Imaging Findings of Primary Malignant Melanoma of the Esophagus: a case report

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Primary melanoma of the esophagus is not frequently encountered clinically but rather high degree malignant that have a poor prognosis. Here we report an uncommon case of primary melanoma of the esophagus with nodal metastasis over the right neck and discuss its radiographic findings, including esophagogram, contrast-enhanced computed tomography (CECT), sonography of neck and positron emission tomography (PET). When characteristic findings as polypoid, intraluminal, and nonobstructive masses are present on image studies, some diagnosis such as spindle cell carcinoma, leiomyosarcoma and lymphoma should be considered first. However, the diagnosis of primary esophageal melanoma always should be kept in mind despite of its rarity.

Key words: Esophagus, CT; Esophagus, neoplasm; Esophagus, PET; Melanoma

Primary melanoma of esophagus is an extremely rare tumor and the prognosis is dismal despite of surgical resection. To our knowledge, although some cases of primary melanoma of esophagus have been reported in the radiology literature, associated full series of imaging findings including esophagogram, contrast-enhanced computed tomography (CECT), positron emission tomography (PET) and sonography are discussed the first time. We believe that early detection of the esophageal tumor is imperative. The staging of esophageal melanoma with nodal metastasis based on the imaging findings is extremely important that may lead to better prognosis.

CASE REPORT

A 40-year-old woman was admitted to our hospital on account of progressive dysphagia for 3 months with body weight loss about 9 kg in recent 2 months. She denied any history of systemic illness or family history of cancer. On physical examination, she appeared generally well nourished and in good health. There were no suspicious pigmented lesions and no ocular abnormalities. Regular laboratory examinations revealed unremarkable finding.

Imaging studies, including esophagogram, contrast-enhanced computed tomography (CECT), positron emission tomography (PET) and sonography of the neck, were performed during the hospitalization.

The esophagogram showed a polypoid intraluminal mass about 6 cm in length with mucosal destruction over the middle third esophagus and without significant obstruction (Fig. 1). The CECT of chest revealed segmental circumferential wall-thickening with adjacent soft tissue lesion over the middle third esophagus from the level above the carina downward extension about 6 cm in length (Fig. 2). For the detection of distal metastasis, the PET was performed and revealed abnormal increased FDG uptake at middle esophagus with focal extension to right prevertebral space. A FDG-avid lesion adjacent to the right jugular vein region of the neck is also identified (Fig.
3). Nodal metastasis was suggested but tumor thrombus in the jugular vein could not be ruled out completely. Sonography of the neck was performed and revealed a well-defined, ovoid-shaped, soft tissue mass about 1.6 cm in long axis in the posterior aspect of the right jugular vein with extrinsic compression (Fig. 4). Finally, the patient received total esophagectomy with reconstruction of esophagus and nodal dissection of the right aspect of neck. The final pathologic findings revealed primary esophageal melanoma with nodal metastasis of neck (Fig. 5). The patient recovered from the operation without complications and was discharged from the hospital. Then she received radiotherapy and chemotherapy, and is still alive for more than 6 months.

**DISCUSSION**

Primary esophageal melanoma is an extremely rare tumor, accounting 0.1%-0.5% of all esophageal malignancies with a poor 5-year survival rate of 4.2% [1, 2]. It could be diagnosed only in patients who have no history of melanoma involving the skin, eyes, anus or vagina. The tumor usually occurs in the fifth decade or later and twice more frequently in men than in women [3]. It is most commonly in the lower half of the thoracic esophagus, which may be due to the greater concentration of melanocytes in this region [4,
The most common symptoms of esophageal melanoma are dysphagia (73%), weight loss (72%), pain (44%) and melena (10%) [2]. The esophagogram is usually performed initially due to the presentation of dysphagia. The tumor usually reveals bulky, polypoid intraluminal mass and focal expanding without obstruction located in middle third or lower third esophagus [6]. The CECT is often performed for demonstrating lesion extent in the adjacent nodes, lung and pleura. It may reveal large mass with circumferential wall thickening of esophagus. In our case, the tumor shows polypoid-shaped with mucosal destruction over the middle third esophagus without significant obstruction on esophagogram and segmental wall thickening over the middle third esophagus. The adjacent soft tissue mass at right prevertebral space on CECT is suggestive of local invasion.

In recent years, it has been reported that PET may be useful in evaluating distant metastases of malignancy [7, 8]. In this patient, PET was performed and showed not only the esophageal lesion but also a metastatic lesion in the right neck. The diagnosis of metastatic node rather than tumor thrombus was depicted on sonography. The result is important for the decision of surgical intervention.

The differential diagnoses of esophageal melanoma include squamous cell carcinoma, adenocarcinoma, spindle cell carcinoma, leiomyosarcoma and lymphoma. Unlike squamous cell carcinoma or adenocarcinoma of esophagus, esophageal melanoma almost shows polypoid intraluminal mass and focal expanding without significant narrowing of lumen.

Levine described it is difficult to differentiate primary esophageal melanoma from other polypoid tumors such like spindle cell carcinoma (the major consideration), leiomyosarcoma and lymphoma based on the imaging findings [9].

The first choice of treatment for primary melanoma of the esophagus is surgical resection with dissection of the lymph nodes. Adjuvant radiotherapy may improve the symptom of dysphagia but its effectiveness is unproved [2]. Some literatures reported that chemotherapy and immunotherapy are useless in treatment [3]. But the numbers of the cases were small. Further evaluation with a large number of patients is needed.

In conclusion, primary melanoma of the esophagus is a rare but aggressive malignancy that has poor 5-year survival. When characteristic findings as polypoid, intraluminal, and nonobstructive masses are present on image studies, the diagnosis of primary esophageal melanoma should be kept in mind despite of its rarity. Preoperative diagnosis and detecting metastasis are important, since more aggressive surgery may be needed to properly treat these patients.

REFERENCES


Figure 5. Histological specimen of the tumor shows hyperchromatic round uniform tumor cells invading the depth of the submucosal layer. (H&E stain, X200)
原發性食道黑色素瘤的影像發現：病例報告

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食道的原發性黑色素瘤在臨床上是不常見的，但卻是極惡性且有不好的預後。我們在這裡報告一個少見的原發性食道黑色素瘤且在右頸部有淋巴結的轉移，並且討論影像學的發現，包括了鉀劑食道攝影，電腦斷層攝影，超音波及正子斷層攝影。當在影像學上有特別的發現像是類腫肉狀，腔室內的，非阻塞性的腫塊時，一些診斷像是梭狀上皮細胞瘤，橫紋肌肉瘤，淋巴瘤都需作第一考慮。但是原發性食道黑色素瘤總是需要納入鑑別診斷雖然它是很罕見的。

關鍵詞：食道，電腦斷層攝影：食道，正子攝影：食道腫瘤，黑色素瘤