Malignant Melanoma of Oesophagus: A Rare Entity

Özofagus Malign Melanomu: Nadir Bir Hastalık

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ABSTRACT

We are discussing a case of malignant melanoma of esophagus, initially presented to us with very atypical presentation. On further history, patient claim having dysphagia for few months, initially to solid then followed by liquid, progressively worsening over few months. On further examination, OGDS reveal pedunculated growth at 30 cm and 32 cm with skip free intact mucosa in between. Subsequently, we optimize his nutritional status and performed open tranhiatal total esophagectomy with cervical anastomosis and feeding jejunostomy. Post-operative recovery was uneventful, and patient was well upon discharge. Histopathology of the specimen revealed malignant melanoma of the esophagus. On subsequent follow up. PET scan was also done and shows stable disease with no evidence of tumor recurrence.

Keywords: Melanoma, Esophagus, Dysphagia

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ÖZET

Başlangıçta çok atıplı bir sunumla bize sunulan bir özofagus malign melanomu vakasını tartışıyoruz. Daha fazla önüne, hasta birkaç aydır disfaji olduğunu iddia ediyor, başlangıçta kati, ardından sıvı, birkaç ay içinde giderek kötüleşen. İleri incelemekte OGDS, 30 cm ve 32 cm’dede pedinküle büyüme ve arada atlamasız intakt mukoza gösterir. Daha sonra beslenme durumunu optimize ettik ve servikal anastomoz ve beslenme jejunostomi ile açık tranhiatal total özofajektomi yaptık. Ameliyat sonrası iyileşme olayındı ve hasta taburcu olmadan sonra iyidi. Önemli histopatoloji yemek borusunun malign melanomunu ortaya çıkardı. Sonraki takipte. PET taraması da yapıldı ve tümörün nüketmesine dair hiçbir kant olmadan stabil hastalık gösterdi.

Anahtar Sözcükler: Melanom, Özofagus, Disfaji

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INTRODUCTION

Melanoma is more frequently found in sun-exposed areas, however it can appear in other sites including the mucosal surfaces, like the esophagus, however the occurrence is rare(1). Melanocytes are present in small amounts in the squamous epithelium or basal membrane of the esophagus and can be the precursors of melanocytosis, leading to primary malignant melanoma of the esophagus. Primary malignant melanoma of esophagus (PMME) is relatively rare tumor, representing approximately 0.1% to 0.2% of all esophageal malignant tumors(2). PMME is characterized by aggressive local invasion and early metastasis. PMME often presents as polypoid lesion and usually causes dysphagia to both solid and liquid progressively, retrosternal and/or epigastric pain. Over the years, the detection rates have improved due to early detection through numerous screening program. The tumour is often located from the middle to lower thoracic esophagus and the characteristic endoscopic finding of PMME is a polyloid lesion that is usually pigmented, in which these both characteristics were seen in our case. The prognosis of PMME is poor relatively because of its highly malignant biological behavior and its tendency to frequently dissemination. The recently reported 5-year survival after surgical resection is 37.5%(3), which is lower than that of esophageal cancer.

CASE REPORT

67 years old gentleman, with no known co-morbidities, presented with history of dysphagia for almost 6 months duration. Dysphagia is associated with constitutional symptoms. He visited general practitioner multiple times but was treated as gastritis before he come to us. He is an otherwise fit with general body built and denies any family history of malignancy running in the family. He also occasionally complains of on and off vomiting secondary to dysphagia. OGDS reveal pedunculated growth at 30cm and 32 cm with skip free intact mucosa in between, bleeding on touch, occupying 3/3 of the esophageal lumen, able to pass through scope.

We admitted patient straight away after OGDS for nutritional assessment and build up, prior to operation. Once optimize, we proceeded with operation, open Transhiatal total esophagectomy with cervical anastomosis and feeding jejunostomy. Patient recovered well from operation and upper gastrointestinal study at day 5 post operation revealed no evidence of anastomotic leak.

Histopathology (HPE) of the specimen revealed malignant melanoma of esophagus. Cut sections of the specimen revealed two polypoidal growths measuring 40×30×14mm and 30×23×27mm. The masses are 15mm apart and there is another small raised lesion measuring 8×5×3mm, 10mm away from polypoidal masses. The masses surfaces are hemorrhagic with ulceration and slough, cut sections show they are confined within the submucosa and are well-demarcated from the underlying muscularis propria. Microscopically, sections of the polypoidal masses and a raised lesion show well-circumscribed hypercellular tumors beneath the mucosa. The cells are large and moderately pleomorphic, displaying round nuclei with vesicular chromatin, prominent nucleoli and abundant cytoplasm. Spindle-shaped tumor cells are also present in areas with melanin pigments seen within the cytoplasm. Immunohistochemical studies show the malignant cells are strongly positive for HMD45 with focal positivity for S100, Melan A and CD117. One out of 11 lymph nodes were positive, it is situated at the posterior surface measuring 20×15×14mm with hemorrhagic tan cut surface.

He was seen by Oncologist and Dermatology team as well. Dermatology team ruled out malignant melanoma of the skin, in view of same pathology noted on the esophagus. Subsequently CT scan were performed about 6 months post operation which revealed stable disease with no local recurrence or distant metastasis with stable lung nodule and liver cyst. One-year post operation PET scan were done and shows stable disease with no evidence of tumor recurrence.

His recovery has been speedy and uneventful. We are continuing our assessment for this patient together with oncology team.

Following are the cut section of computed tomography of our patient discuss in this case with comparison of pre-operation and post-operation CT.

Figure 1 – Sagittal view of CT scan showing the tumor location at the mid-oesophagus (pre-operation)

Figure 2 – Sagittal view of CT scan showing the gastric pull up, stomach noted to be within the thoracic cage (post-operation)
Primary melanoma of the gastrointestinal tract is exceedingly rare, whereby cutaneous melanoma is the most common malignancy to metastasize to the gastrointestinal tract. Primary malignant melanoma of the esophagus (PMME) is a rare disorder and has a reported prevalence of 0.1–0.5% of all esophageal malignancies. The overall prognosis is poor, however early detection of PMME allows better prognosis for patients. Like all cancers, if early detection can be done, the disease progress can be controlled by performing esophagectomy with or without chemoradiation, case to case basis(5).

PMME is most commonly located in the middle or distal third of the esophagus, in approximately 90% of cases, probably because of the greater concentration of melanocytes in these regions. The diagnostic criteria for PMME require the presence of melanin granules within the tumor cells as well as melanocytes in the overlying epithelial layer and areas of junctional activity within squamous mucosa and the adjacent epithelium.

Diagnostic criteria for PMME are defined by Allen and Spitz(6) as follows, (1) a typical histological pattern of melanoma, with melanin granules inside the tumor cells, and an (2) origin in an area of junctional activity in the squamous epithelium. Junctional activity is defined as some nests of melanocytes with varying degrees of atypia found at the mucosal-submucosal junction adjacent to the tumor mass. Immunohistochemical studies also offers accurate diagnosis of PMME, in which it reveals a positive antibody-specific cytoplasmic reactivity to HMB-45 and S-100 proteins, similar to our case discussed earlier. In the present case, melanocytosis and junctional activity were surrounding the main tumor, and positive results of HMB45, S-100 and Melan-A were revealed by immunohistochemical staining, which led to definitive diagnosis of PMME.

Treatment of PMME should be tailored based on each patient. The choice of intervention should be based on tumor size and location, presence of metastases, age, and co-morbidities of the patients(7). Kimura et al(8) on the other hand reported the first case of PMME treated by EMR and discussed the indications for EMR of superficial-type PMME. On the other hand, for patients with PMME at T1a, curative surgical resection could improve their overall prognosis. Although there is no absolute indication of adjuvant therapy for stage T1a and negative lymphoid metastatic cases because of low risk of metastasis and recurrence, as with regards to esophageal cancer, careful follow-up such as blood tests including tumor marker and contrasted enhanced imaging such as CT scan or PET scan is warranted. Table 1 below compares various study and the treatment approach in tackling PMME.

**Table 1: Adapted from Japanese Classification of Esophageal Cancer, 11th edition.**

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Size (cm)</th>
<th>Depth*</th>
<th>Treatment</th>
<th>Course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kido [16]</td>
<td>2000</td>
<td>60</td>
<td>M</td>
<td>Lt</td>
<td>4.0 × 2.0</td>
<td>T1a</td>
<td>CR</td>
<td>33 months</td>
</tr>
<tr>
<td>Mikami [17]</td>
<td>2001</td>
<td>41</td>
<td>F</td>
<td>Mt</td>
<td>0.8 × 0.6</td>
<td>T1a</td>
<td>CR</td>
<td>31 months</td>
</tr>
<tr>
<td>Kimura [18]</td>
<td>2005</td>
<td>73</td>
<td>M</td>
<td>Lt</td>
<td>1.8 × 1.3</td>
<td>T1a-LPM</td>
<td>EMR</td>
<td>15 months</td>
</tr>
<tr>
<td>Suzuki [19]</td>
<td>2008</td>
<td>62</td>
<td>M</td>
<td>Mt</td>
<td>7.0 × 4.5</td>
<td>T1a-EP</td>
<td>CR</td>
<td>33 months</td>
</tr>
<tr>
<td>Suzuki [19]</td>
<td>2008</td>
<td>67</td>
<td>M</td>
<td>Lt</td>
<td>5.5 × 5.5</td>
<td>T1a-LPM</td>
<td>CR</td>
<td>53 months</td>
</tr>
<tr>
<td>Miyatani [11]</td>
<td>2009</td>
<td>64</td>
<td>F</td>
<td>Lt</td>
<td>0.5</td>
<td>T1a-LPM</td>
<td>EMR</td>
<td>20 months</td>
</tr>
<tr>
<td>Wang [14]</td>
<td>2013</td>
<td>62</td>
<td>M</td>
<td>Mt</td>
<td>7.0 × 4.5</td>
<td>T1a</td>
<td>CR</td>
<td>93.7 months</td>
</tr>
<tr>
<td>Yamanoto [21]</td>
<td>2015</td>
<td>75</td>
<td>M</td>
<td>Lt</td>
<td>1.5 × 1.0</td>
<td>T1a-MM</td>
<td>CR</td>
<td>83 months</td>
</tr>
<tr>
<td>Our case</td>
<td>2015</td>
<td>78</td>
<td>F</td>
<td>Lt</td>
<td>5.7 × 3.8</td>
<td>T1a-MM</td>
<td>CR</td>
<td>39 months</td>
</tr>
</tbody>
</table>

**Notes:**


* All reported cases are still alive after the treatment, and none have had any symptoms of relapse or distant metastasis.

**Conflict of interest**

No conflict of interest was declared by the authors.
REFERENCES


6. Allen AC, Spitz S. Malignant melanoma; a clinicopathological analysis of the criteria for diagnosis and prognosis. Cancer. 1953; 6:1–45