Flat-type primary malignant melanoma of the esophagus

Case report

A 63-year-old woman was referred to our hospital for further investigation of a gastric mucosal abnormality in an upper gastrointestinal series. Esophagogastroduodenoscopy (EGD) demonstrated two areas of flat, widespread blackish pigmentation situated 30 to 33 cm, and 34 to 38 cm from the incisor teeth (Fig. 1), and no gastric mucosal abnormality. Distinguishing malignant melanoma from diffuse melanocytosis is difficult due to the absence of polypoid morphology. Several parts of these flat lesions were biopsied at random. However, biopsies could not be diagnosed as a malignant melanoma in situ because of the loss of neoplastic proliferation (Fig. 2). Computed tomography demonstrated no abnormal lesion in the esophagus and no enlarged regional lymph nodes. Positron emission tomography-computed tomography (PET-CT) showed no metastases, and a skin survey revealed no cutaneous melanoma. Follow-up examination or surgical resection with a three-stage esophagectomy were discussed; however, the patient wished to proceed with a definite diagnosis of these lesions. The patient underwent endoscopic submucosal dissection (ESD) of these lesions. After the endoscopic resection with a three-stage esophagectomy were confirmed to proceed with a definite diagnosis of these lesions. The patient underwent endoscopic submucosal dissection (ESD) of these lesions. After the endoscopic resection, the lesions were confirmed to be a PMME. Computed tomography demonstrated no abnormal lesion in the esophagus and no enlarged regional lymph nodes. Positron emission tomography-computed tomography (PET-CT) showed no metastases, and a skin survey revealed no cutaneous melanoma. Follow-up examination or surgical resection with a three-stage esophagectomy were discussed; however, the patient wished to proceed with a definite diagnosis of these lesions. The patient underwent endoscopic submucosal dissection (ESD) of these lesions. After the endoscopic resection with a three-stage esophagectomy were confirmed to proceed with a definite diagnosis of these lesions. The patient underwent endoscopic submucosal dissection (ESD) of these lesions. After the endoscopic resection, the lesions were confirmed to be a PMME.

Discussion

Primary malignant melanoma of the esophagus (PMME) is rare and accounts for less than 0.1–0.5% of esophageal malignancies [1]. Overall, 90% of the cases are located in the middle or lower thirds, and 10% in the upper third [2]. PMME is usually single, but multiple lesions have been reported in 12% of cases [3]. The etiology of PMME has not been well investigated because of the rarity of the disease. However, melanocytosis has been indicated as a predisposing factor [4]. Therefore, clinical management of cases, where there is difficulty in distinguishing between early stage PMME and melanocytosis, has not been investigated in detail. In the present case, biopsies could not be diagnosed as a malignant melanoma. Because the patient wished to obtain a definite diagnosis before the optimal treatment of PMME, she underwent endoscopic submucosal dissection (ESD) of these lesions. After the endoscopic resection, the lesions were confirmed to be a PMME. This is the first report of a flat-type PMME to be diagnosed with ESD and treated with radical surgical resection.

The endoscopic characteristic of PMME is a polypoid, irregularly pigmented, obstructive esophageal tumor, which might also be ulcerated [5]. The non-polypoid form, as in the present case, is extremely rare. In the present case, it was also difficult to distinguish PMME from diffuse melanocytosis at EGD because of the absence of the polypoid form. The diagnosis of PMME was confirmed histologically and immunohistochemically by typical cytologic features and the presence of melanin pigment, respectively [6–8]. However, reduced aggressiveness, loss of neoplastic proliferation, and small specimens may not always allow a definite diagnosis of early stage PMME [9, 10]. Therefore, in patients with a small lesion, endoscopic mucosal resection (EMR) can be performed both to obtain a definite diagnosis and to
treat the patient [6, 9]. In contrast, in patients with flat and widespread lesions, as in the present case, ESD may be more useful than EMR to obtain a definite diagnosis.

In general, the optimal treatment approach for PMME is surgical resection with dissection of regional lymph nodes, but total or near-total esophagectomy offers the best survival outcome (about 5 years, versus 9 months for local resection) [2]. Therapeutic options such as chemotherapy, immunotherapy, and radiotherapy provide limited benefits, and are generally not recommended as first-line treatment options for patients with operable PMME. In contrast, there are five case reports of PMME treated with EMR [6, 8, 9, 12, 13]. These were three polypoid and two flat lesions, 22.8 (5–50) mm in size, and all cases were stage IA according to TNM7 classification. The size of the present case was larger than these reported cases. Morphological type was not related to the depth of invasion in these reported cases. All of the reported cases have had no recurrence or metastasis after EMR; however, surgical resection with dissection of the regional lymph nodes should be selected even though PMME may be early stage and treated by endoscopic resection, because no reports have discussed the risk of recurrence, and metastasis of a large number of early stage PMME [4].

In summary, we describe a patient with flat-type, early stage PMME diagnosed with ESD, and treated successfully with radical surgical resection. In cases in which the biopsies could not be distinguished between early stage PMME and melanocytosis, EMR and ESD may be useful to obtain a definite diagnosis of flat-type, early stage PMME. However, the decision for endoscopic treatment without radical surgical resection should be considered carefully, even though the PMME was clinically an early stage tumor.
Competing interests: None

References