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Case Report

Concurrence of primary malignant melanoma of the esophagus with adenocarcinoma of sigmoid colon and villous adenoma of cecum: a case presentation

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Abstract: In this paper, a 74 years old male patient with complaints of dysphagia and hemoptysis is presented. Endoscopy revealed black colored mass protruding to the lumen at distal esophagus. Diagnosis of malignant melanoma was confirmed with biopsy. Examinations for staging purposes revealed masses at sigmoid colon and cecum. Biopsy was performed with colonoscopy. The mass at the sigmoid colon was diagnosed as adenocarcinoma and the mass at the cecum was diagnosed as villous adenoma. Although the treatment strategy is not straightforward, surgical treatment is the most important step. For this reason, patient underwent three field esophagectomy, anterior resection and right hemicolectomy in the first place. The patient is currently receiving his adjuvant chemotherapy and immunotherapy at postoperative 6th month. According to our knowledge, concurrence of these tumors with two different origins has only been reported in 1 patient before. Our patient has the significance of being the second reported case.

Keywords: Primary esophagus malignant melanoma, sigmoid colon cancer

Background

Primary malignant melanoma of the esophagus (PMME) is a rare disease with very poor prognosis. It is very rarely a part of synchronous tumors originating from different organs of the gastrointestinal system. For that matter, concurrence of PMME with adenocarcinoma of sigmoid colon has only been reported in one case [1]. It is generally seen in sixth or seventh decade, and two times more frequent in men than in women [2]. Among all malignancies of the esophagus, its frequency is approximately 0.1-0.2% [3]. Additionally, among non-cutaneous melanomas, its frequency is approximately 0.5% [4]. Since its first report in 1906 by Baur, nearly 400 cases have been reported [5]. Like the other malignancies of the esophagus, dysphagia, retrosternal pain and weight loss are the most common symptoms. It is generally localized to lower esophagus [6]. Average survival is 14.2 months following radical resection, and 9 months following local excision [7]. Five year survival rates have been reported and 4% [7, 8]. As is the case with our patient, treatment algorithm in synchronous tumors of the gastrointestinal tract is unclear. We aimed to present a case who had concurrent primary malignant melanoma of the esophagus and adenocarcinoma of the sigmoid colon.

Case presentation

Seventy-four years old male patient presented to gastroenterology department with complaints of dysphagia and hemoptysis. There was no abnormality in his systemic examination. He had no history of previous operation or medication use. His routine blood tests and tumor markers were normal. An upper gastrointestinal system endoscopy was performed in the first place. There was a black colored lesion with ulcerated top at 1/3 middle section of esophagus which was protruding to lumen and approximately 3 cm in diameter; a biopsy was performed (**Figure 1**). Biopsy result was reported as malignant melanoma. Abdominal CT was performed to determine whether the tumor was

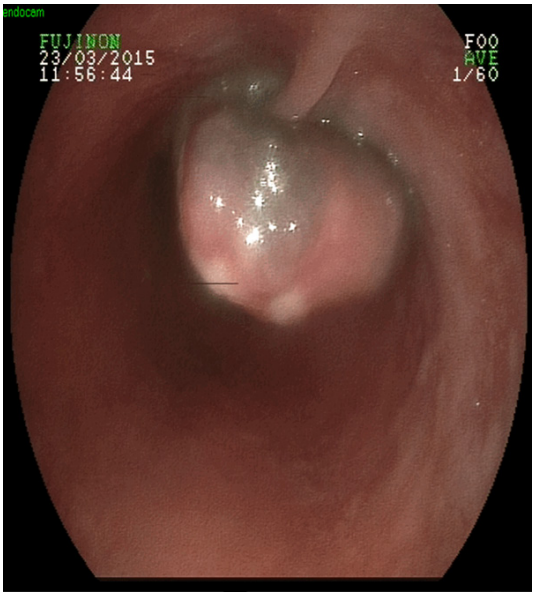

Figure 1. Endoscopic image of the black colored polypoid lesion prior to treatment.

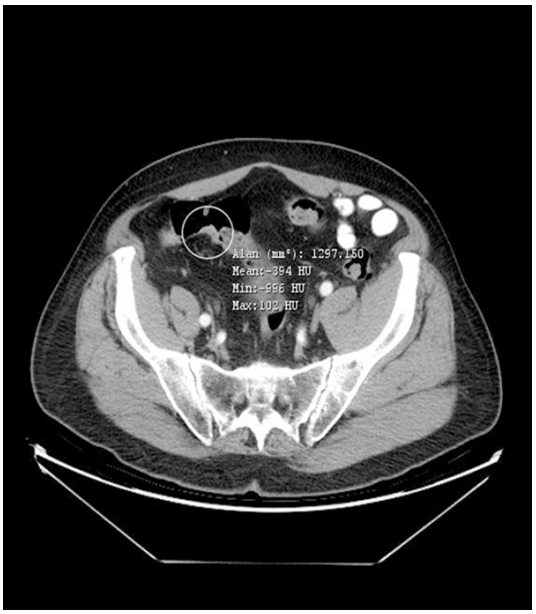

Figure 2. Abdominal CT image of the adenocarcinoma of the sigmoid colon.

primarily from esophagus or metastatic and for staging purposes, which revealed irregular wall thickening at approximately 2 cm segment of distal sigmoid colon (**Figure 2**). After this finding, colonoscopy was performed, and a mass was observed at sigmoid colon which was approximately 2.5 cm. in size, had wide base and protruded to the lumen; a biopsy was taken. In addition, there was a wide based lesion in cecal wall that was approximately 2 cm in size, which was also biopsied. The lesion at the sigmoid colon was diagnosed as well differentiated adenocarcinoma arising in villous adenoma, whereas the lesion at cecum was diagnosed as severe dysplasia arising in villous adenoma. PET-CT confirmed tumoral lesion at distal esophagus (**Figure 3**). No other primary origin was detected in dermatological screening for cutaneous melanoma. After these findings, endoscopic biopsy from the mass at esophagus was reported as possible primary malignant melanoma of the esophagus (**Figure 4A-D**). Additionally, according to results of biopsy taken from the mass at sigmoid colon at colonoscopy, there was adenocarcinoma at sigmoid colon and villous adenoma at cecum (**Figure 5A**, **5B**).

Patient's treatment plan was discussed at multi-disciplinary council. It was planned to perform three field esophagectomy with dissection, surgical resection for adenocarcinoma of sigmoid colon and villous adenoma at cecum in the first place, and chemotherapy, radiotherapy and immunotherapy later. In the resected esophagectomy specimen, tumor diameter was 3 cm, tumor was infiltrated to mucosa and submucosa (pT1), there was angiolymphatic invasion, surgical borders were negative, and there was 1 metastatic lymph node, According to IUCC staging system, the tumor at esophagus was determined as Stage IIB (T1bN1M0). In sigmoid colon resection material, tumor diameter was 1.5 cm, tumor was infiltrated to mucosa and submucosal layer (pT1), there was no angiolymphatic invasion or metastatic lymph node. In segmentary colon resection material, the mass at cecum was histopathologically diagnosed as villous adenoma. Patient is at postoperative 6th month and currently receiving chemotherapy and immunotherapy for treatment.

Discussion

Primary malignant melanoma of the esophagus is a quite rare tumor, and its concurrency with synchronous tumors originating from the gastrointestinal tract is even rarer. Although there have been approximately 400 reported PMME cases up to day, its concurrency with adenocarcinoma of the sigmoid colon has only been reported in one case [1].

PMME is particularly observed during sixth and seventh decades and is 2 times more frequent

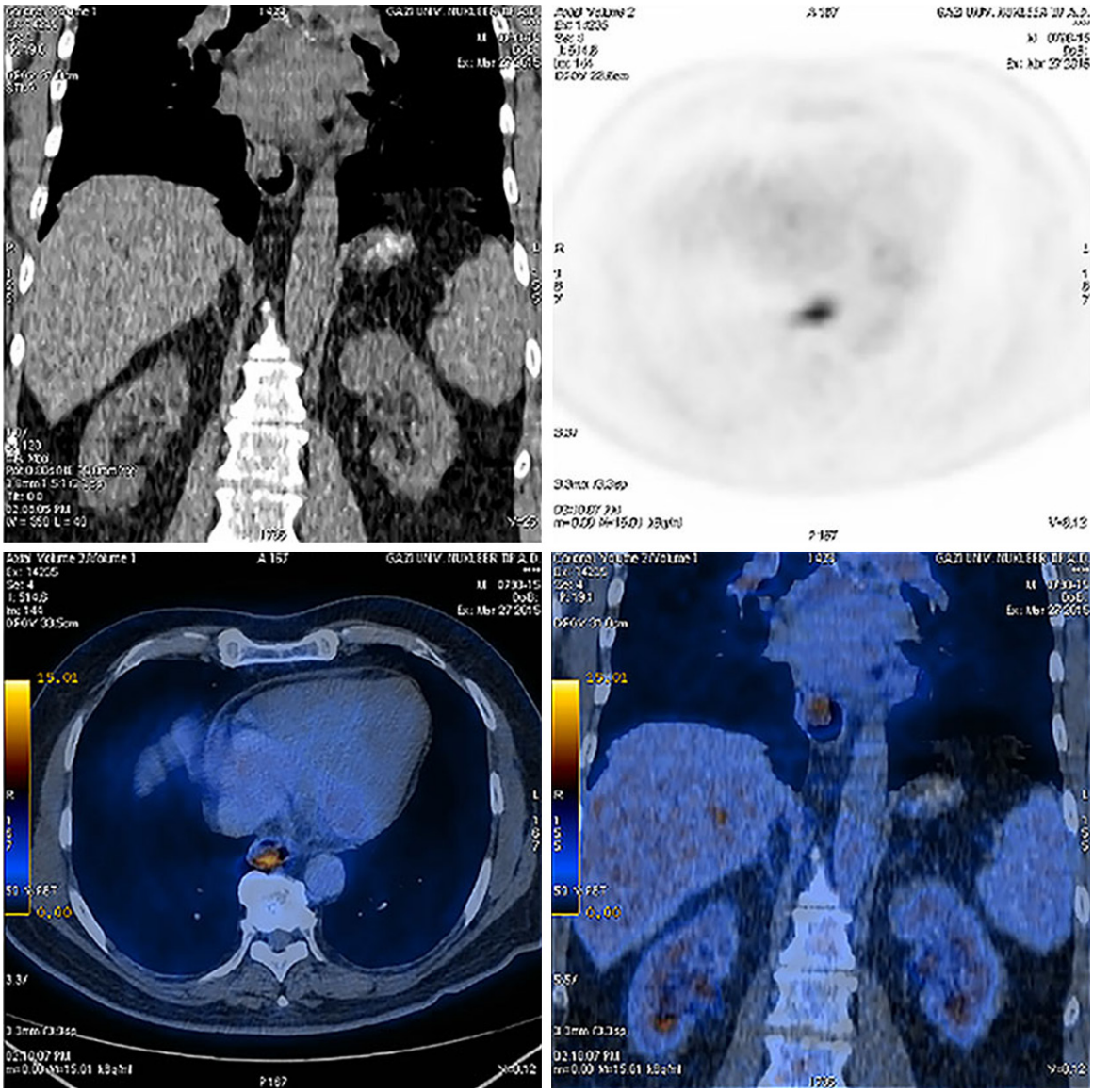

Figure 3. FDG PET image of the increased uptake by primary malignant melanoma of the esophagus.

in men than in women [2]. Our case is 74 years old, which is an age similar to other reports in literature. Among all malignancies of the esophagus, its frequency is approximately 0.1-0.2% [3]. Additionally, among non-cutaneous melanomas, its frequency is approximately 0.5% [4]. Its genetic variance has not been determined. Esophageal melanocytosis is particularly thought to be a precursor for melanoma [10]. Similar to other esophageal cancers, dysphagia, non-specific retrosternal pain, weight loss and less frequently, hemoptysis are observed clinically in patients [11]. Our case had dysphagia and hemoptysis. Endoscopy is gold standard for diagnosis. CT and PET-CT are important to determine its relation with surrounding structures, lymph node metastasis and for staging. In our case, CT that was performed for staging purpose revealed a suspicious synchronous tumoral mass at sigmoid colon. It should be noted here that malignant melanoma is more likely to be cutaneous in origin, and metastatic lesions can be detected frequently at the time of diagnosis.

As it is a rare condition, there is no standard treatment plan for PMME. There are contradictions about in what conditions should neoadjuvant treatment be administered, to what extent should surgical resection be made and how adjuvant treatment plan should be; and presence of synchronous adenocarcinoma at sig-

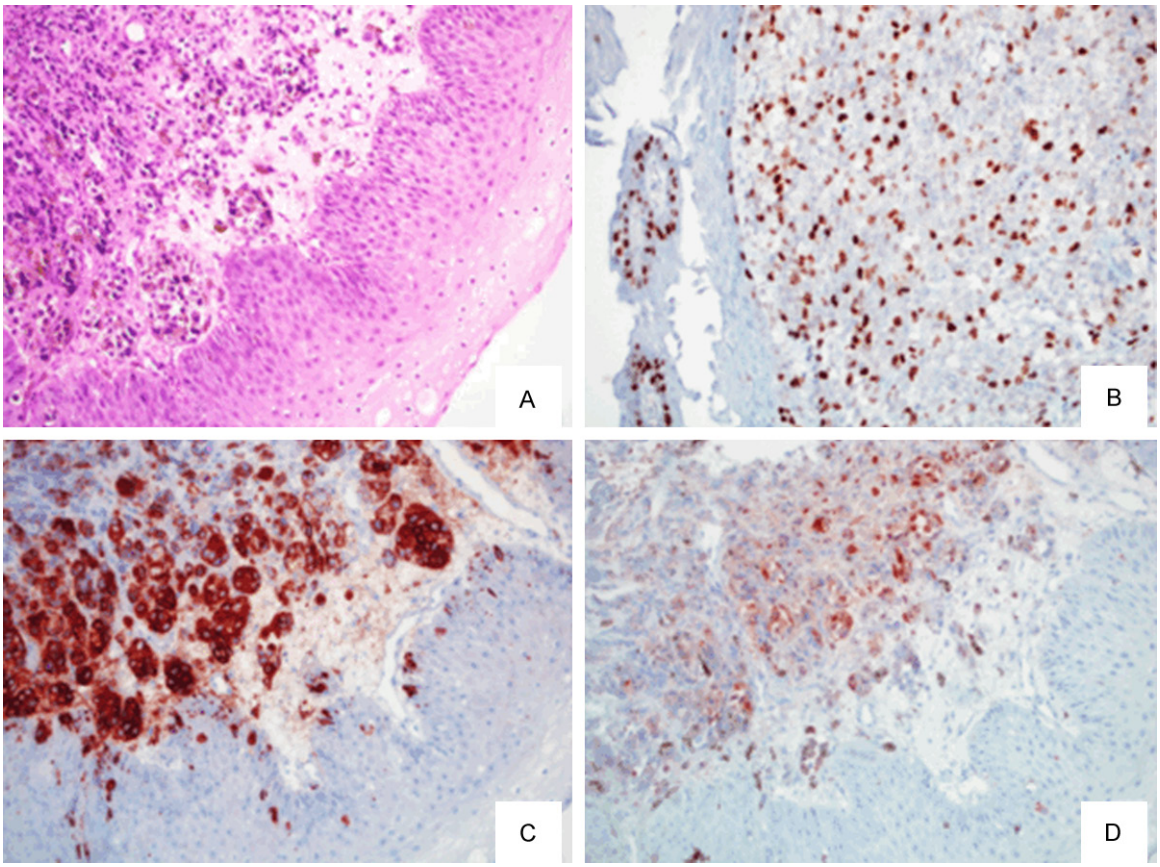

Figure 4. Histopathological images of the malignant melanoma of the esophagus. (A. H&E × 200); Nest of melanocytes containing melanin pigment are observed infiltrating lamina propria under stratified squamous epithelium of the esophagus. (B. Streptoavidin/biotin immunperoksidaz × 200); 75% proliferative activity was detected in tumor cells with Ki-67. (C. Streptoavidin/biotin immunperoksidaz × 200); Nests of melanocytes are positive for S-100 protein and HMB-45. (D. Streptoavidin/biotin immunperoksidaz × 200); Nests of melanocytes are positive for HMB-45.

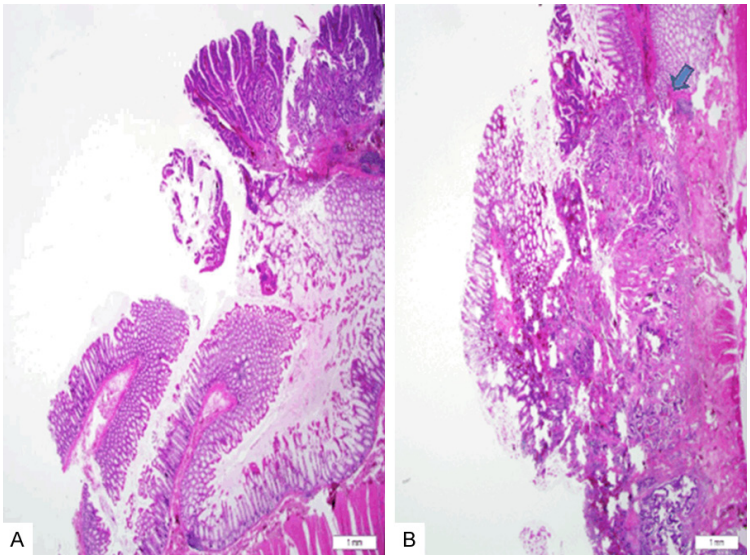

Figure 5. (A. H&E × 12.5); Histopathological image of well differentiated adenocarcinoma arising in villous adenoma. (B. H&E × 12.5); well differentiated adenocarcinoma arising in villous adenoma (Arrow: submucosal invasion area).

moid colon in our case further complicated this treatment strategy. Unfortunately 30% of patients are unresectable at the time of diagnosis [12]. Whether there is metastasis to lymph node or not, en bloc resection of the lesion is very important in terms of survival. Even in the best centers, average survival despite radical resection is 14.2 months [7]. On the other hand, average survival in cases who undergo endoscopic local resection has been reported as approximately 9 months. In addition, five year overall survival has been reported in the rate of 4% [7, 8]. In one study including 13 patients in a single center, lymph node metastasis ratio was reported

as 0% when the tumor infiltrated only mucosal layer, but as 42.9% when tumor infiltrated submucosal layer [13]. The same study reported presence of recurrence within 1 year in all patients at stage 1b or higher. One year overall survival was determined as 54%, whereas five year overall survival was 35.9%. Additionally, lymph node metastasis state was determined to be an independent prognostic factor. Our patient was T1bN1M0 (Stage IIB) for PMME and T1N0M0 (Stage I) for adenocarcinoma of sigmoid colon according to IUCC. Although these results suggest the tumor is at early stage, we think presence of angiolymphatic invasion and lymph node metastasis are poor prognostic criteria. Additionally, we think synchronous colon cancer detected in our case is unfavorable in terms of prognosis though dependent on the stage. Concurrence of PMME and adenocarcinoma of colon, and in addition, detection of villous adenoma at cecum further complicated already hard-to-decide treatment strategy. Our case had three field dissection total esophagectomy for PMME, anterior resection for adenocarcinoma of the sigmoid colon and right hemicolectomy for villous adenoma at cecum since endoscopic resection was not possible.

In conclusion, PMME is a rare tumor with poor prognosis. Presence of concurrent adenocarcinoma of the colon complicates the situation. Therefore, the diagnosis, staging and treatment strategy for the disease should be carefully planned.

Disclosure of conflict of interest

None.

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