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A Rare Primary Malignant Melanoma of the Esophagus with Hemorrhage: A Case Report

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Abstract

Primary malignant melanoma of the esophagus (PMME) is a highly aggressive tumor with a poor prognosis. Here we describe the case of an 87-year-old man who presented with poor appetite, dysphagia, hematemesis, and weight loss. He had a history of primary malignant melanoma of the esophagus 5 years ago. PMME with hemorrhage and anemia were diagnosed by the esophagogastroduodenoscopy (EGD). After hemostatic medications, the hemorrhage in the esophagus was successfully controlled. The patient refused further surgery or radiotherapy, and he chose palliative care. This article reported a rare case with PMME with hemorrhage, and the findings of the morphological changes of the PMME from a flat-type melanoma to a bulging tumor during a 5-year period are extremely rare. (J Intern Med Taiwan 2021; 32: 297-300)

Key Words: Primary malignant melanoma of the esophagus, Esophageal hemorrhage, Treatment, Prognosis

Introduction

Primary malignant melanoma of the esophagus (PMME) is a rare and aggressive tumor. PMME accounts for 0.1–0.3% of all esophageal tumors and is prevalent in male and older adults with a mean age of onset of 60.5 years[1,2]. Patients with PMME are usually diagnosed in the late stage (LS), and the incidence of metastasis at initial diagnosis is about 40–80%[3]. The 5-year survival rate after surgery was around 4–37%, and overall survival varied between 9–18 months[2,3]. Esophageal melanocytosis is viewed as the precursor for PMME, resulting from chronic esophagitis or proliferated melanocytes in the basement membrane or squamous epithelium of esophagus[1,3,4]. Melanocytes migrate from the neural crest to the epithelium of the skin, meninges, nasopharynx, inner ear, and oral cavity during the early embryonic process[3,4]. The presence of melanocytes rarely exist in the normal esophagus, and 8% of subjects with esophageal melanocytes have been reported by Tateshi et al[5]. Yokoyama et al. discovered older age, smoking, and alcohol use were associated with esophageal melanosis, esophageal dysplasia, and squamous cell carcinoma of the upper aerodigestive tract[6]. In this article, a case with a flat-

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type melanoma of the esophagus progressing to a bulging melanoma with hemorrhage was reported after a 5-year follow-up.

Case report

An 87-year-old man with hypertension and alcohol consumption presented with a 3-day history of hematemesis, and a 6-month history of poor appetite, dysphagia, and weight loss of 20 kg. He had a history of melanoma of the esophagus, which was diagnosed by biopsies from an esophagogastroduodenoscopy (EGD) in a medical health check-up 5 years ago. The morphology revealed a flat and blackish-pigmented lesion in the lower third of the esophagus situated 33 cm from incisor teeth (Fig. 1). Histopathological analysis revealed atypical melanocytic cells in the basal layer of the esophageal squamous tissue. The immunohistochemical stainings were positive for Melan-A, HMB-45, and increased proliferative index of Ki-67. Positron-emission tomography (PET) showed no detection of distant metastasis. The clinical stage according to the 7th edition of American Joint Committee on Cancer (AJCC) staging manual was cT1N0M0, stage I. The patient refused surgical treatment.

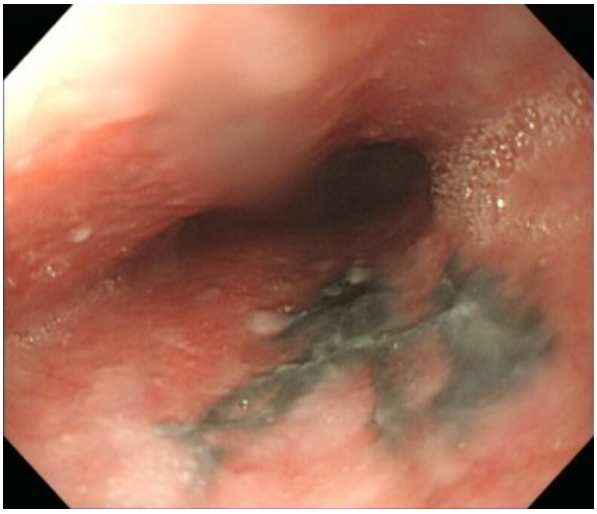

Figure 1. Gastroscopy showed flat, extended blackish-pigmented lesions of the esophagus 33 cm from incisor teeth.

On admission, physical examination revealed pale conjunctiva and cachexia appearance. Laboratory examinations showed a hemoglobin level of 9.5 g/dL and a C- reactive protein level of 1.93 mg/dL. Serum AFP, CEA, CA 19-9, and CYFRA21 were within normal values. Repeat EGD revealed a bulging esophageal mass with necrotic tissue, blackish-pigmentation, and active hemorrhage on the surface 33 cm from incisor teeth (Fig. 2). Chest CT showed a

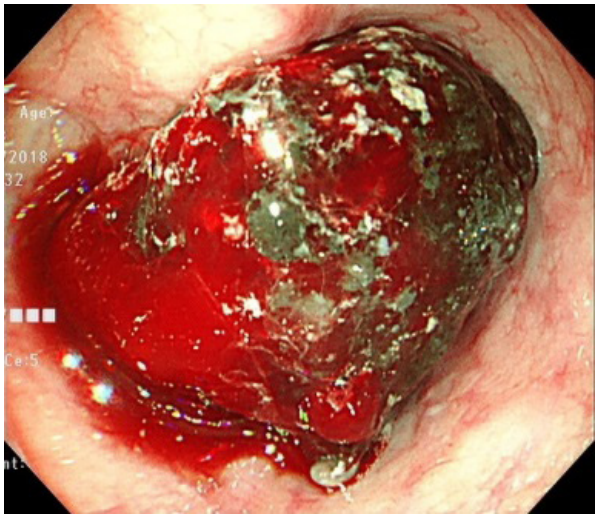

Figure 2. Gastroscopy showed a bulging esophageal mass with necrotic tissues, necrosis, and blackish-pigmentation, hemorrhage on the surface situated 33 cm from incisor teeth.

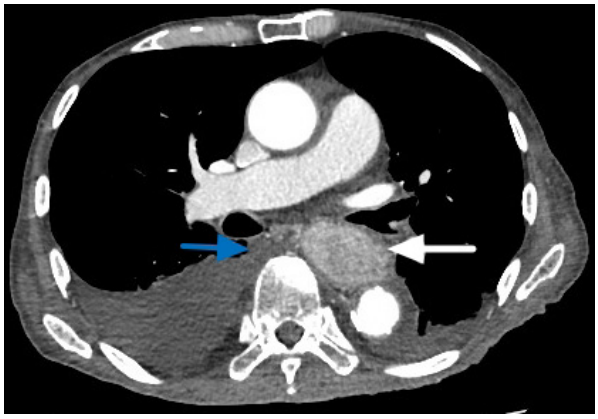

Figure 3. Contrast-enhanced chest computed tomography (axial view, white arrow) showed a tumor measuring 9.8 × 4.1 × 2.1 cm in the middle to lower thoracic esophagus with enlarged paraesophageal and subcarinal lymph nodes (blue arrow).

heterogeneous mass measuring 9.8 × 4.1 × 2.1 cm in the middle to lower thoracic esophagus with paraesophageal and subcarinal lymph nodes (LN) metastasis (Fig. 3). TNM stage based on the 8th edition of AJCC staging system was rcT3N2M0, stage III. The patient was treated with intravenous hemostatic medications for the hemorrhage and peripheral parenteral nutrition for 5 days. He refused surgical resection or radiotherapy and opted for palliative treatment. After successful control of the esophageal hemorrhage, he was discharged after 2 weeks.

Discussion

Primary malignant melanoma of the esophagus is a highly aggressive neoplasm with a dismal prognosis and widespread metastases at initial diagnosis[1-3]. PMME is mostly located in the lower third than the upper or middle third of the esophagus[1-3]. Despite the uncertainty of the natural course and etiology of PMME, the mean duration of detected esophageal metastasis from primary origin varies from 11 to 62 months[3,7]. Endoscopic findings of PMME include irregular, flat, rough, pedunculated, sessile, pink, pigmented or non-pigmented polypoid appearance, friable, eroded, and easily superficial bleeding features[2-4]. The diagnosis of PMME is based on the histopathological and immunohistochemical results. Histopathology of melanoma consists of melanin granules, spindle cells, and pigmented epithelioid[3,4,7]. Immunohistochemically, melanocytes are positive for S-100 protein, neuron-specific enolase, HMB-45, and Melan-A[3,4,7]. Clinical presentations of PMME individuals include dysphagia, retrosternal pain, epigastralgia, weight loss, hematemesis, and melena.[1,3] Treatment strategies for PMME remain uncertain, especially for non-aggressive esophageal melanoma[1-3]. Radical resection might be beneficial for resectable PMME at early stage[2,3]. Additionally, esophagectomy for PMME individuals without lymph node involvement reached a higher disease-free survival than those with lymph node involvement[8]. Lymph nodes metastases could be an independent prognostic factor for PMME[2,3]. The invasion depth of the esophageal tumor beyond the submucosa layer (T2-T4) is inclined to have lymph node and distant metastasis and poor outcome[2,3]. The efficacy of adjuvant therapy with chemotherapy, radiotherapy, or immunotherapy is unclear for PMME in the advanced stage. Long-term survival among patients with PMME was uncommon. One case with T1aN0M0 PMME survived 17 years after radiotherapy and surgical resection[2]. The 5-year survival of the present case without surgery or radiotherapy might be related to no lymph node involvements and organ metastases at initial diagnosis.

Conclusion

The reported case with a flat-type to a bulging PMME after a 5-year follow-up is extremely rare. Regular endoscopic follow-up might play an important role in a better understanding of the morphological changes of PMME. The comprehensive following of guidelines is critical for early detection and timely treatment to prevent disease progression and improve prognosis among patients with primary malignant melanoma of the esophagus.

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罕見的原發性食道惡性黑色素瘤併出血：病例報告

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摘 要

一位87歲男性五年前因健康檢查診斷原發性食道惡性黑色素瘤(primary malignant melanoma of the esophagus)，病患無接受進一步手術切除、化學或放射治療。主訴體重下降、食慾不佳、吞嚥困難及嘔血，胃鏡發現食道惡性黑色素瘤併出血，胸部電腦斷層檢查此腫瘤侵犯中下段食道合併食道旁及氣管隆凸下淋巴結轉移。該病患經止血藥物及周邊靜脈營養處置後，成功控制出血並出院。原發性食道惡性黑色素瘤為一罕見食道惡性腫瘤，預後不佳且常合併遠處轉移，此病患的食道惡性黑色素瘤於五年期間型態上由不規則扁平狀進展成腫塊狀，引起臨床病徵，定期的胃鏡檢查很重要，初期發現原發性食道惡性黑色素瘤並切除，避免進展成晚期病灶及淋巴轉移對於增加病患預後有助益。