Black pigmented dysphagia

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Case description

A 63-year-old male of Turkish origin was referred to our department for further investigation of dysphagia for solids since 2 years with recent occurrence of food impaction. The patient had no significant medical history besides smoking. There was no mention of weight loss or other systemic complaints. Upper gastrointestinal endoscopy showed a large subobstructive pedunculated mass in the lower thoracic esophagus. The lesion was remarkably pigmented and seemed to be infused with Chinese ink (see figure 1). Biopsies were taken.

Question

What is your diagnosis?

Answer

The biopsies showed a proliferation of strong pigmented melanocytes with atypical features, suggestive for a malignant melanoma. After a thorough clinical search for a primary skin tumor and no evidence for metastatic disease on PET-CT and MRI of the brain, diagnosis of a primary malignant melanoma (PMM) of the esophagus was made (cT3N0M0). A minimal invasive esophagectomy was performed where, at histopathological analysis, one affected gland was found within the resection area (pT1N1). Given the high risk of relapse in mucosal melanoma at this stage, adjuvant treatment with anti-PD1 was started (NGS ongoing, given subtype no BRAF mutation was expected).

Malignant melanomas are malignant tumors from the melanocytes and can arise from both cutaneous and mucosal surfaces (1). Mucosal melanomas only account for 0.8-3.7% of the malignant melanoma and have a worse prognosis than the cutaneous form, possibly due to late diagnosis and patient's delay (1). They occur in places not exposed to UV light, etiology remains unknown. A PMM of the esophagus is an extreme rare entity accounting for 0.1-0.2% of all primary neoplasms of the esophagus (2). Most common symptoms are dysphagia and weight loss (2). Wide excision surgery is the treatment of choice (1). The overall prognosis is poor with five-year survival of 0-45% (1). The exact role of adjuvant chemo, immuno- and radiotherapy remains unknown given their very low prevalence. Adjuvant pembrolizumab during 12 months was efficient in a large placebo-controlled trial (3).

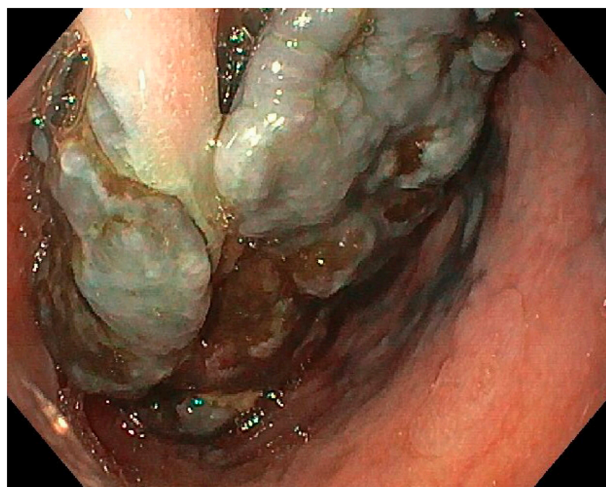

Figure 1. — Upper gastrointestinal endoscopy showed a large subobstructive pedunculated mass in the lower thoracic esophagus. The lesion was remarkably pigmented.

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