

Primary malignant melanoma at esophagogastric junction

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ABSTRACT

A 63-year-old-man was admitted to the hospital with two months history of abdominal pain, anorexia and weight loss. The endoscopy demonstrated a 2 cm in size, dark blue, pediculated and polypoid tumor almost under the esophagogastric junction with a diagnosis of primary malignant melanoma of the esophagus (PMME). Patient died of progression of liver metastases 2 months after the diagnosis. Clinical and biological characteristics of this rare tumor with its management opportunities are also reviewed in the light of pertinent literature. [Turk J Cancer 2006;36(4):182-184].

KEY WORDS:

Malignant melanoma, esophagus, chemotherapy

INTRODUCTION

Only 4-5% of all malignant melanomas originate from primarily non-cutaneous sites. They commonly originate from the mucous membranes or in the eyes (1). Primary malignant melanoma in the gastrointestinal tract is mostly originated from the esophagus and from an anorectal lesion (2). However, primary malignant melanoma of the esophagus (PMME) accounts for less than 0.1-0.2% of all malignant tumors in this organ, and is an aggressive tumor with very poor survival. Because its clinical findings are usually similar to other neoplasms in the esophagus, and histopathologic examination is needed to confirm definitive diagnosis (3). We report herein a patient diagnosed as PMME without evidence of a cutaneous and/or ocular origin. We discussed why the melanoma in our case was considered primary tumor.

CASE REPORT

A 63-year-old-man was admitted to the hospital with two months history of abdominal pain, anorexia and weight loss (5 kg in 2 months). The endoscopy demonstrated a 2 cm in size, dark blue, pediculated and polypoid tumor almost under the esophagogastric junction (Figure 1). Multiple endoscopic biopsies of the polyp were

obtained. Histopathological features were consistent with malignant melanoma. Fontana Mason (Figure 2), and Melan A staining (Figure 3) showed a positive reaction in the tumor. Notable pigmented lesions of the skin, the anal region and both eyes were not found in physical examination, and a diagnosis of PMME was made. Computerized tomography (CT) showed multiple liver metastasis with no evidence of pulmonary metastases. The patient received temozolomide 150 mg/m²/day for 5 successive days orally in every 28-day. Twenty days after the last day of chemotherapy, the patient was admitted to the hospital with abdominal pain, vomiting and jaundice. His serum conjugated and unconjugated bilirubin concentration (cBI/uBI), gamma-glutamyltranspeptidase activity (GGT), alkaline phosphatase (ALP), aspartate aminotransferase (AST), and alanine aminotransferase (ALT) were all elevated (cBI: 14 mg/dl, uBI: 10 mg/dl, GGT: 381 U/L, ALP: 395 U/L, AST: 396 U/L, ALT: 210 U/L). Ultrasonography (USG) suggested progression of liver metastases. Hence, conservative management was initiated. His clinical condition continued to deteriorate despite these efforts, and he died on the 10th day of hospital stay.



Fig 1. Endoscopic view of malignant melanoma polyp

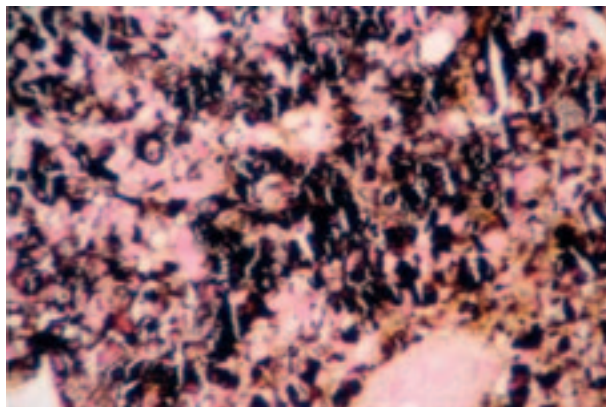


Fig 2. Positive Masson Fontana staining of tumor cells (Masson Fontana stain, x200)

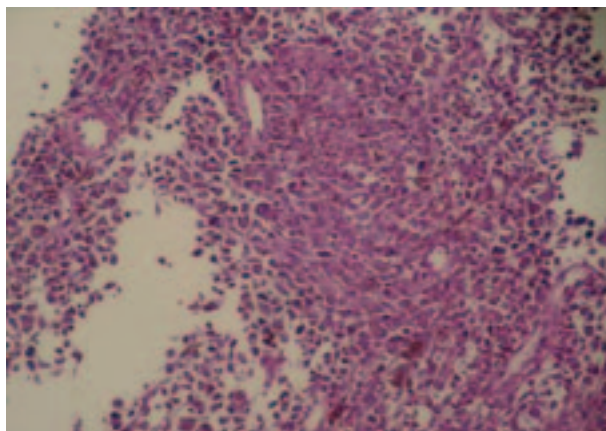


Fig 3. On Melan-A immunostaining of specimen, the tumor cells are positive for Melan-A (Melan A stain; x400)

DISCUSSION

Malignant melanoma of the gastrointestinal tract are more aggressive than cutaneous melanoma (1). The prognosis is usually poor even after a radical treatment due to the aggressive biological behavior of tumor with a high incidence of local failure and advanced stage at the time of presentation. The esophagus usually lacks benign melanocytes, and the occurrence of PMME was controversial for many years. With the documentation of benign melanocytes in normal esophagus, the existence of this tumor is accepted, albeit as an extremely rare tumor. Approximately 200 cases have been published in the world literature since its first description by Baur (4) in 1906. It should be differentiated from other

esophageal tumors and also mandates a thorough investigation to rule out the possibility of being a metastasis from another more common primary site melanoma. Patients affected by this tumor are usually older with a male predominance (5). In our patient, a thorough investigation did not reveal a primary lesion in any other sites. As in our case, the duration of symptoms is generally short before diagnosis. The characteristic finding at endoscopy is a polypoid irregularly pigmented tumor (6). It may be found in all areas of the esophagus, but mostly located in the lower two-thirds of the esophagus (7). In our case the tumor was located at the esophago-gastric junction, and abdominal CT and USG showed liver metastases. Hematogenous and lymphatic metastases are common (7,8). Approximately 50% of patients with PMME present with metastatic disease. The most com-

mon sites for metastasis are the liver, mediastinum and lung (7). The mean survival time of the majority of patients has been less than 15 months due to metastases to vital organs (5,6,8).

The ideal management of PMME is surgical resection, even for patients with recurrence. Different adjuvant therapies such as chemotherapy, radiotherapy and immune modulation have not been proven to be of benefit; however, they are recommended for palliative treatment if surgery is not considered (8-10).

In conclusion, PPME is a very uncommon, aggressive malignant tumor arising from esophagus that have a long natural history characterized by frequent vital organ metastasis. This case report emphasizes the poor prognosis of patients with esophageal melanoma.

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