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Case Report

Endoscopic resection as a primary palliative therapy for advanced primary malignant melanoma of the esophagus

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Abstract

Primary malignant melanoma of the esophagus is a rare aggressive malignant tumor with poor prognosis. It usually presents as dysphagia and retrosternal chest pain. Diagnosis is made by endoscopy and biopsy, and staging is done by computed tomography (CT) scan and fluorodeoxyglucose positron emission tomography scan. The mainstay of treatment is usually surgical with curative or palliative intent since radiotherapy and chemotherapy do not improve the outcome. Here, we report a case of 50-year-old female patient who presented with dysphagia. Esophagogastroduodenoscopy was done, which revealed a large black-colored polypoid lesion occluding the entire lumen of the esophagus. Histopathology confirmed it as malignant melanoma. CT of the chest was performed which showed a tumor mass extended into mediastinum abutting aorta and multiple mediastinal lymph nodes suggesting an advanced disease. As surgery could not be performed and radiotherapy and chemotherapy have no role, endoscopic resection of mass within the esophagus was done and the defect in the esophageal wall was closed with the clips. The patient was symptomatically improved following the resection. She expired after 3 months due to disseminated diseases.

Key words Endoscopic resection, esophagus, malignant melanoma

Introduction

Primary malignant melanoma of the esophagus (PMME) is a malignant tumor which occurs in the melanin cells of esophageal mucosal epithelial basal layer. PMME is a rare neoplasm that accounts for 0.1%–0.2% of all primary neoplasms of the esophagus and 0.5% of visceral melanomas.[1]

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Although PMME was first reported in 1906[2] and confirmed with histological evidence in 1952,[3] the diagnosis remained controversial until 1963 when the presence of benign melanocytes within the esophageal mucosa was demonstrated.[4] This observation was confirmed later in two other studies.[5]

Risk factors are not yet defined, but esophageal melanosis has been indicated as predisposing factor.[6] The PMME most often affects patients between the sixth and seventh decade of life. There is a 2.02:1 male predominance. Most patients are symptomatic on presentation, and dysphagia is the most common symptom, followed by weight loss, substernal, or

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epigastric discomfort. The duration of symptoms before presentation is short approximately 3 months in most cases.[7]

A polypoidal, pigmented lesion in the lower two-thirds of the esophagus is the typical endoscopic finding.[8] Less frequently, they are nonpigmented, but melanin can be identified in histological examination in a large proportion of those cases, and the true incidence of amelanotic melanoma of the esophagus is <2%.[9]

Thoracic and abdominal computed tomography (CT), fluorodeoxyglucose positron emission tomography-CT, and endoscopic ultrasonography are useful as staging methods.[10]

Most PMME are diagnosed at advanced stage of the disease.[8] Although there is no formal recommendation, surgery is the preferential method of treatment, but the prognosis remains poor.

Case Report

A 50-year-old female patient presented with progressive dysphagia for 2 months with dysphagia score of 3 at the time of presentation. She had one episode of hematemesis associated with malena and retrosternal chest discomfort and constitutional symptoms. Her Glasgow–Blatchford score was 11 at presentation. The patient was hemodynamically stabilized and was subjected to endoscopy. Esophagogastroduodenoscopy was performed with Optera Video Gastrointestinal Scope GIF-H17 and found to have a large pigmented polypoidal mass at 28 cm from incisors occluding the entire lumen [Figure 1]. It was reported as spontaneous esophageal hematoma or aortoenteric fistula with large hematoma or ulcerated mass with blood clot. CT of the chest with oral contrast was performed, and it revealed a large mass with heterogeneous echogenicity in the esophagus extending into the mediastinum closely abutting the aorta [Figure 2]. Multiple lymph nodes were noted in the mediastinum with a small metastatic deposit in the liver [Figure 3]. After excluding the possibility of hematoma, repeat endoscopy was done. Endoscope was passed gently and found that the mass extended from 28 to 35 cm from incisors. Beyond the mass, few satellite lesions with smooth black surface were noted [Figure 4] giving a clinical impression of malignant melanoma. Biopsy was taken from the mass and satellite lesions and sent for histopathological examination. Microscopic examination revealed multiple tumor cells with intra- and extracellular pigmentation consistent with malignant melanoma [Figure 5]. One section showed that tumor cells arise from junctional complex [Figure 6] which suggests that the tumor arises from esophagus. Immunohistochemical staining confirmed it as malignant melanoma. Workup for primary site of origin did not reveal any other primary focus. As the tumor is in advanced stage with mediastinal involvement, surgery cannot be performed. Malignant melanoma is not responsive to chemotherapy or radiotherapy. Her Eastern Cooperative Oncology Group performance status was 2. To improve her dysphagia, endotherapy was planned as a palliative treatment. Endoscopic resection of polypoidal lesion

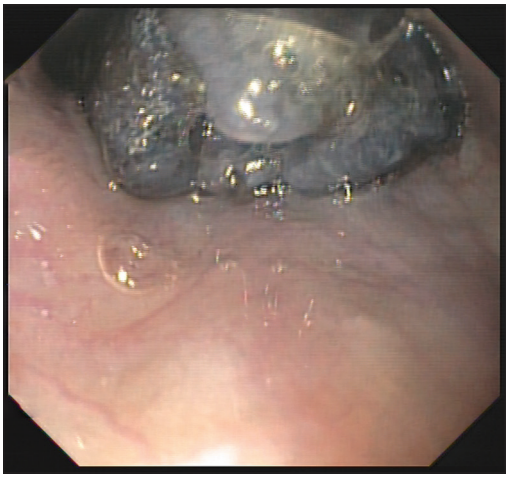

Figure 1: Endoscopy showing large pigmented polypoidal lesion

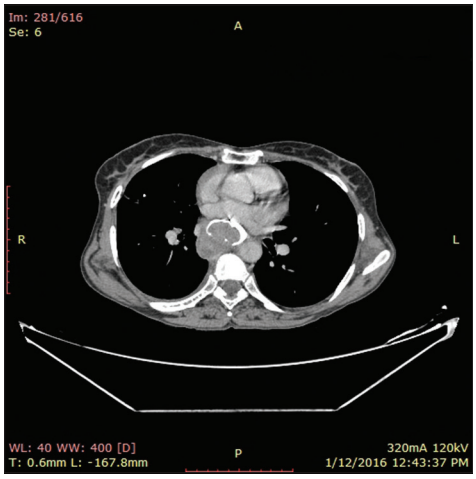

Figure 2: Contrast-enhanced computed tomography showing large heterogeneous mass in the esophagus extending into mediastinum abutting aorta

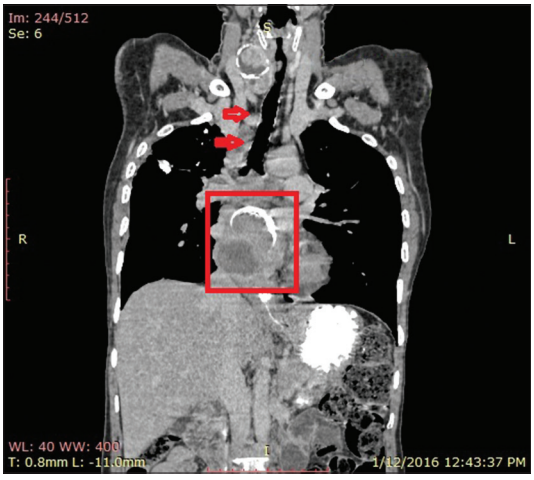

Figure 3: Contrast-enhanced computed tomography showing mass in the esophagus and multiple mediastinal lymph nodes

was done in piecemeal using snare. Oozing of blood from the tumor bed was controlled with argon plasma coagulation. The defect in the esophageal wall was closed using the cook instinct endoscopic hemoclips [Figure 7]. After endoscopic resection, the dysphagia score improved to 1. The patient expired after 3 months due to disseminated disease.

Discussion

We presented a rare case report of PMME and for the first time reported endoscopic palliative resection of the tumor mass.

By the year 2011, only 337 cases had been reported worldwide, most of which being single case reports and most of the cases being males.[11] Here, we report a female patient with malignant melanoma of esophagus.

The majority of PMME patients presents with the complaints of dysphagia, nonspecific retrosternal pain, and weight loss. This patient presented with dysphagia and upper gastrointestinal tract bleeding which is an unusual presentation.

The PMME diagnostic criteria defined in the study by Allen and Spitz[12] include: (i) a typical histological pattern of melanoma and the presence of melanin granules within the tumor cells; (ii) an origin in an area of junctional change within the squamous epithelium; and (iii) junctional activity with melanotic cells in the adjacent epithelium. These criteria, along with the presence of *in situ* melanoma and/or satellite tumors with no previous history of cutaneous melanoma, and a systematic investigation with pathological examination are required for a definitive diagnosis of PMME. All these criteria were fulfilled in the present case.

No gold standard has been established for the treatment of PMME. Kimura *et al*.[13] reported the use of endoscopic mucosal resection for the treatment of early PMME and recorded no recurrence after 18 months. Consistent with this finding, Cheung *et al*.[14] evaluated the surveillance, epidemiology, and end results database of primary gastrointestinal melanomas (1973–2004) and identified complete surgical resection as the only identifiable treatment strategy that significantly improved survival in PMME patients. The present patient has extensive disease which is not possible for resection.

Alternative nonsurgical or surgical adjuvant therapies for PMME patients include chemotherapy, chemoradiotherapy, endocrine therapy, and immunotherapy; however, these treatment strategies have not shown any benefit, and they are

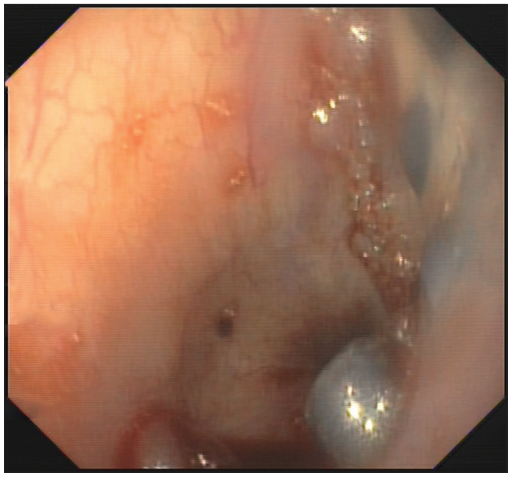

Figure 4: Repeat endoscopy showing satellite lesions beyond the mass

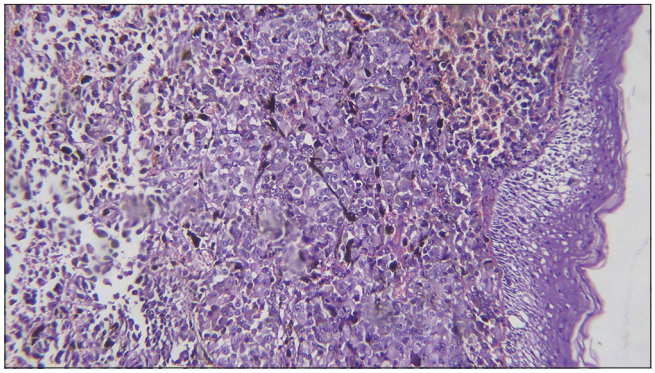

Figure 6: HPE showing junctional activity

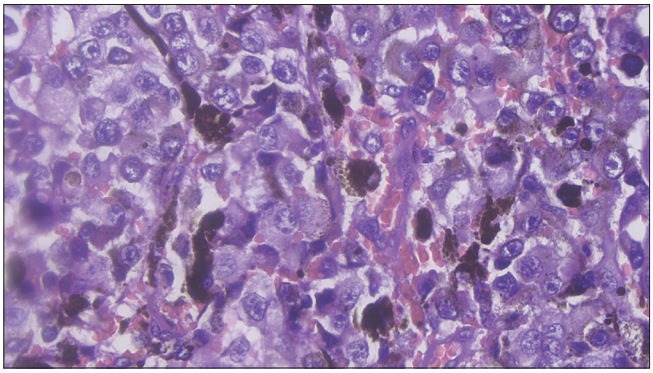

Figure 5: Histopathological Examination (HPE) showing large tumor cells with intracellular and extracellular melanin deposition

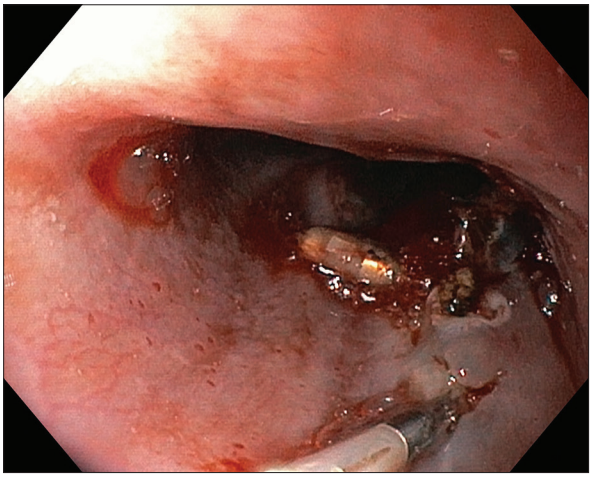

Figure 7: After endoscopic resection, hemoclips were placed

not fully evaluated. Hence, chemoradiotherapy was not given to this patient.

As her performance status was good, we did endoscopic resection to improve her dysphagia as a palliative therapy. After endoscopic resection, her dysphagia scores improved to 1 and went home. She expired after 3 months due to disseminated disease.

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Conflicts of interest
There are no conflicts of interest.

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