

Primary Malignant Melanoma of the Esophagus: Diagnostic Challenges, Therapeutic Decisions, and Outcomes in a Rare Case Report

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BACKGROUND

Primary Malignant Melanoma of the Esophagus (PMME) is a rare and aggressive neoplasm, 0,1-0,2% of all esophageal malignancies, Its nonspecific presentation and the need to exclude other primary sites pose significant diagnostic challenges. Management guidelines are not well established due to the rarity of the disease and the patient prognosis remains poor.

CASE PRESENTATION

A 58- year-old woman was admitted with progressive dysphagia and weight loss of 15 kg. Upper gastrointestinal endoscopy revealed a 5cm polypoid mass extending 30-35cm from the incisors (Z-line at 38cm). Histopathological analysis and immunohistochemistry confirmed malignant melanoma with a strong positivity of S-100 and Melan-A. The mass was also identified in the middle-distal esophagus on the CT scan. MRI, PET-CT,colonoscopy, full body dermatoscopy, and slit-lamp ophthalmoscopy were negative for distant metastases and cutaneous melanoma, leading to the diagnosis of PMME. The Multidisciplinary Team Meeting decided the administration of 7 cycles of neoadjuvant immunotherapy with nivolumab and ipilimumab before the surgical treatment but there was no therapeutic response as proven by the subsequent upper GI endoscopy. The surgical management included total esophagectomy with two-field lymphadenectomy, gastric tube reconstruction and cervical esophagogastric anastomosis.The histopathological analysis confirmed the diagnosis of PMME, R0 resection and one positive periesophageal lymph node while the immunohistochemical analysis showed strong positivity for Melan-A, S100, SOX10, HMB45 and negative results for AE1/AE3 and DOG-1. The patient remained in a good clinical condition and was discharged on the 11th postoperative day with adjuvant immunotherapy. At the 6 month follow up, the patient remained clinically well with no evidence of locoregional recurrence or distant metastasis.

CONCLUSIONS

This case report highlights the importance of the surgical management of PMME with esophagectomy and two-field lymphadenectomy and the administration of adjuvant immunotherapy, despite the lack of response to neoadjuvant, in order to achieve the best outcome .

