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Case Report

Internal Medicine Section

Primary Spindle Cell Malignant Melanoma of Esophagus: An Unusual Finding

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ABSTRACT

Malignant melanoma of esophagus is usually a metastatic tumour rather than a primary tumour. Primary malignant melanoma accounts for less than 0.2% of all esophageal neoplasm. We report a case of primary spindle cell malignant melanoma of esophagus in a 69-year-old male who presented with history of dysphagia since 1 month. Radiological examinations revealed polypoidal growth at lateral aspect of esophagus. Biopsy was reported as grade III squamous cell carcinoma. Video assisted thoracoscopic esophagectomy was performed. Histopathological examination along with immunohistochemistry gave confirmed diagnosis of primary spindle cell malignant melanoma of esophagus. Though a rare entity, due to its aggressive nature and poor prognosis primary malignant melanoma should be one of the differential diagnoses in a patient with polypoidal esophageal mass lesion. Despite radical surgical treatment prognosis is extremely poor.

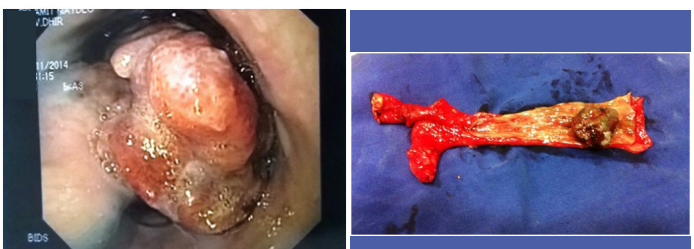
Keywords: Malignant, Melanocytes, Polypoidal mass

CASE REPORT

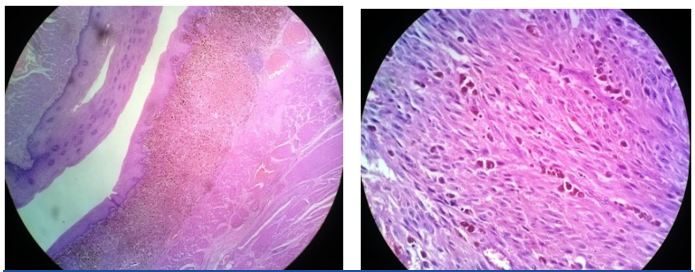
A 69-year-old male presented with complaint of discomfort and gradual progressive dysphagia especially to liquids since 1 month. Dysphagia was associated with cough, on swallowing on and off since few months. Patient also complained of burning micturition intermittently since 1 month. There was no history of vomiting, fever, haematemesis, malena or lymphadenopathy. There was no past history of cutaneous, ocular or other site melanoma. Patient was operated for appendectomy 30 years back and had no complications. Patient was a tobacco chewer since childhood. Laboratory investigations including complete blood count, liver function test, renal function test, coagulation studies were all within normal limits. Barium swallow showed mild fusiform widening involving retro cardiac esophagus with lobulated filling defect distending the lumen on mucosal release film. Esophago-gastroscopy showed polypoidal growth 3x3 cm at lateral wall of esophagus 25 cm from incisor teeth. Endoscopic ultrasound (EUS) showed mid esophageal polyp limited to submucosa with impression of Fibro vascular polyp or malignant tumour [Table/Fig-1]. Computed tomography of chest showed well defined heterogeneously enhanced lobulated 3.3x2.8x4.1 cm in retrocardiac esophagus at the level of 1.5 cm inferior to tracheal bifurcation causing near total obliteration of lumen and dilatation of proximal esophagus. Biopsy taken from polypoidal growth was reported as grade III Squamous cell carcinoma. Based on the radiological findings and biopsy report clinical diagnosis of carcinoma esophagus was made. Video assisted thoracoscopic esophagectomy with feeding jejunostomy was done and specimen was sent for histopathological diagnosis. Grossly specimen comprised of esophagogastrectomy with cut margins and various group of lymph nodes including bronchial, upper, mid and lower paraesophageal, subcarinal, paragastric, lesser curvature and left gastric nodes. The esophagus measured 12 cm in length and stomach measured 4cm & 13.5 cm along lesser and greater curvature. On cutting open brownish mucosa covered polypoid tumour noted occupying one third circumference of esophagus measuring 4x4x2.5cm [Table/Fig-2]. Histopathological examination of tumour showed spindle cell malignant melanoma infiltrating submucosa and sparing muscularis propria [Table/Fig-3&4]. Mitotic count was 4-5/10hpf. Cut margins and all lymph nodes were free of tumour. Immunohistochemistry study showed that tumour cells expressed S100, HMB45 and were negative for CK, EMA, CD34 and CD117. A final diagnosis of primary spindle cell malignant melanoma was made. Postoperative period was uneventful. No metastasis and recurrence was observed after 7 month of follow up of patient.

DISCUSSION

Primary malignant melanoma of esophagus is rare and accounts for less than 0.1-0.2% of all esophageal tumours [1]. Melanoma is considered to be a malignant tumour arising from the melanocytes of skin and eyes mainly. Melanoma arising from the mucosal lining the gastrointestinal tract, respiratory and genitourinary tract is unusual for its location. Approximately 337 cases of primary malignant melanoma of esophagus have been published in the literature [2]. Mucosal melanomas carry poor prognosis than cutaneous due to delay in diagnosis and metastatic spread. Aberrant migration



[Table/Fig-1]: Endoscopic ultrasound (EUS) showed mid esophageal polypoid mass limited to submucosa of 4x4x2.5 cms. **[Table/Fig-2]:** Gross photograph showing esophagus measured 12 cm in length. Cut section showed brownish mucosa covered polypoid tumour of 4x4x2.5 cm size noted occupying one third circumference of esophagus.



[Table/Fig-3]: Histopathological examination of tumour showed spindle cell malignant melanoma infiltrating submucosa and sparing muscularis propria (H&E,100).
[Table/Fig-4]: Histopathological examination of tumour showed spindle cell tumour arranged in fascicles with intracytoplasmic melanin pigment (H&E, 400).

of esophageal melanocytes may be attributed to development of primary malignant melanoma in the esophagus [2,3].

During development melanocytes originate from neural crest and migrate to skin, hair follicles, uvea, leptomeninges and squamous epithelium [3]. Diagnosis of primary esophageal melanoma remained doubtful because melanocytes do not migrate to esophageal epithelium usually. De La Pava et al., first described presence of scattered melanocytes at the junction of lamina propria and squamous epithelium of esophagus [4]. Esophageal melanosis is found to be a premalignant lesion in 25% of cases [5]. Both metastatic and primary melanoma are rare tumour in esophagus; metastatic more common than primary tumour. Most common site of melanoma is in the mid and lower one third of esophagus. It is usually a solitary lesion but multifocal lesions have been reported in the literature [6].

The commonest clinical symptoms of patient are dysphagia and discomfort on swallowing of short duration. Other presentations may include anorexia, abdominal or retrosternal pain, weight loss, haematemesis or malena. It occurs in 6^{th} -7^{th} decade of age group with male preponderance. Radiological examinations like barium swallow, upper gastrointestinal tract endoscopy helps in diagnosis. On endoscopic findings melanoma usually appear as an intraluminal, exophytic, polypoidal mass covered with ulcerated or intact squamous mucosa. It may or may not be obstructive and in about 85% cases lesions are pigmented [7]. But multiple, ulcerative and amelanotic lesions have been reported in the literature [8]. Amelanotic melanoma mislead to diagnosis of high grade or undifferentiated squamous cell carcinoma until unless proved by histopathological Examination (HPE) and immunohistochemistry (IHC).

Grossly most cases show intraluminal polypoidal, lobulated, brownish black mass causing obstruction of lumen of mid or lower esophagus. The mass is covered with ulcerated or intact mucosa. HPE remains the gold standard for diagnosis of melanoma. Microscopically tumour tends to grow in a lentiginous manner in the mucosal and submucosal layers. But deeper penetration into the muscle layer and lymphovascular invasion is also common. Tumour cells are epithelioid or spindle shaped arranged in nests and fascicles respectively with or without melanin pigment production. Biopsy specimen especially superficial may reported as poorly differentiated carcinoma or high grade squamous cell carcinoma if the tumour do not contain melanin pigment as occurred in our case. Special stain like Masson's Fontana, DOPA reaction and IHC plays crucial role in confirmation of diagnosis in such cases. IHC markers like S100 protein; HMB-45 and Melanin A are immunopositive and tumour cells are immunonegative for cytokeratin [7].

Differential diagnosis of polypoidal masses in esophagus includes malignant tumours like spindle cell carcinoma, leiomyosarcoma, squamous cell carcinoma and rarely adenocarcinoma. Benign lesions include squamous cell papilloma, leiomyoma, fibroma, fibrovascular polyp. These lesions can be ruled out by their characteristic histomorphological features [5,7].

Primary Malignant Melanoma of Esophagus (PMME) has to be differentiated from metastatic melanoma from other sites like cutaneous, ocular or genitourinary system. Diagnostic microscopic criteria of primary malignant melanoma proposed by Allen and Spitz [8] include a typical melanoma structure, presence of melanin pigment in the tumour cells, origin from squamous epithelium with junctional activity or junctional activity with melanotic cells in the adjacent epithelium. Metastatic spread from cutaneous melanoma to GI tract occurs commonly to small intestine, less often to stomach, colon and very rarely to esophagus [9]. PMME is a biologically aggressive tumour. Chalkiadakis G et al., revealed that at the time of presentation metastasis is present in approximately 50% of cases, 31% hepatic, 29% mediastinal,18% pulmonary, 13% cerebral and regional lymph nodes [6].

The treatment of choice for PMME is enbloc resection with regional lymphadenectomy with restoration of gut continuity. Role of utility of adjuvant or neoadjuvant radiotherapy is not proven [10]. Additional immunomodulatory therapy may be used in metastatic disease. The prognosis of PMME is poor. The mean survival time for PMME is approximately 10-14 months with a 5 year survival rate of 4-5% [11].

In our case intraluminal polypoid mass was seen in the mid-esophagus obstructing the lumen of esophagus. On the basis of endoscopic ultrasound and clinical findings and biopsy findings clinical diagnosis of carcinoma esophagus was made. No evidence of metastasis was seen on histopathological and radiological examination.

CONCLUSION

PMME is an extremely rare neoplasm of esophagus. It is biologically aggressive tumour with poor prognosis and difficult to distinguish from other esophageal malignancies.

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