Esophageal Melanocytosis - An Unusual Melanocytic Entity

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Abstract

Esophageal melanocytosis is a rare benign condition characterized by melanocytic proliferation in esophageal squamous epithelium and melanin deposition in the mucosa. Because of its uncommon nature, pathologists and gastroenterologists lack experience with this entity. We present a case of esophageal melanocytosis in a 66 years old male patient who presented with atypical chest pain and dysphagia. Endoscopic guided biopsy was done, provisional diagnosis of esophageal melanocytosis was made. Bleaching and immunohistochemistry confirmed the diagnosis.

Introduction

Esophageal melanocytosis is a rare benign clinicopathological entity characterized by melanocytic proliferation in esophageal squamous epithelium and melanin deposition in the mucosa.¹,² Little is known about the etiology and natural course of this condition, although it is hypothesized that it may result from chronic irritant stimuli such as gastroesophageal reflux disease, chronic esophagitis, which would cause mucosal damage and subsequent reactive melanocytic hyperplasia.² We report a rare case of esophageal melanocytosis in 66 year old male who presented with atypical symptoms.

Case Report

A 66 years old male presented with history of atypical chest pain and dysphagia since 5 months. There were no other complaints or any co-morbidity. Past and family history were not significant. Routine investigations, ECG, USG of abdomen and X-ray chest were within normal limits. Upper gastro-intestinal endoscopy showed tiny black pigmented patchy spots in the mid-esophagus. Endoscopic-guided biopsy bit was submitted for histopathological examination.

Gross morphology shared multiple tiny grey-brown to grey-black soft tissue bits altogether measuring 1 cc. Microscopic examination revealed strips of stratified squamous epithelium of esophagus with focal hyperplasia and melanin pigmentation in the basal layer. Subepithelial stroma showed edematous fibrocollagenous tissue with melanophages (Figure 1: Images 1 and 2). There was no atypia/ necrosis/mitoses. The melanin pigment was bleached completely with potassium permanganate.

Immunohistochemistry with HMB-45 was positive in the pigment present areas in the basal layer and also in the melanophages (Figure 1: Images 3 and 4).

Based on histomorphological findings and positive IHC marker, a final diagnosis of esophageal melanocytosis was made.

Discussion

Esophageal melanocytosis is a rare condition of benign melanocytic proliferation found in 0.07-2% of gastrointestinal biopsies. Like their cutaneous counterparts, basal melanocytes of esophagus lack desmosomes and tonofilaments but possess long dendritic cytoplasmic processes that extend between the keratinocytes. Esophageal melanocytosis is a rare benign condition first described by De La Pava et al in 1963.³ Its natural history is unknown and some have suggested that it may be a precursor of esophageal melanoma, although such transformation has never been reported. Esophagus does not normally contain melanocytes but aberrant migration from the neural crest may occur.⁵

During early embryogenesis, melanocytes migrate from the neural crest to the epidermis, hair follicle, oral cavity, nasopharynx, uvea, leptomeninges and inner ear. Aberrant migration of melanocytes during embryogenesis does occur in small numbers to esophagus, thus the occurrence of benign and malignant melanocytic lesions of esophagus are now well accepted.

Other postulated theories regarding esophageal melanocytosis are differentiation of the esophageal stem cells to melanoblasts and chronic stimuli causing keratinocytic hyperplasia, that can result in increased melanogenesis.⁵

The term melanosis used in previous reports, does not accurately describe the increased number of melanocytes found in this condition and does not imply that the underlying pigment is specifically melanin. Esophageal melanocytosis is endoscopically characterized by a circular, linear or oval lesions of dark brown colour with smooth surface and jagged edges.⁶ It has been documented that esophageal melanocytosis is mostly located in the middle or lower third of esophagus⁵ and is frequently associated with reactive epithelial changes such as acanthosis and basal cell hyperplasia.

Etiology of this condition is poorly understood but it has been suggested to be related to chronic irritation and inflammation. It has been associated with a number of conditions like Addison’s disease,⁷ oral melanoma and esophageal squamous cell carcinoma. But in our case, the patient had no such associations.

Differential diagnosis of esophageal melanocytosis includes benign naevi and primary malignant melanoma of the esophagus. Absence of atypia and presence of heavily pigmented dendritic melanocytes in the subepithelial stroma differentiates it from esophageal benign nevus.

Biphasic growth pattern with spindle

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and epithelioid cells, cytologic atypia, prominent eosinophilic nucleoli, brisk mitosis differentiates melanoma from melanocytosis.9

In recent review only 34 cases of esophageal melanocytosis were found in the literature, 21 of whom were Indians. A single case of esophageal melanocytosis progressing to malignant melanoma has been reported in the literature who underwent follow-up endoscopies over a period of 11 years.9

There is no report of esophageal melanocytosis that resulted in specific symptoms and special treatment.

**In conclusion, esophageal melanocytosis is a rare benign condition.** Because of its uncommon nature, many pathologists and gastroenterologists lack experience with this entity. More widespread recognition by gastroenterologist and histopathologist would be worth while for a better understanding of esophageal melanocytic lesions. Due to the presumed benign course, neither treatment nor surveillance is currently available, however follow-up endoscopies are required to observe for any malignant transformation.

**References**