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Case Report

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Primary Malignant Melanoma Oesophagus: A Rare Cause of Dysphagia

GS Zacharia[1], A Rajendran[2], A Jacob[3]

[1]Department of Gastroenterology, Ahalia Hospital, Abu Dhabi, United Arab Emirates
[2]Department of Gastroenterology, Kozhikode District Co-Operative Hospital, Calicut, India
[3]Department of Anesthesiology, Ahalia Hospital, Abu Dhabi, United Arab Emirates

Correspondence:
GS Zacharia
E-mail: george.lenx@yahoo.com
https://orcid.org/0000-0001-5705-3275

ABSTRACT

Primary malignant melanoma, though rare, is believed to be the most common non-epithelial neoplasm of the oesophagus. Compared to epithelial esophageal malignancies, melanoma is more aggressive and metastatic, resulting in poor therapeutic response and prognosis. Immunohistochemical staining is often diagnostic; conventional staining classically reveals epithelioid cells with pigment deposits. We report a case of primary esophageal melanoma in an elderly male who presented with dysphagia, treated palliatively with esophageal stenting.

Keywords: *Melanoma, Esophageal melanoma, Primary melanoma, Dysphagia*

INTRODUCTION

Primary malignant melanoma is an exceedingly rare neoplasm accounting for about 0.1% to 0.2% of all malignant esophageal neoplasms [1]. It is mostly reported in elderly males, typically present as dysphagia. The tumor typically involves the middle and lower third of the oesophagus, and about 40% are metastatic at diagnosis [2-4]. Only a few hundred cases have been reported to date, and likely owing to the rarity, there is no consensus on management. Surgical resection is considered the most accepted modality of treatment in operable cases. Despite the advancements in surgery and chemotherapy, the overall prognosis remains dismal, even in operable cases [1-3].

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CASE REPORT

A 70-year-old male patient presented with painless progressive dysphagia of three months duration associated with anorexia and weight loss. He had no history of heartburn, vomiting, dyspepsia, or overt gastrointestinal bleeding. His history was remarkable for coronary artery disease and atrial septal defect (ASD). He has thirty a thirty-pack-year history of smoking but quit two years back. The physical examination was unremarkable except for features of ASD and pulmonary hypertension. There were no skin lesions or lymphadenopathy. The hemogram revealed mild microcytic anemia and elevated erythrocyte sedimentation rate (26 mm/hour). Serum electrolytes and renal and liver function tests were within normal limits. Barium swallow revealed a fixed filling defect in the mid-oesophagus (Figure 1).

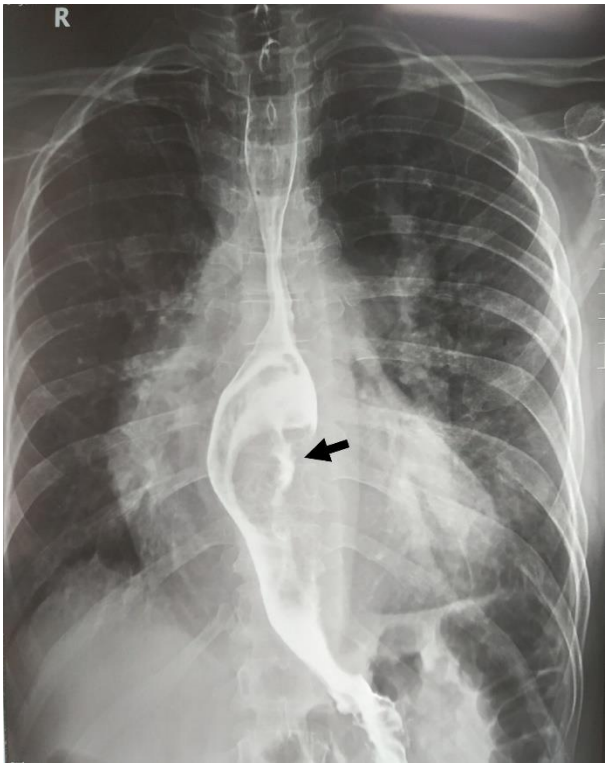

Figure 1: Barium swallow image showing a filling defect in mid-oesophagus (black arrow)

Contrast-enhanced computerized tomography (CECT) of the chest and abdomen revealed an enhancing lesion (2.5 x 3.8 x 5.8 cm) with luminal narrowing, prominent right upper paratracheal node (1.2 cm) and sub-centimeter paraaortic and bilateral hilar nodes (Figure 2).

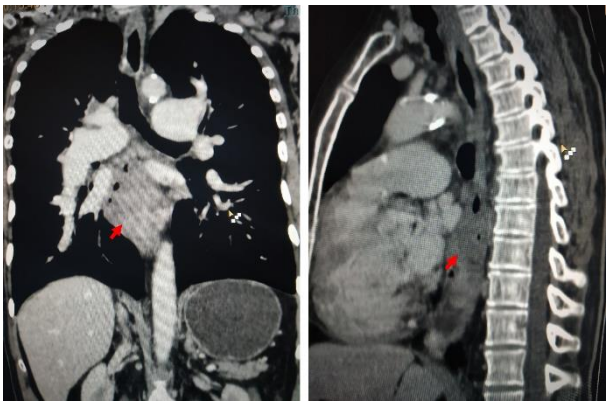

Figure 2: Contrast-enhanced CT coronal and sagittal images reveal enhancing mass lesions in the mid-oesophagus (red arrows)

Esophago-gastro-duodenal endoscopy (EGD) revealed a polypoidal mass at 33 cm from the incisors with luminal narrowing, extending five centimeters distally; the lesion was biopsied. Hematoxylin and eosin staining revealed atypical cells arranged diffusely and in sheets, suggesting poorly differentiated neoplasm (Figure 3).

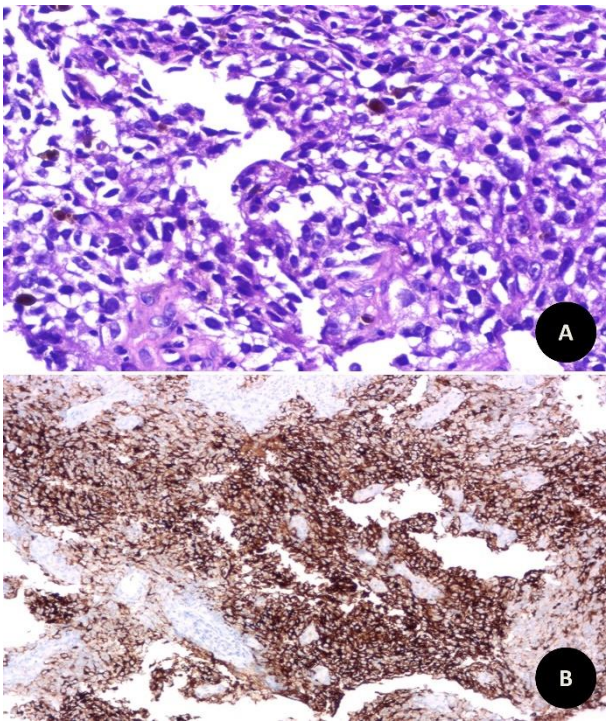

Figure 3: Hematoxylin and eosin stain (A) demonstrate atypical hyperchromatic cells in sheets. Immunohistochemistry (B) demonstrates positive HMB-45 reactivity.

Immunohistochemistry showed positive HMB-45 and cytokeratin-negative cells, which are suggestive of melanoma (Figure 3). Cardiology evaluation revealed 14 mm ASD with bidirectional shunt, severe pulmonary hypertension, and dilated right atrium and ventricle. Following a multi-disciplinary and tumor board meeting, it was decided not to proceed with surgery, considering the age, cardiac comorbidity, and regional adenopathy. The patient opted for palliation and dysphagia relief; hence, the patient underwent esophageal self-expanding metallic stenting.

DISCUSSION

Esophageal cancer is the seventh most common cancer worldwide, with about 50% of total cases reported from China [5]. More than 95% of esophageal neoplasms are epithelial in origin, primarily squamous cells, or adenocarcinoma. Melanomas commonly involve the skin but are also reported in the eyes and viscera. Primary esophageal melanoma is exceedingly rare, and only a few hundred cases have been reported in the literature.

Esophageal melanoma typically affects older people in the 6^{th} or 7^{th} decade of life with a male-to-female ratio of 2-3:1 [1]. About 90% of cases involve the middle or lower third of the oesophagus and typically present as dysphagia. [1,2,4]. Pain and bleeding are uncommon manifestations [6]. Morphologically, most lesions are polypoidal and pigmented; however, superficial, ulcerated, and non-pigmented lesions may occur. Diagnosis of primary esophageal melanoma requires histological features consistent with melanoma. Histology reveals atypical cells with abundant cytoplasm, cytoplasmic melanin deposits, heterochromatic nuclei, and high mitotic activity arranged as nests, sheets, or cords.

Nests of melanocytes and cellular atypia at the junction of mucosa and submucosa [7]. Junctional changes, nests of atypical melanocytic cells at the junction of mucosa and submucosa, are early and classical features [8,9]. Immunochemical staining improves diagnostic accuracy, without which about 40% of cases may be misdiagnosed, most often as poorly differentiated carcinoma [6]. Melanomas stain positive with HMB-45 and S-100 proteins and negative for cytokeratin or carcinoembryonic antigen [4].

Diagnostic imaging reveals esophageal enhancing mass with luminal narrowing. Magnetic resonance imaging may show high intensity in T1-weighted images due to the presence of melanin. Approximately 40% are metastatic at diagnosis, most commonly to the adjacent nodes [4]. There are no standard staging systems or treatment consensus for primary esophageal melanoma. The TNM staging for esophageal carcinoma is often adopted for melanomas, and there is limited data on its relation to survival. Radical surgery and lymph node dissection are the most accepted treatments in operable cases. The efficacy of chemotherapy, radiotherapy, novel therapies, and immunotherapy remains unclear [1,6,8]. Being highly aggressive tumors with limited therapeutic armamentarium, primary esophageal melanomas carry an abysmal prognosis, with 5-year survival rates of less than 5%. Patients with dysphagia may receive palliative esophageal stenting for symptom relief.

In line with published literature, our patient was also an elderly male who presented with dysphagia. He was misdiagnosed as poorly differentiated carcinoma on hematoxylin and eosin staining; however, he was diagnosed accurately with HMB-45 stain. Owing to advanced age, nodal involvement, and comorbidities, he was palliated with esophageal stenting, which provided him with symptom relief and improved quality of life till his death eight months later.

CONCLUSION

Primary malignant melanoma is a rare esophageal neoplasm often confirmed with biopsy and immunohistochemical staining. The tumor tends to be highly aggressive with a high propensity to metastasis. Owing to the rarity, there is a shortage of data regarding optimal management, and the prognosis remains abysmal.

Author declaration

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Authors' contributions:
Investigating, diagnosing, and managing the patient: G.S.Z., A.J., A.R.; Drafting of the manuscript: A.J.
Critical revision of the manuscript for important intellectual content: G.S.Z and A.R

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