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Case Report

Primary Gastric Melanoma: A Rare Diagnostic Entity – A Case Report

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Abstract: *Primary gastric melanoma is an extremely rare malignancy, with the majority of gastrointestinal melanomas being secondary to cutaneous lesions. Its diagnosis is challenging due to nonspecific symptoms and the need for histopathological confirmation.* ***Case Presentation:*** *We report the case of a 55-year-old male presenting with a year-long history of hematemesis, vague chest discomfort, epigastric pain, fatigue, melena, and significant weight loss. Endoscopic examination revealed multiple pigmented polypoid masses in the stomach. Biopsy and immunohistochemistry confirmed malignant melanoma. No cutaneous, ocular, or anorectal primary lesion was identified. The patient underwent total gastrectomy with Roux-en-Y esophagojejunostomy and regional lymphadenectomy.* ***Conclusion:*** *Primary gastric melanoma should be considered in the differential diagnosis of upper gastrointestinal bleeding, mainly when pigmented lesions are observed. Early diagnosis and surgical resection are critical, although prognosis remains poor due to its aggressive behavior and early metastatic potential.*

Keywords: Gastric melanoma, Primary melanoma, Hematemesis, Gastrectomy, Immunohistochemistry

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Introduction

Primary gastric melanoma (PGM) is an exceptionally rare neoplastic entity, representing a mere fraction of all malignant melanomas. The incidence of PGM is notably low, with primary gastrointestinal melanomas occurring at a frequency of approximately 0.5 to 1 case per million individuals, of which PGM accounts for about 12.7% (1). The clinical presentation of PGM is often nonspecific, commonly manifesting symptoms such as abdominal pain, weight loss, melena, and anemia, which can mimic other upper gastrointestinal pathologies (2,3).

Diagnosing PGM poses significant challenges, primarily due to the necessity of excluding other potential sources of melanoma, particularly cutaneous metastases. The diagnostic criteria established for confirming PGM include: 1. the absence of any identified primary melanoma from the skin or other organs; 2. the lack of unexplained skin tumors or extraintestinal melanoma metastases; and 3. no other related intestinal mucosal lesions (4,2). Without adherence to these criteria, the risk of misdiagnosis remains high, as demonstrated in previous case reports where PGM was initially mistaken for metastatic disease or other malignancies (3).

Moreover, the prognosis for PGM is notably poor, exhibiting a five-year survival rate that is significantly lower than that of cutaneous melanoma, largely attributed to late-stage presentation at diagnosis (3-5). The specificity of characteristics seen in PGM, such as the presence of distinct mucosal lesions, adds another layer of complexity to diagnosis, necessitating careful pathological examination to differentiate between primary and metastatic lesions (2,6). This case report underscores the importance of heightened awareness and clinical vigilance regarding PGM to facilitate timely diagnosis and management.

In light of its rarity, limited literature exists regarding PGM, highlighting the need for increased reporting of such cases. Each instance serves to enhance our understanding of this unique melanoma variant, contributing to better clinical recognition and improved outcomes for affected patients. The significance of this case report lies in its potential to add to the scant body of knowledge surrounding primary gastric melanoma and to guide future research and clinical practices.

Case Presentation

A 55-year-old male presented with a one-year history of intermittent hematemesis, progressive fatigue, exertional dyspnea, generalized body aches, and significant weight loss of approximately 10 kg. He also reported vague retrosternal and epigastric discomfort, heartburn, and lethargy. The patient had sought medical care at multiple centers due to recurrent anemia, requiring frequent blood transfusions. His past medical history was unremarkable, and there was no history of cutaneous lesions, prior malignancies, or gastrointestinal surgeries.

Upper gastrointestinal endoscopy revealed multiple soft, friable, and easily bleeding polypoid lesions along both the greater and lesser curvatures of the stomach. The esophagus appeared normal (Figure 1). Contrast-enhanced computed tomography (CT) of the chest and abdomen demonstrated marked circumferential and asymmetric wall thickening involving the fundus and body of the stomach, predominantly on the posterior wall, extending distally to the antro-pyloric region. Multiple enlarged perigastric and porta hepatis lymph nodes were also observed, along with minimal ascites (Figure 2).

Histopathological analysis of endoscopic biopsies revealed a malignant neoplasm composed of large, atypical tumor cells with hyperchromatic nuclei, a high nuclear-to-cytoplasmic ratio, and abundant melanin pigment within the cytoplasm and extracellular spaces. Immunohistochemical staining demonstrated diffuse positivity for HMB-45 and SOX10, confirming the diagnosis of malignant melanoma involving the gastric mucosa.

A comprehensive dermatological examination revealed no suspicious cutaneous lesions, and the patient denied any prior removal of pigmented skin lesions. Ophthalmologic evaluation, including fundoscopy, showed no evidence of ocular melanoma. Proctoscopic assessment was also unremarkable.

In the absence of a cutaneous, ocular, or anorectal primary, the diagnosis of primary gastric melanoma was established. The patient subsequently underwent exploratory laparotomy, total gastrectomy with Roux-en-Y esophagojejunostomy and jejunojejunostomy, and regional

lymphadenectomy involving nodes at the celiac axis, splenic hilum, porta hepatis, and body of the pancreas.

Gross examination of the resected specimen demonstrated large, ulcerated, pigmented masses involving both the lesser and greater curvatures of the stomach (Figure 3). Microscopic evaluation of the specimen revealed sheets of pleomorphic, plump tumor cells with abundant eosinophilic cytoplasm, coarse chromatin, prominent nucleoli, and black-brown intracellular pigment consistent with melanin. Occasional mitotic figures were noted. All surgical margins were free of tumor infiltration.

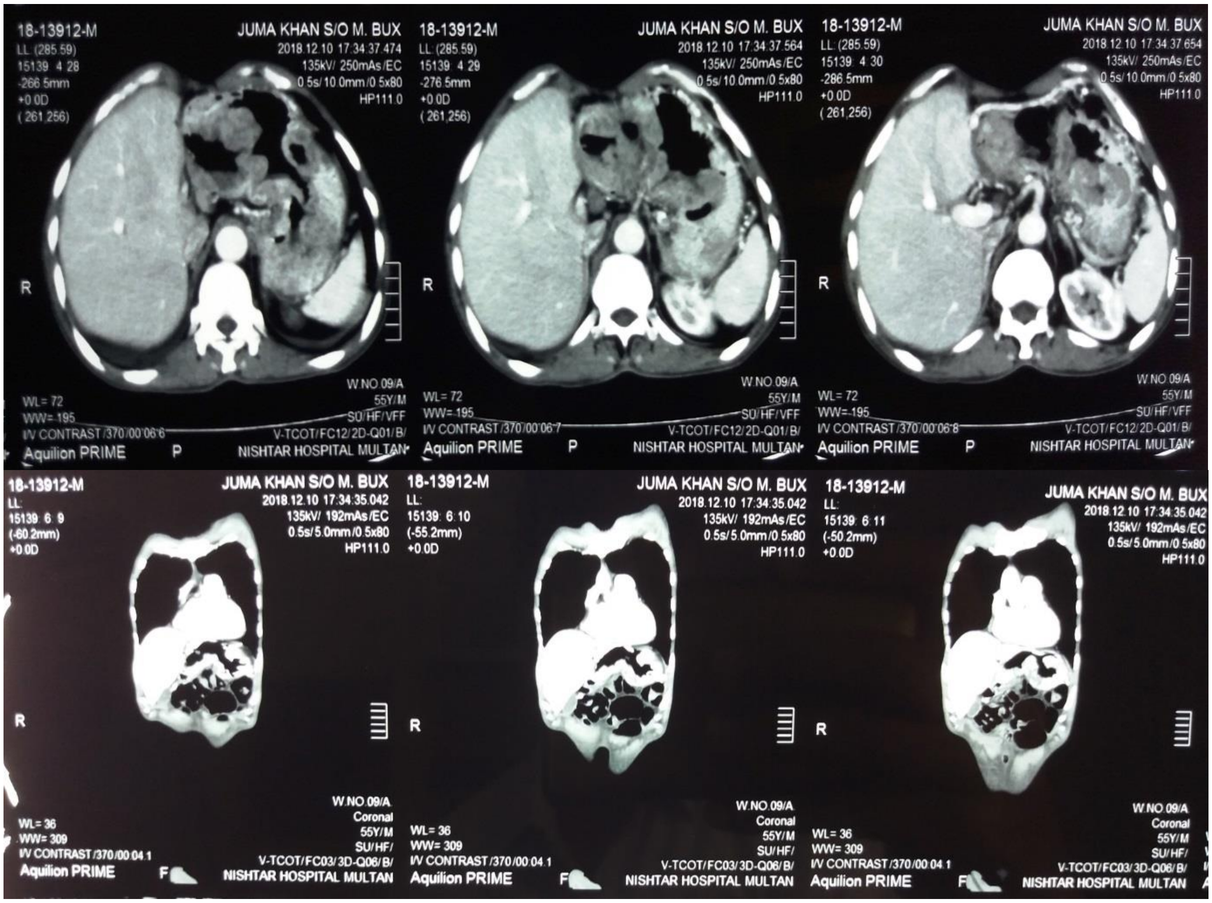

Figure 1: *Axial contrast-enhanced CT images of the abdomen demonstrate significant circumferential and asymmetric wall thickening involving the fundus and body of the stomach, predominantly along the posterior wall, extending toward the antrum. Multiple enlarged perigastric and porta hepatis lymph nodes are visible, along with minimal ascites. These findings are suggestive of an aggressive infiltrative neoplasm.*

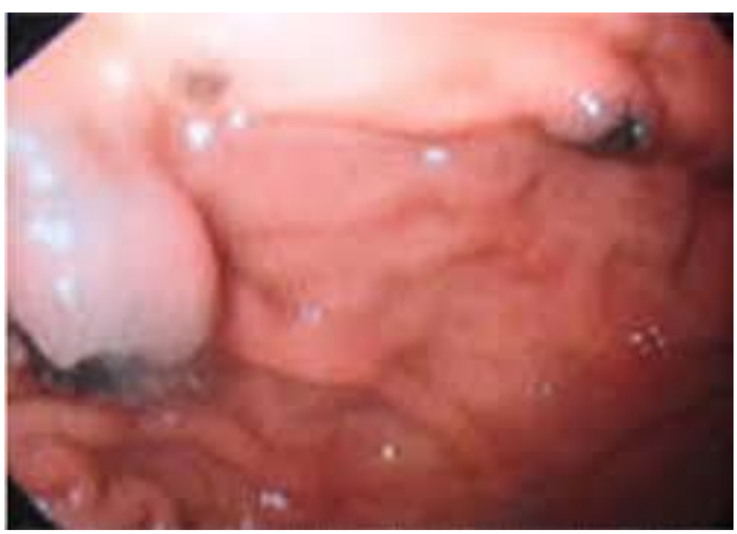

Figure 2: *Upper gastrointestinal endoscopic image showing multiple polypoid, friable, and pigmented lesions involving the gastric mucosa along the greater and lesser curvature. The lesions appeared soft and bled easily on contact, raising suspicion of a malignant process. These findings prompted biopsy and further histopathological evaluation.*

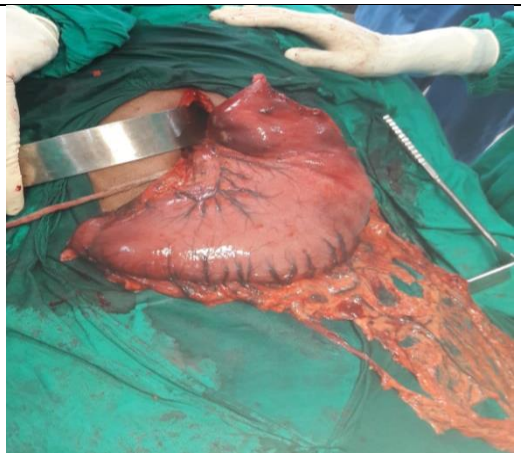

Figure 3: *Intraoperative image showing the gross appearance of the stomach during exploratory laparotomy. The specimen reveals a distended stomach with multiple pigmented, ulcerated, and nodular lesions involving the greater and lesser curvature. Total gastrectomy with Roux-en-Y esophagojejunostomy and extensive lymphadenectomy was performed for complete tumor excision.*

Discussion

Malignant melanoma, primarily recognized for its cutaneous manifestations, can also present in the gastrointestinal tract, particularly in the stomach. While most gastric melanomas are metastatic lesions stemming from a primary site, primary gastric melanoma is a rare entity with specific diagnostic criteria. It has been historically asserted that melanoma could not originate in the stomach due to the traditionally believed absence of melanocytes within the normal gastric wall (7). However, recent evidence indicates that primary gastric melanomas may occur and are characterized by specific criteria: the absence of concurrent melanoma lesions, no prior history of melanoma, and a disease-free interval of at least 12 months following the curative resection of the gastric tumor (8).

The clinical manifestations of primary gastric melanoma often mirror those of other gastric malignancies, with frequent symptoms including weight loss, upper gastrointestinal bleeding, and anemia. Dysphagia has been reported in a subset of patients, but prevalence rates can vary in literature (9,10). Early diagnosis remains challenged; imaging techniques such as computed tomography (CT) and upper gastrointestinal endoscopy are critical for detecting these lesions, which may present as mass-like structures with pigmentation (7,11). Additionally, immunohistochemical staining using markers such as SOX10, S-100, and HMB-45 significantly enhances diagnostic sensitivity, aiding in distinguishing primary gastric melanoma from other gastric neoplasms (12-14).

The prognosis for patients diagnosed with primary gastric melanoma is generally poor due to the typically advanced stage at the time of diagnosis and the aggressive nature of the disease, with many patients experiencing metastasis shortly after diagnosis (15,16). The overall survival rate for primary gastric melanoma is low, with reports indicating 5-year survival rates around 25% (7), highlighting the critical need for early detection and intervention to improve long-term outcomes (10,14).

Distinguishing between metastatic gastric melanoma and primary gastric melanoma is vital, especially considering that metastasis from cutaneous melanoma commonly affects the stomach. Research indicates that gastric metastases are often identified within a year following the diagnosis of the primary melanoma, underscoring the necessity for thorough assessment in patients with known melanoma histories (17,18). Therefore, comprehensive diagnostic criteria are essential for accurately classifying these lesions to ensure appropriate management (19).

Thus, primary malignant melanoma of the stomach is a rare but clinically significant entity that necessitates enhanced awareness for early detection and treatment. The strict diagnostic criteria help differentiate primary lesions from metastases, and the application of immunohistochemical staining is crucial for accurate diagnosis. Timely surgical intervention remains the primary hope for extending survival, despite the overall poor prognosis associated with this condition.

Conclusion

Primary gastric melanoma, though exceedingly rare, does occur and should be considered in the differential diagnosis of patients presenting with vague upper abdominal pain, nausea, hematemesis, melena, fatigue, anemia, or unexplained weight loss. The presence of a pigmented (black) lesion on upper gastrointestinal endoscopy should raise clinical suspicion and prompt a biopsy with immunohistochemical analysis. A diagnosis of primary gastric melanoma should only be made after thorough dermatological and ophthalmological examinations to exclude more common primary sites such as the skin and retina. We report a rare case of malignant melanoma originating in the stomach and emphasize that clinicians must remain vigilant of this entity. Although surgical resection and primary gastrointestinal reconstruction may provide palliation, the long-term prognosis remains poor due to the aggressive nature of the disease and its tendency for early metastasis, compounded by the challenges in early detection.

Declarations

Data Availability statement
All data generated or analysed during the study are included in the manuscript.

Ethics approval and consent to participate
Approved by the department concerned. (IRBEC-MMS-033-24)

Consent for publication
Approved

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Author Contribution

MR (Consultant)
Manuscript drafting, Study Design,
AT (Consultant)
Review of Literature, Data entry, Data analysis, and drafting articles.
AR (Assistant Professor)
Conception of Study, Development of Research Methodology Design,

All authors reviewed the results and approved the final version of the manuscript. They are also accountable for the integrity of the study.

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