

Case Report: Primary Malignant Melanoma of the Liver, A Rare Entity

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Abstract

Case Report

Background: Primary hepatic malignant melanoma is a rare clinical entity, with few reported cases and an unclear pathogenesis. Diagnosis remains challenging due to its nonspecific clinical presentation and radiological appearance that can mimic other malignant hepatic tumors. **Case Presentation:** We report a case of a 61-year-old North African male presenting with a 4-month history of abdominal pain, anorexia, and weight loss. Imaging revealed two hepatic lesions in segments V–VIII and III, characterized by heterogeneous enhancement and necrosis. Liver function tests and tumor markers [AFP, CEA, CA19-9] were within normal limits. Histological evaluation of core biopsies identified malignant cells with abundant intracytoplasmic melanin pigment. Immunohistochemistry demonstrated strong expression of MelanA, confirming melanoma. A comprehensive clinical, radiological, and laboratory workup excluded any other primary site, leading to the final diagnosis of primary hepatic melanoma. The patient was referred for systemic chemotherapy and continues to be followed. **Conclusion:** Primary hepatic melanoma is an exceptionally rare malignancy with no established treatment guidelines. Accurate diagnosis requires thorough exclusion of other primary sites and relies heavily on histopathological and immunohistochemical findings. Increased awareness is crucial for timely recognition and management of this rare tumor.

Keywords: Primary hepatic melanoma, Malignant melanoma, Immunohistochemistry, MelanA, Liver neoplasms.

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INTRODUCTION

Malignant melanoma occurs most frequently in skin but also in many organs and tissues of the body. However, primary hepatic malignant melanoma is exceedingly rare with poor survival outcomes ranging from months to few years. Diagnosis of primary hepatic melanoma based on clinical and radiological features is difficult due to its nonspecific characteristics. Microscopically, it may be easily misdiagnosed because of morphological heterogeneity and hypomelanotic appearance. The present case represents the only case encountered in our department. 1, 2, 3

CASE REPORT

We report the case of a 61-year-old North African man admitted in the department of gastroenterology with the complaint of pain in the abdomen for 4 months associated with the loss of appetite and subsequent weight loss.

The liver function tests were within normal limits. Serum tumor markers of AFP, CEA, and CA19-9 were within normal ranges. An initial abdominal ultrasound demonstrated a heterogeneous, iso-echoic hepatic lesion in segments VI and VII, exhibiting significant vascularity on color Doppler imaging. The remaining hepatic parenchyma appeared homogeneous, with a normal size liver and regular contours. No additional abdominal abnormalities were identified, including the absence of deep lymphadenopathy, deep vein thrombosis, ascites, or any abdominal masses.

A subsequent contrast-enhanced computed tomography of the abdomen was done showing a mass spanning segments V, VI, and VIII, fairly well-defined, with lobulated contours, measuring 77 x 81 x 70 mm [anteroposterior x transverse x craniocaudal]. It was isodense on non-contrast images and contains coarse peripheral calcifications. After contrast injection, it demonstrated heterogeneous enhancement, outlining central areas of necrosis. A second lesion was identified in segment III, measuring 10 x 9 mm, fairly well-defined, with regular contours, spontaneously hypodense, and

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displaying mild heterogeneous enhancement after contrast injection.

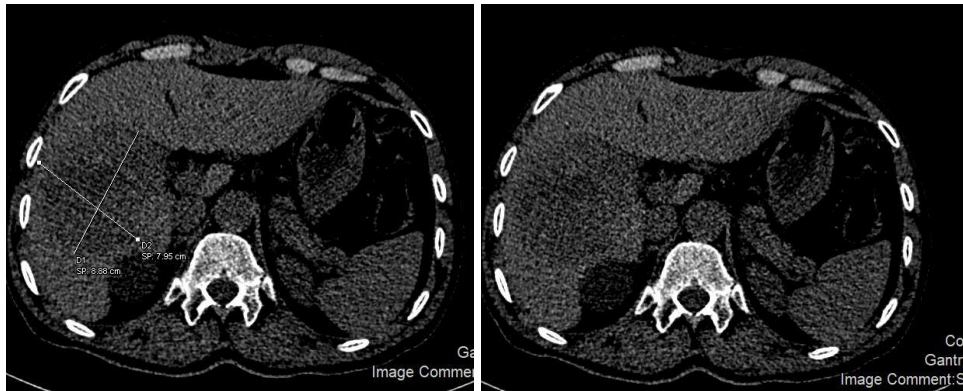


Figure 1: Abdominal SC scan images

An ultrasound-guided liver biopsy was done and histopathological evaluation identified a malignant tumor with abundant black intracytoplasmic pigment. An immunohistochemical analysis was conducted for further characterization, utilizing the immunoperoxidase

technique [Automated method: ONINIS station, Agilent]. The results revealed strong and diffuse cytoplasmic expression of tumor cells with the anti-MelanA antibody [Clone A103, IgG1 Kappa], confirming the diagnosis of melanoma.

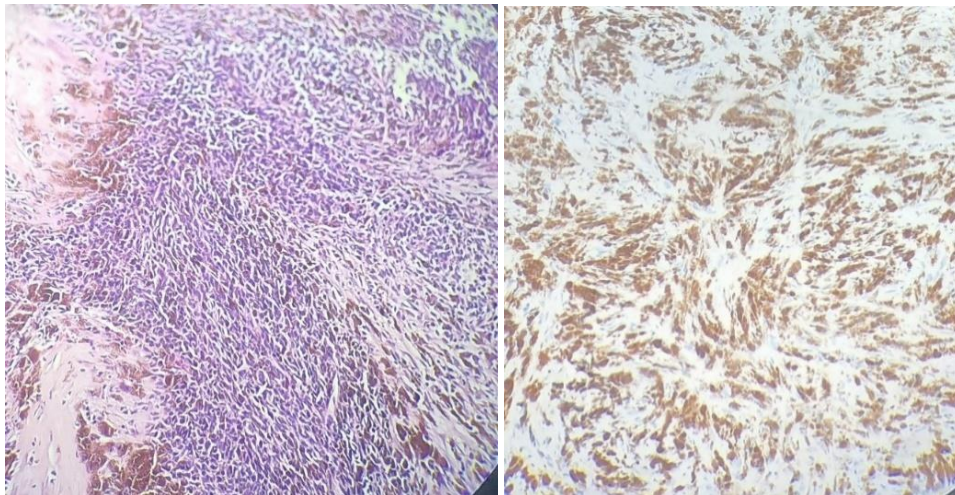


Figure 2: 2A, histopathological exam showing an infiltrative tumor proliferation with a diffuse architecture. The tumor cells are sometimes round, sometimes spindle-shaped. They have anisocaryotic, hyperchromatic nuclei, occasionally nucleolated, with irregular contours. Abnormal mitoses are present. The cytoplasm is eosinophilic, with areas containing melanin pigments. The stroma is fine and fibrous and with large areas of tumor necrosis.

2B, immunohistochemical complement of the biopsies showing an intense cytoplasmic expression with Anti-Melan A antibody

The patient then underwent a full workup with a detailed dermatological examination, ophthalmological examination, a thoraco-abdominopelvic cross-sectional imaging that didn't objectify any other possible site of a primary melanoma.

Thus, the case was then presented in a multidisciplinary meeting with oncologists, pathologists, gastroenterologists and radiologists and a final diagnosis of primary hepatic melanoma with hepatic metastatic lesion was thus retained. The patient was then addressed to the oncology department of our institution to start his chemotherapy, and was regularly followed up since then.

DISCUSSION

Primary hepatic melanoma is extremely uncommon and has been rarely reported. In this case report, we discuss the clinicopathological features of primary hepatic melanoma so as to increase the awareness and improve our understanding of the disease.

Malignant melanoma is a cancer of melanocytes, usually in skin. It can be highly aggressive, with as many as 20% of patients developing metastases [4].

Such melanomas of unknown primary site [MUP] are estimated to comprise between 3.7% and 6% of all incident melanomas [6-5].

Diagnostic criteria for MUP initially were proposed in 1963 by Dasgupta *et al.*, [6]. Contemporary criteria for the diagnosis include the following: 1] metastatic melanoma confirmed clinically, histologically, and immunohistochemically; 2] the absence of a previous cutaneous tumor, pigmented or not, destroyed or excised without histologic examination; and 3] exclusion of unusual primary sites, including urogenital, otolaryngologic, or ophthalmologic sites.

The origin of mucosal malignant melanoma is unclear. Most experts hold that malignant melanomas of non-sun-exposed tissues are linked to stimulation by the blood-borne sunlight circulation factor, expressed in sun-exposed melanoblasts [7]. Based on the origin of primary melanomas of the esophagus and stomach, some authors believe that these alimentary tract neoplasms originate from migrating melanocytes invaginated by digestive tract epithelial cells during embryogenesis [8], which is supported by the distribution of melanocytes in other typically-occurring mucosal cells of the rectum [8]. However, the origin and pathogenesis of primary melanomas arising in parenchymal organs are still unclear.

It is difficult to show the clinical characteristics of primary hepatic malignant melanoma. Pathologically, hepatic malignant melanoma resembles that of the skin or mucosa, exhibiting morphologic variability within the tumor sample. Microscopically, the tumor mass is comprised of epithelioid cells arranged in nests, or spindle cells arranged in fascicles, with or without melanin pigment deposition. Mitotic figures are readily apparent [9].

Given the rarity of this tumor, the optimal therapeutic regimen is not known. In fact, the natural history of these cases also remains unclear. Since surgical therapy is usually palliative, a more aggressive oncologic regimen consisting of chemotherapy, immunotherapy and radiotherapy may be required. With the natural history largely unknown, it is necessary to find treatment modalities for other deep primary melanomas.

CONCLUSION

Primary hepatic malignant melanoma is an exceptionally rare tumor. Due to its nonspecific clinical presentation and the absence of characteristic imaging features, definitive diagnosis relies on histopathological examination and immunohistochemical confirmation, coupled with the exclusion of other primary sites. This case underscores the importance of maintaining a high index of suspicion in atypical hepatic lesions and highlights the critical role of a multidisciplinary approach in its management due to absence of established therapeutic guidelines. Given the rarity and aggressive nature of the disease, further case reports and clinical studies are essential to enhance our understanding and guide future therapeutic strategies.

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