

## Primary Malignant Melanoma of Esophagus

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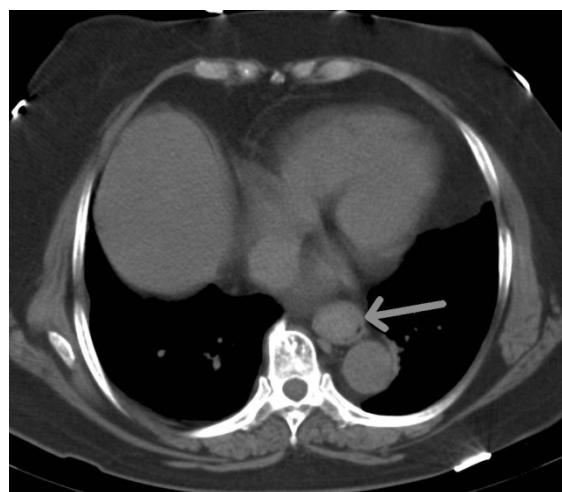
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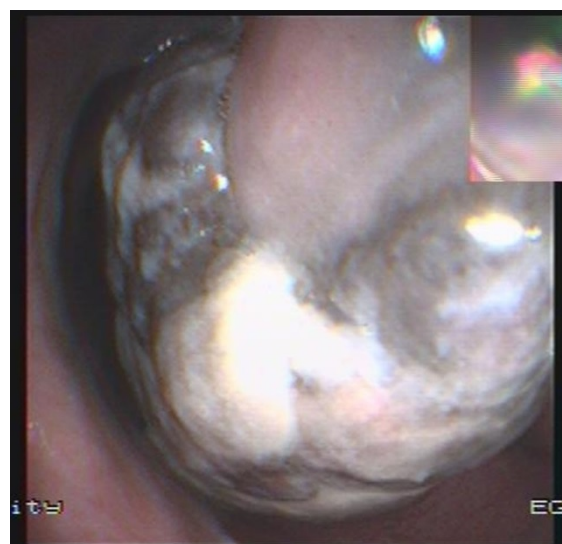
### Clinical Image

A 63-year-old woman presented with progressive dysphagia for 1 month, and had lost approximately 2 kg in weight since the onset of his illness. Physical examination showed no abnormality. MSCT scan revealed a solid mass in the distal third of the esophagus, where local fat space around the esophagus still remained (Figure 1). Endoscopy revealed an irregular dark endoluminal and lobulated mass suited 30-35 cm from the incisors (Figure 2). Endosonography showed esophageal mass to be solid, heterogenous in echo-architecture and arising from the deep mucosa (Figure 3). The biopsy specimen suggested suspected malignant melanoma. Extensive detailed examination revealed no other skin, anal, facial or rectal lesions. The patient underwent an Ivor-Lewis esophagogastrectomy and lymph node dissection. The surgical specimen showed a 5.0 cm × 4.0 cm × 4.0 cm, polypoid and pigmented lesion located in the distal esophagus. The tumor was composed of plump spindle-shaped cells with pigmented cytoplasm, prominent nuclei and numerous mitosis (Figure 4). Immunohistochemical stains were positive for S-100 protein and HMB-45. Therefore, a diagnosis of primary malignant melanoma of esophagus (PMME) was made. The patient had an uneventful recovery and was in good condition on the follow-up of one year.

Malignant melanoma is a typical cutaneous tumor, originating from melanocytes. However, the origin of melanocytes in the esophagus remains hypothetical. PMME is exceedingly rare, varying its incidence from 0.1% to 0.2% of all esophageal malignant tumors [1]. PMME may be found in all areas of the esophagus, but has predominantly been located in the lower two-thirds of the esophagus. PMME occurs twice as frequently in men as in women and usually presents during the sixth and seventh decades of life. Imaging studies such as barium meal and CT only reveal esophageal occupying lesion. The characteristic appearances is a polypoid irregular pigmented and friable lesion at endoscopy, but only 54.7% of cases are diagnosed preoperatively as malignant melanoma. Endoscopic ultrasound is seldom applied and common a hypoechoic or a mixed echogenicity mass, as described in our case. Because of the predominant submucosal location of the tumor in more than 50% of the cases, endoscopic biopsies were negative or misinterpreted as poorly differentiated squamous cell carcinoma [2]. PMME is a rare disease that is characterized by aggressive invasion, early metastasis and poor prognosis [3]. Treatment protocols are not well-established, but an esophagectomy is still the treatment of choice.



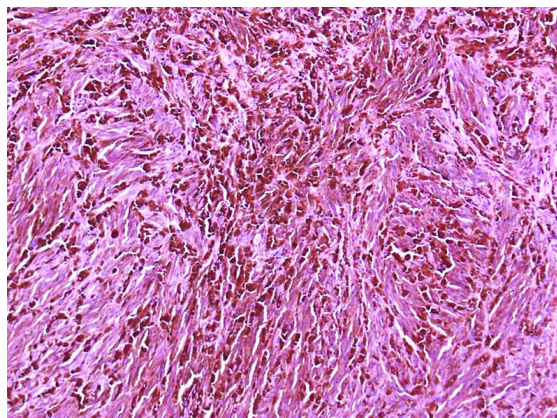
**Figure 1:** Chest axial CT showed a solid mass in the esophageal lumen.



**Figure 2:** Endoscopy showed an irregular dark endoluminal mass.



**Figure 3:** Endosonography showed a solid, heterogenous in echo-architecture mass.



**Figure 4:** The tumor was composed of plump spindle-shaped cells with pigmented cytoplasm, prominent nuclei and numerous mitosis.

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