International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Primary Malignant Melanoma of Esophagus: A Rare Entity

Dr Rajanikanth Balakrishna[1], Dr Shamila Mohamed Ali[2], Dr Reddy Polu Somashekara[3]

[1]Consultant Pathologist, Acura Diagnostic Centre and Health Care, Padmanabanagar, Bangalore.
[2]Junior Pathologist, Department of Pathology, Sagar Hospitals, Jayanagar, Bangalore.
[3]Lab Director and Head, Sagar Hospitals, Jayanagar, Bangalore.

Corresponding Author: Dr Reddy Polu Somashekara

ABSTRACT

Primary malignant melanoma of oesophagus (PMME) is a rare entity accounting only for 0.1%-0.2% of all oesophagal malignancies. Only 340 cases are reported worldwide with limited literature from India. The prognosis is poor as it is an aggressive tumor usually diagnosed at an advanced stage with metastasis. We report a case of 51 year old male who presented as a polypoidal growth in oesophagus. Total surgical excision was performed. Clinicopathological, histological and immunohistochemical examinations are discussed.

Keywords: Melanoma, Esophageal neoplasms, immunohistochemistry.

INTRODUCTION

Primary malignant melanoma of esophagus (PMME) accounts for only 0.1% to 0.2% of all esophageal malignant tumors and 0.5% of all non–cutaneous melanomas. [1] The oesophagus is a rare site for the origin of malignant melanoma with only 340 cases published till date. [2] It exhibits demographic and ethnic predisposition being detected more in caucasians. PMME is rare in Asians, particularly in Indian population with a limited literature. [3] This tumor disseminates early via blood stream & lymphatics. [1] It is a highly aggressive tumor with bad prognosis and treatment outcome is generally poor.

CASE REPORT

A 51 year old male presented with history of dysphagia for solid foods, retrosternal burning sensation and odynophagia of 2 months duration. There was no history of fever, weight loss, vomiting or alteration in bowel habits. Physical examination revealed a moderately built and nourished individual. X-ray chest and CT scan were done outside which revealed a widening at mid-1/3 of oesophagus extending up to cardiac end. Perioesophageal fat spaces were normal. Radiological and C.T impression was leiomyoma of esophagus. Endoscopy was done and biopsy was taken elsewhere and was reported as poorly differentiated carcinoma. Patient was admitted to our hospital for surgery. Trans-hiatal total esophagectomy with esophago-gastric anastomosis was done and specimen was sent to histopathology department.

Gross Examination showed an esophagectomy specimen measuring 18cm along with cardiac end of stomach. Cut section showed a polypoidal tumor at mid-1/3 arising from the wall of esophagus measuring 10x 4 cm (Figure 1). The tumor was protruding into lumen, reaching

uptocardiac end. Surface was brownish to grayish in color with focal area of ulceration. Surrounding mucosa was normal.

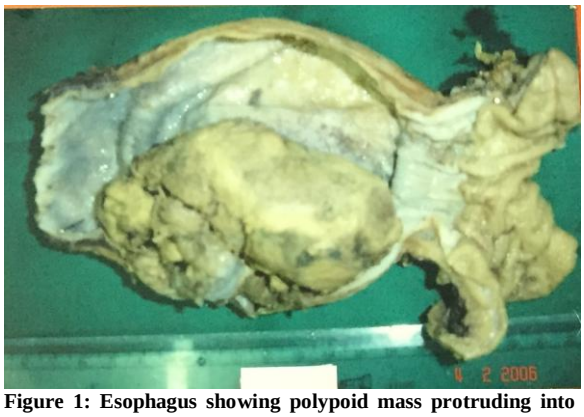

Figure 1: Esophagus showing polypoid mass protruding into the lumen. Surrounding mucosa unremarkable.

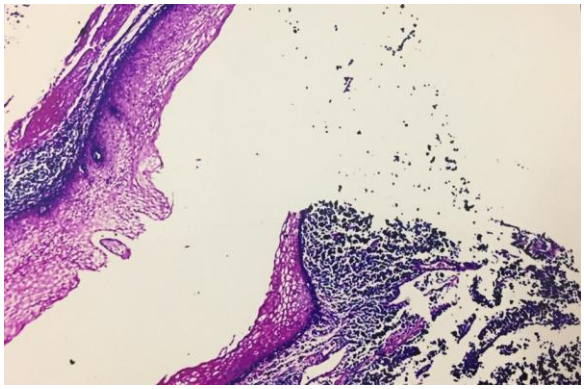

Figure2: Scanner view showing sheets of round cells covered by squamous epithelium and also showing ulceration. (Haemotoxylin and Eosin).

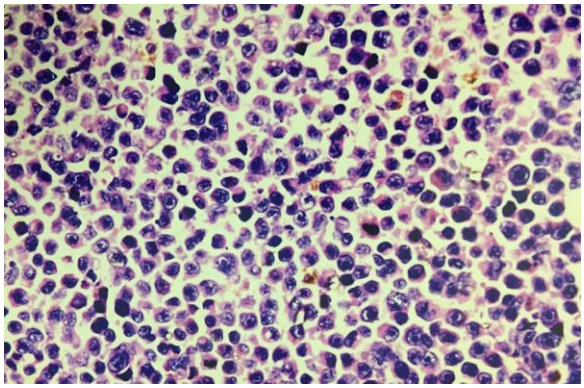

Figure 3: Cells are round with areas of pleomorphism. Hyperchromatic and irregularly clumped chromatin. Pigment sparse (Haemotoxylin and Eosin, 400X).

Microscopic Examination revealed a tumor composed of nests and sheets of round to polygonal cells with abundant clear to amphophilic cytoplasm .The nucleus was round to oval with centrally located prominent eosinophilic nucleolus. Chromatin was irregularly clumped (Figure 2&3). Melanin pigment was sparse. Tumor was mitotically active and very vascular. Tumor cells were seen involving muscle and serosal layer. Both resected margins and lymph nodes salvaged did not show tumor involvement.

On immunohistochemistry, cells were positive for S100, HMB45 and Vimentin and negative for EMA (Figure 4&5). Patient was followed up and was asymptomatic for 6 months. After one year he presented with wide spread metastasis and succumbed to the disease.

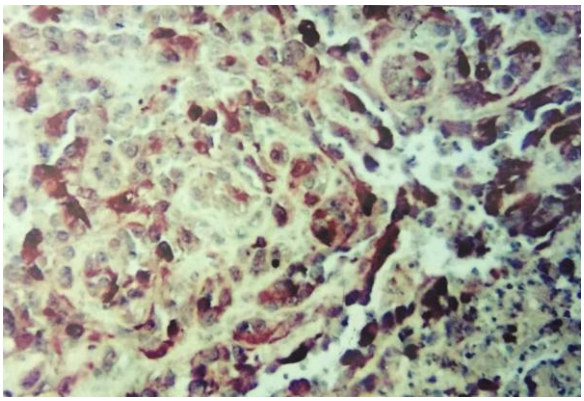

Figure 4: Immunohistochemistry-S100 cytoplasmic and nuclear positivity.

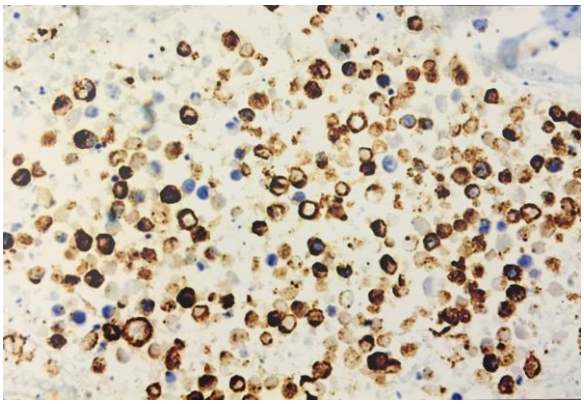

Figure 5. Immunohistochemistry-HMB45-cytoplasmic positivity.

DISCUSSION

The first case of malignant melanoma of oesophagus was reported by Baur in 1906.However, it was considered to be metastatic. This perception changed in 1963 when De pa Lava reported presence of melanocytes in esophageal epithelium. [3]

Allen and spitz defined the diagnostic of PMME.1) A typical histological pattern of melanoma and

presence of melanin granules within tumor cells 2) the origin from an area of junctional change within squamous epithelium 3) Junctional activity within melanotic cells in adjacent epithelium. [3] However, these criteria were met in only 40% of the cases reviewed by Kreuser although he suggested that tumor growth may have obliterated junctional activity in some cases. [4] In our case there was no junctional activity identified though ulceration was noted. Melanin pigment was sparsely distributed. A detailed history and clinical examination is important in distinguishing between primary and secondary disease. In this case there was no cutaneous, ocular or anal lesion. Biopsy specimen can be misinterpreted as poorly differentiated carcinoma because of the submucosal nature of the tumor and the fact that the melanoma cells may contain little or no melanin pigment as seen in this case. A number of patients have been previously misdiagnosed due to absence of melanin granules. Thus immunohistochemical markers like S-100, HMB-45 and Melan A are required for a definitive diagnosis. [3] Even though it is a rare lesion it must be considered in differential diagnosis of polypoidal lesions of oesophagus like leiomyomas, lipomas, fibroma, neurofibroma, epidermoid carcinoma, sarcoma, small cell carcinoma, carcinosarcoma and metastatic melanoma. [1] PMME arise in 6th and 7th decades with a male: female ratio of 2:1. [3] Basque et al reported the only case in a child who was 7 year old. [1]

PMME usually presents as a solitary tumor but multiple lesions have been reported in 12% of the cases. It typically presents as a polypoidal mass or well circumscribed, elevated and pigmented tumor. Although black pigmentation is an established characteristic of PMME, 10-25% of melanomas may be tan, dark brown, blue or gray depending on the melanin quantity. [3] They are non-pigmented in 20-25% of the cases. The true incidence of amelanotic melanoma is less than 2%. [1]

The neoplasms occur in distal 2/3 rd. of the oesophagus in 90 %of the cases. The clinical presentation is usually similar to that of epithelial tumors of oesophagus with dysphagia, retrosternal discomfort, weight loss, malena and hematemesis. [5] These symptoms typically manifest only a few months before diagnosis, usually less than 3 months. [1] Our patient too had short history.

The mean survival time from diagnosis is only 13.4 months and 5 year survival rate is 4.2% worldwide. [3] The outlook is dismal unless the tumor is limited to esophageal wall. Symptoms typically develop only when the tumor has become large or metastasized. At the time of diagnosis, metastatic disease is present in approximately 50% of the patient with, 31% hepatic, 29% mediastinal, 18% pulmonary and 13 % cerebral metastasis documented. [1] Recent studies recommend the 18-flourodeoxy glucose PET CT to be used as the first line modality to stage the disease and detect metastasis. [2] Risk factors are not yet defined. Melanosis, a benign condition defined as an increase in number of melanocytes in the basal layer and with increased in quantity of melanin in these melanocytes, is a predisposing factor. Oshiro et al in 2007 described a case of melanosis which transformed into malignant melanoma on follow up. [1] A higher incidence of esophageal malignant melanoma has been described in Japanese and this can be attributed to a greater number of esophageal melanocytes observed in these population. [1] Reflux of gastric juice, increase in melanocytes in hyperplastic epithelium and chronic esophagitis may act as precursor lesions for malignant melanoma. [3]

Lager et al detected mutations in C-KIT genes in two patients with PMME. Mutations in BRAF are detected in metastatic malignancies. These are currently under study as targets for molecular therapy. [6]

Surgical treatment with radical resection with wide margins is advised as tumor has a tendency to spread

longitudinally through the submucosa and satellite nodules are reported. Other modalities of treatment are radiotherapy, chemotherapy, chemoradiotherapy and endocrinotherapy. The only treatment that influence the survival is surgery where as other option is palliative or helps in loco regional control. [5]

REFERENCES

1. Machado J, Ministro P, Araújo R, Cancela E et al. Primary malignant melanoma of the esophagus: A case report. World J Gastroenterology . 2011; 17(42):4734-38.
2. Ballester AN, De Molina S D, Morales J G. Primary Malignant Melanoma of the Esophagus: A Case Report and Review of the Literature. Am J Case Rep 2015; 16:491-95.
3. Jiang W, Zou Z, Liu B. Primary malignant melanoma of the esophagus: A case report and review of the literature. Oncology Letters. 2015; 9(5):2036-40.
4. Ishizaki M, Aibara Y, Furuya K. Primary malignant melanoma of the esophagogastric junction: Report of a case. International Journal of Surgery Case Reports. 2013;4(8):700-703.
5. Yonal O, Ibrisim D, Songur Y, et al. Primary Malignant Melanoma of the Esophagus. Case Reports in Gastroenterology. 2013;7(2):272-76.
6. Hiroshi F, Ribeiro U, Sallum RA. Primary malignant melanoma of oesophagus: a rare and aggressive disease. World J Surg Oncology. 2013; 11:210.

How to cite this article: Balakrishna R, Ali SM, Somashekara RP. Primary malignant melanoma of esophagus: A rare entity. Int J Health Sci Res. 2017; 7(7):396-399.

****