

Malignant Melanoma of Esophagus and its Prognosis: A Case Report and Review of Literature

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Abstract

Primary malignant melanoma of the esophagus (PMME) is a rare entity, representing only 0.1-0.2% of all esophageal malignancies. The most common location of melanoma in esophagus is in the middle to lower thoracic part. Dysphagia or retro sternal discomfort or pain is the most common initial presentation. It commonly metastasizes via hematogenic and lymphatic pathways. Esophagectomy and lymph node dissection is the treatment of choice in localized cases. Survival of the patients is poor and usually <1 year after diagnosis. The purpose of this study was to describe a case of 60-year-old man with a history of progressive dysphagia since 12 months, the investigation of which led to a diagnosis of PMME. The patient was treated by radical resection, and now survived with no evidence of disease since 12 months after surgery.

Keywords: Diagnosis, Esophagus, Malignant melanoma, Prognosis

INTRODUCTION

Malignant melanoma is more frequently found in sun exposed areas; however, it can occur in other sites including the mucosal surfaces. The primary malignant melanoma of esophagus (PMME) is a rare entity and accounts for 0.1-0.2% of all malignant tumors in this organ, with a total of 337 reported cases up to the year 2011.¹ It occurs mostly in the elderly age group with an average age at diagnosis of 60.5 years, and a male to female prevalence ratio of 2:1, as in the present case.² Small amount of melanocytes are present in the normal squamous epithelium or basal membrane of the esophagus. These melanocytes can act as precursors of melanocytosis and primary malignant melanoma. Most of the cases are diagnosed in the advanced stages and with poor prognosis. Prognosis is not related to the tumor thickness.³ Mean survival period is 10 months after diagnosis. The present case is the esophageal melanoma, which underwent radical resection and survived till 22 months of the initial appearance of symptoms.

CASE REPORT

A 65-year-old male patient was presented with dysphagia since 12 months. Upper gastrointestinal (GI) endoscopy showed a friable pedunculated polyp on the posterior aspect of esophagus 30 cm from the incisor, which bleeds on touch (Figure 1). Biopsy from the growth was taken, and histopathological examination revealed melanin containing malignant epithelial cells in large sheets and also in small clusters along with adjacent areas of normal looking stratified squamous epithelial cells (Figure 2). Individual tumor cells are round to polygonal in shape having abundant amount of cytoplasm containing melanin, large round to oval nucleus and single central prominent macro-nucleoli (Figure 3). A contrast-enhanced computed tomography thorax revealed a pedunculated well defined enhancing lobulated lesion (of size 30 mm × 24 mm × 20 mm) in the lumen of mid esophagus at the level of D6-7 disc and is attached to the posterior wall of esophagus by a narrow peduncle of size 5 mm × 6 mm. Total esophagectomy and lymph node dissection

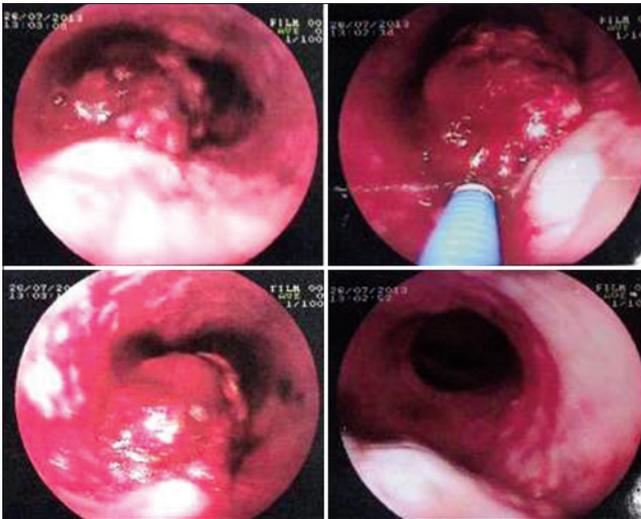


Figure 1: Upper gastrointestinal endoscopy showed a friable pedunculated polyp on posterior aspect of esophagus

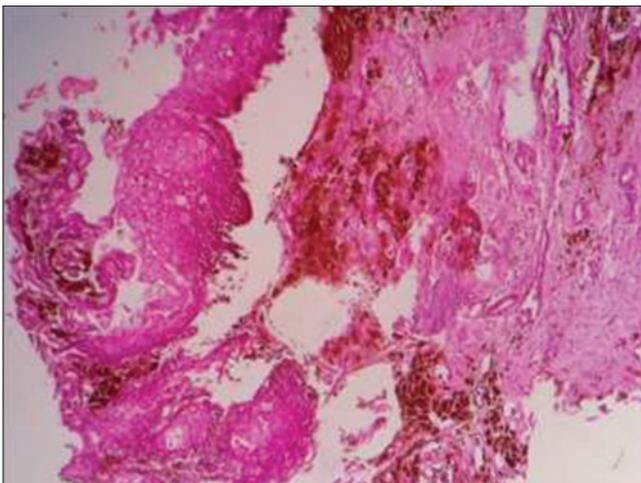


Figure 2: Presence of stratified squamous epithelial lining along with melanin containing malignant epithelial cells in sheets and small clusters (H and E, x100)

done and a polypoidal fleshy blackish growth found in the mid esophagus of size 3.5 cm × 3 cm × 2.5 cm with multiple mucosal dark intransit lesions, the growth is 7 cm away from proximal resection margin and 9 cm away from distal resection margin. Microscopic examination revealed malignant melanoma of esophagus invading up to submucosa, all cut margins free and all lymph nodes examined free of tumor. Immunohistochemistry showed positive for human melanoma black-45 (HMB-45). After 6 months of surgery, the patient developed a stricture at anastomotic site. Repeated dilatation done, but stricture persists, for which endoscopic retrograde cholangiopancreatography and stenting done. Now, patient is survived since 12 months after surgery without any recurrence of the disease.

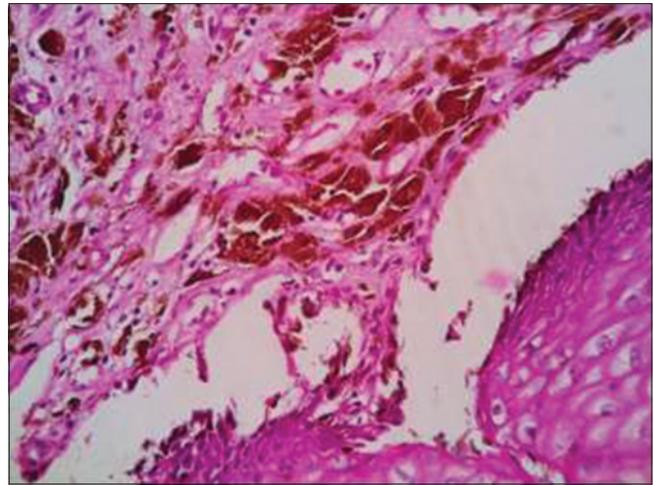


Figure 3: Presence of stratified squamous epithelial lining along with melanin containing malignant epithelial cells in sheets and small clusters. Individual cells are large having abundant amount of melanin containing cytoplasm, large round to oval nucleus and single prominent macro-nucleoli (H and E, x400)

DISCUSSION

Skin is the most common site for primary malignant melanoma. However, it can occur in other sites like in the GI tract and eyes. Among the GI tract, anorectal area is the most common.⁴ It is rare in the esophagus. PMME has a worse prognosis than cutaneous melanoma. The mean survival rate is reported to be <5% at 5 years and a mean survival rate of 10 months with a disease related mortality of 85%.⁵ It occurs most frequently in the middle and lower esophagus, as reported in more than 90% of cases in some series. This is due to a greater concentration of melanocytes in this location.⁶ PMME mostly have seen in solitary form. According to the literature, multiple lesions are seen in 12% of cases.⁷ Most of the cases are seen in pigmented form (85%). Only few cases of amelanotic melanoma have been reported. PMME grows in a lentiginous radial manner and involves mucosal and submucosal layers in most of the cases. Involvement of lymphovascular space invasion is common.

Dysphagia is the most common symptom in malignant melanoma similar to esophageal carcinoma, as in our case. The occurrence of hematemesis or melena is unusual. It may easily be missed by small biopsy of an esophageal growth. The diagnosis of malignant melanoma can only be established by upper gastrointestinal endoscopy with biopsy and immunohistochemical studies.⁸ Endoscopic findings usually show a pedunculated friable polypoidal masses, and pigmented tumor, covered by normal mucosa and rarely accompanied by ulcers.² Its color varies depending on the amount of melanin, which can be absent. Most often it

is non-pigmented, and histological examination showed the presence of epithelioid, spindle and anaplastic cells. In some cases, melanin granules are not detected in the cytoplasm. In these situations, immunohistochemical positive study of S-100 protein, HMB-45, neuron-specific enolase and negative for cytokeratin and carcinoembryonic antigen confirm the diagnosis of melanoma and exclude carcinoma. Distinguishing primary from metastatic melanoma is difficult, but absence of history of malignant melanoma elsewhere, presence of radial growth phase, and epithelioid and spindle cell histology are in favor of primary esophageal melanoma.⁹

In most cases, it is diagnosed at advance stage like aggressive local invasion, and lymphatic or distant metastases; survival is no more than a few months, despite multimodality treatments.^{10,11} The most common sites of metastasis are adjacent lymph nodes, liver, adrenal glands and lung.¹⁰ The PMME is prognostically poor due to aggressive behavior, late diagnosis, and advanced stage at presentation.¹² Recently, the prognosis seems to be improved due to early detection of the tumor. The primary treatment of esophageal melanoma is surgical excision with lymph node dissection inoperable cases. Total near-total esophagectomy offers the best survival outcome (about 5 years, vs. 9 months for local resection).⁵ A combination of esophagectomy and 3-field lymph node dissection as periesophageal, mediastinal and celiac trunk is the treatment of choice in PMME.¹ Due to the presence of frequent satellite lesion, broader surgical margins should be taken in comparison to other esophageal melanoma. In this procedure, it is difficult to determine cost benefit due to high post-operative morbidity and mortality and most patients die from disease recurrence in a short period of time.¹² In case of obstructive unrespectable tumors radiation therapy can be used as palliative treatment, and endoscopic metallic stents can also be used. The efficacy of treatment with radiotherapy, chemotherapy and immunotherapy are controversial.

CONCLUSION

PMME is a rare entity with aggressive behavior. Small biopsy may miss the diagnosis; so larger biopsy or multiple sites of biopsy should be taken. A better survival rate can be achieved if the diagnosis is made early. The treatment of choice is surgical resection, even in cases of recurrent or metastatic disease. After radical treatment close, surveillance is necessary to detect local and distant recurrence.

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How to cite this article: Sahoo TK, Dhal I, Majumdar SKD, Parida DK. Malignant Melanoma of Esophagus and its Prognosis: A Case Report and Review of Literature. *Int J Sci Stud* 2014;2(6):151-153.

Source of Support: Nil, **Conflict of Interest:** None declared.