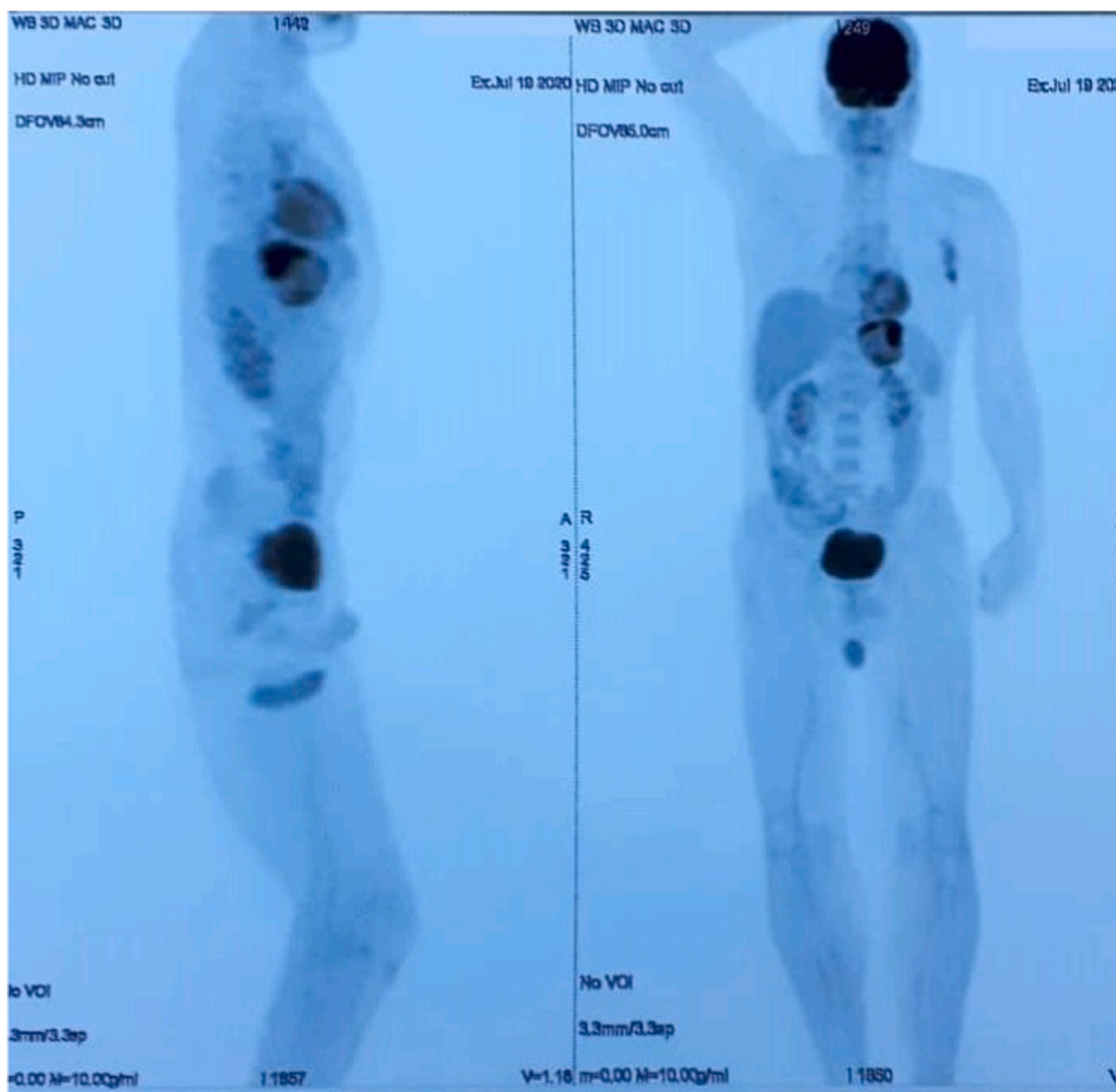


Fig. 1. PET/CT scan demonstrating hyper metabolic primary tumor in distal of esophagus, gastroesophageal junction and cardia with extension to lesser curvature.

2. Case presentation

A 47-year-old male patient presented to the emergency department with chief complaint of massive rectorrhagia. His past medical and family history was not significant except for some prior episodes of dyspepsia, melena, malaise and weight loss, his drug and habitual history was negative, and he did not have any allergic status. Patient was admitted in hemorrhagic shock, tachycardic and hypotensive status. After resuscitation with crystalloid fluid, blood transfusion and emergent endoscopic intervention, bleeding was controlled. The patient needed transfusion of 7 units packed RBC to reach hemodynamic stability. Endoscopy revealed a large pedunculated polypoid lesion with long pedicle in distal third of esophagus and a suspicious subepithelial lesion in gastroesophageal junction with mucosal ulceration. Few biopsies were obtained from mucosal lesions and presence of malignant cells was reported on pathologic investigations. Immunohistochemistry (IHC) study was performed on biopsy specimens and tumor cells were positive for S100 protein, Melanin A (some cells) and HMB-45 (some cells). No Pan-CK, CK5/6 or P63 were detected in tumor. Thus, IHC study was consistent with diagnosis of malignant melanoma. Considering the fact that GI melanoma are most often metastatic lesion from a

primary cutaneous or ocular lesion, thorough physical examination was performed to find probably missed lesions. The only positive findings were left axillary lymphadenopathy and a suspicious skin discoloration at right posterior auricular area. Excisional biopsies of both lesions revealed lymph nodes with benign reactive changes and atypical nevus respectively. Spiral CT scan of chest was normal. Spiral CT scan of the abdomen and pelvis with intravenous and oral contrast showed a 17 × 18 mm hypo dense mass lesion in the distal part of esophagus with luminal narrowing and a 44 × 69 mm hypo dense mass with sharp and regular border in lesser curvature of stomach. Since investigations to this point revealed no primary oculocutaneous source of melanoma, PET/CT scan was performed. PET/CT demonstrated hyper metabolic primary tumor in distal of esophagus, gastroesophageal junction and cardia with extension to lesser curvature (standard uptake value max = up to 9.3) (Fig. 1); with mild hyper-metabolic gastro-hepatic lymph nodes with no evidence of distant metastasis; so the lesions were assumed as primary GI melanoma.

Existence of tumoral lesion that once caused massive GI bleeding and hemodynamic instability, prompted to an interventional plan to prevent rebleeding. For gastrointestinal lesions that are highly susceptible for rebleeding, angioembolization may be performed as therapeutic or



Fig. 2. figure 2A, surgical field, showing *trans*-hiatal esophagectomy and proximal gastrectomy. Distal part of stomach was pulled up for cervical anastomosis. Fig. 2 B resected tumor with dimension of 10×8×4 cm and 5 cm margin.

Table 1

Results of definite post-surgical pathology.

Tumor configuration: elevated polypoid
Tumor size: 10×8×4 cm
Tumor site: esophagus, cardia, fundus, lesser curvature
* Invasive melanoma
*tumor penetrates the subserosal connective tissue without invasion of visceral peritoneum
*mitotic rate: 5 mitosis/mm ²
*lymphovascular invasion: present
* neurotropism: present
*tumor-infiltrative lymphocyte: present, brisk
*all surgical margins are free of tumors
*distance of tumor from the closest (lesser omental) margin: 1.5 cm
Thoracic dissection:
• 4 out of 17dissected nodal lymph nodes involved by tumor
• Extra nodal extension: present
Duodenum, D2 dissection:
• 2 out of 11 lymph nodes are involved by tumor
• Extra nodal extension: present
Omentum resection:
• Congested fibroconnective tissue free from tumor
• 2 dissected reactive lymph nodes free from tumor

palliative procedure based on nature of lesion [4]. In this patient, upper endoscopic therapy with epinephrine and thermocoagulation, controlled first episode of massive bleeding without further rebleeding during work up time. Patient’s age and the fact that the tumors were proven to be primary lesions, guided us to choose surgical resection as therapeutic plan of choice, rather than angioembolization. We assumed two different surgical plans to be suitable for the patient: the first, total esophagectomy and proximal gastrectomy with gastric pull up, and the second, total esophago-gastrectomy with colon interposition. The patient was prepared for both procedures before surgery. Because of COVID-19 pandemic, we preferred minimal thoracic cavity manipulation and cervical anastomosis rather than thoracic one. Hence, *trans*-hiatal esophagectomy and proximal gastrectomy with 5 cm safe margins of tumor site, was performed (Fig. 2A; Fig. 2B). The proximal and distal margins were sent for intraoperative frozen section study which were free of tumor. Therefor without further resection, distal part of stomach was pulled up for cervical anastomosis. Thoracic and D2 lymph node dissection were also performed. Results of definite post-surgical pathology is summarized in Table 1. The operation done by an expert surgeon and the faculty member of the most referral educational hospital. The patient’s general condition was acceptable after surgery, and he was discharged after 5 days. Afterwards, the patient was referred to oncologist for adjuvant chemotherapy, for which he received

subcutaneous interferon (Targeted therapy was not available). On follow up visits, during first few months he gained weight and his general condition improved significantly. Approximately 10 months later, the patient complained of constant epigastric pain. CT scan of abdomen with intravenous and oral contrast revealed multiple foci in liver and spleen consistent with metastasis without any evidence of recurrence at primary tumor excision site. Absence of local recurrence may depict that sufficient margin was resected primarily and delayed metastases are due to some of intrinsic features of tumor such as lympho-vascular invasion.

3. Discussion

Despite cutaneous melanoma, staging system prior to surgery, exact surgical plan and case specific outcomes of existent surgical technique are not well established for primary GI melanomas. On the other hand, American Joint Committee of Cancer staging system for cutaneous melanoma is not applicable on this category of solid tumors [5]. Surgical resection plus lymphadenectomy with median survival rate of 19 months is the therapeutic approach of choice in comparison with non-operative management which results in average survival rate of only 8 months [1]. As in cutaneous melanoma, early lymphadenectomy may help in staging, allows identification of possible lymph node metastasis and necessity of adjuvant treatments [6].

Both gastric pull up and colon interposition can be performed for reconstruction after esophagectomy in case of esophageal cancer. Each method has its own costs and benefits that surgeon must consider them all together to decide which one might be the best for each patient. Broad vascular supply, ease of mobilization and need for single anastomosis, make stomach first choice for esophagus reconstruction when sufficient gastric tissue is available [7,8]. On average 41.8% patients encounter post-operative complication such as reflux, anastomosis leak or stricture, pneumonitis, etc. [9]. On the other hand, when tumors are located in gastroesophageal junction and wide gastric resection is needed, colon with its long length and acid resistance, makes a favorable substitute for esophagus but Long surgery duration, complicated pre-operative preparation and need for 3 anastomoses are some disadvantages [7]. About 64% of patients may suffer from pulmonary complications, anastomosis leakage or stricture and other post-operative morbidities [10]. Peri-operative mortality of gastric pull up is 11% which is lower than colon interposition which is 15.8% [11,12]. Unfortunately, outcomes of these two techniques specifically for gastro-intestinal melanoma are not available in literatures.

For the discussed patient, after frozen section study stated that margins were free of tumor, availability of sufficient gastric tissue for conduit construction, shorter surgical duration and generally lower morbidity and mortality rates, guided surgical team to perform gastric pull up and regional lymph node dissection. Another reason for choosing this technique was coincidence with COVID-19 pandemic that imposed exceptional conditions on the hospital and forced us to perform less complicated procedures with shorter length of stay.

In conclusion, due to the rarity of this category of melanoma; the amount of margin to be resected and effects of some tumoral features such as lympho-vascular invasion on adequate removal and long-term results of surgeries, are not well known and further information must be gathered to standardize procedures.

Ethical approval

Nothing to declare.

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Author contribution

The surgery was performed by 'Narjes Mohammadzadeh' and 'Mohammad Ashouri'. The manuscript was mainly written by 'Neda Nilforoushan' and reviewed and edited by 'Narjes Mohammadzadeh' and 'Mohammad Ashouri'. All authors read and approved final manuscript.

Registration of research studies

1. Name of the registry: N/A.
2. Unique Identifying number or registration ID: N/A.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): N/A.

Guarantor

Mohammad Ashouri.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Declaration of competing interest

All authors state that there are no conflicts of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2022.104195>.

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