A case of early-stage primary malignant melanoma of the esophagus

Tatsuya Mikami, MD, Shinsaku Fukuda, MD, Tadashi Shimoyama, MD, Ryo Yamagata, MD, Daisuke Nishiya, MD, Yoshihiro Sasaki, MD, Yoshiharu Uno, MD, Hiroshi Saito, MD, Shun’ichi Takaya, MD, Yoshimasa Kamata, MD, Akihiro Munakata, MD

Primary malignant melanoma of the esophagus is an extremely rare disease throughout the world. It has a poor prognosis because the tumor is relatively large and has metastasized at the time of diagnosis. This is a case of primary malignant melanoma of the esophagus without evident metastasis detected by chance at mass screening. There has been no local recurrence or distant metastases after esophagectomy.

CASE REPORT

A 41-year-old asymptomatic Japanese woman underwent a barium esophagogram as part of a mass screening program and a polyloid filling defect was detected in the esophagus. Upper endoscopy disclosed a black polyloid lesion in the esophagus and biopsy specimens were interpreted as suggestive of malignant melanoma. She was referred to our hospital for further evaluation 1 month later.

Hematologic and serum biochemical studies including tumor markers and urinalysis at the time of admission were within normal limits. There were no pigmented lesions in the skin, no abnormalities in the eyes, and evidence of a primary malignant melanoma. The patient’s personal medical history and family history were unremarkable.

Endoscopic examination revealed a polyloid, smooth-surfaced tumor about 0.5 cm in size located at 28 cm from the incisors. The appearance of the tumor was mostly black and seemed to consist of normal esophageal mucosa in part (Fig. 1). There was also evidence of melanosis in the surrounding mucosa. No abnormalities were found in the stomach or the duodenum. A barium esophagram was consistent with the esophageal smooth-surfaced polyloid tumor (Fig. 2). No enlarged lymph nodes were detected by CT of the chest and abdomen. Colonoscopy revealed normal mucosa from the rectum to cecum.

Considering these results, a diagnosis was made of primary malignant melanoma of the esophagus without metastatic lesions (T1N0M0, stage 1 in the TNM classification). The patient underwent subtotal esophagectomy by thoracotomy, 3-field lymph node dissection, and cervical anastomosis with stomach substitution by a posterior mediastinal route. The resected specimen contained a tumor 0.8 × 0.6 cm in size. Histologically, the tumor was localized to the mucosa and was diagnosed as a malignant melanoma (Fig. 3A and B), according to the criteria by Allen and Spitz.1 The tumor was also positive for immunohistochemical stains, S-100 protein, and HMB-45, which is specific for melanoma cells (Fig. 3C).2 The presence of esophageal melanosis was also confirmed histologically. There was no intramucosal spread or lymph node metastases microscopically. After obtaining informed consent, the patient was treated with 2 courses of adjuvant systemic chemotherapy consisting of dacarbazine, cisplatin,
and vindesine, which has been shown to have some beneficial effect. At 31 months after diagnosis there was no evidence of recurrence of the tumor.

DISCUSSION

Malignant melanoma usually arises in a cutaneous or ocular site, but rarely can occur in the esophagus, accounting for only 0.1% to 0.2% of all esophageal neoplasms. Although malignant melanoma can metastasize to the esophagus, it was formerly unclear whether the tumor could arise primarily from the esophageal mucosa. However, since the detection of melanocytes in the esophagus, it is generally accepted that malignant melanoma can arise primarily from the esophagus.

Melanosis of the esophagus is thought to be a predisposing factor when this is discovered, the patient has to be followed carefully. It is seen in the one fourth of the patients with esophageal melanoma. In this case, melanosis was also present in the esophagus. Of particular interest was the presence of melanosis in the basal mucosa of the small tumor.

At endoscopy, esophageal malignant melanoma is described as pedunculated, polypoid, and often pigmented. The tumor is usually located in the distal two thirds of the esophagus. Most patients had dysphagia and/or retrosternal discomfort caused by enlargement of the tumor. Esophageal melanomas are usually large in size. The tumor in the present case is the smallest among those reported and the only case in which the tumor was superficial to the muscularis mucosae. The esophageal malignant melanoma in the present case had the characteristic features described above even though it was an early-stage lesion.

Because primary malignant melanoma of the esophagus frequently has distant metastases at the time of diagnosis and even after the resection of the primary tumor, the prognosis is extremely poor. Even when the tumor is limited to submucosa, there are usually lymph node metastases. Furthermore, there was widespread recurrence after operation in a patient without lymph node metastases.
Only one third of patients survive for more than 1 year after the initial diagnosis. If the general condition of the patient is good, surgical resection could be the best choice for treatment inasmuch as adjuvant chemotherapy has not been proved to be effective. Thus the only way to improve the prognosis for patients with esophageal malignant melanoma is to detect the tumor in an early stage without metastasis. Endoscopists should take care not to miss small malignant melanomas especially in patients with esophageal melanosis.

REFERENCES