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A Leiomyosarcoma of Esophagus : Report of Two Cases

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Abstract:

Leiomyosarcoma is a rare tumor of esophagus that accounts for 0.5% esophageal sarcomas. This paper presents two cases of leiomyosarcoma of esophagus in 45 years old lady and 52 years old gentleman who presented with dysphagia and they were evaluated to rule out malignancy, but they turned out to be rare leiomyosarcoma which were confirmed by immunohistochemistry. One patient underwent oesophago-gastrectomy and other was treated with self-expandable metal stent (SEMS) in view of inoperability. Case presentations, diagnostic modalities and various management options are discussed.

Key words: Leiomyosarcoma, Esophageal Neoplasms, Immunohistochemistry, Stents, Deglutition Disorders.

Introduction

Leiomyomas of esophagus are the most common esophageal mesenchymal tumors, by contrast, esophageal leiomyosarcomas are tumors that have been described anecdotally in medical literature [1]. We report two cases of leiomyosarcoma, which have been diagnosed by histopathology and immunohistochemistry (IHC) treated with different modalities.

Case Reports

Case 1

A 45 years old lady presented with 1 month history of dysphagia, 2 weeks history of fever and vomiting. Her clinical examination was essentially normal and routine biochemical, hematological parameters were within normal limits. Upper gastrointestinal endoscopy showed grade 1 esophageal varices and a doubtful lower esophageal diverticulum. CECT abdomen showed large mass in left lower thorax arising from distal esophagus. Centre of the mass was ulcerated and it was communicating with the esophageal lumen [Fig.1]. Esophago-gastrectomy was done using left thoraco-abdominal approach. Intra operatively supradiaphragmatic growth was seen in lower thoracic esophagus [Fig.2]. Cut section showed intact mucosa and a fleshy growth arising from the wall. Cavity in the center of growth was communicating with esophageal lumen. Patient had an uneventful post-operative recovery. Histopathology was suggestive of leiomyosarcoma

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of esophagus with spindle cells arranged in sweeping fascicles and intervening bundles. Mitotic activity of 2/10 Hpf was noticed. All nodes examined were reactive and the resected margins negative for malignancy. Immunohistochemistry showed strong positivity for SMA and desmin with negative CD 34 and CD 117 [Fig.3]. Patient is asymptomatic; tolerating feeds and is on regular monthly follow up since last 6 months.

Case 2

52 years old gentleman was admitted with complain of dysphagia for 3 months. Clinical examination was normal. Biochemical and hematological parameters were within normal limits. Upper gastrointestinal endoscopy showed a smooth narrowing in esophagus from 28-34 cms. CECT abdomen showed a large heterogenous mass in the subcarinal esophagus causing luminal narrowing and proximal dilatation along with periesophageal extension with bilateral bronchial and aortic infiltration [Fig.4]. CT guided percutaneous biopsy showed features of malignant spindle cell tumor. Mitotic activity of 4/10 Hpf was also noticed. Immunohistochemistry showed strong positivity for SMA and desmin with negative CD 34 and CD 117 suggestive of leiomyosarcoma. Since the tumor was unresectable, covered self-expandable metal stent (SEMS-Nitinol-S) was placed to palliate dysphagia and radiotherapy(45 Grays) given. Now patient tolerating semisolid food without difficulty and he is in regular follow up.

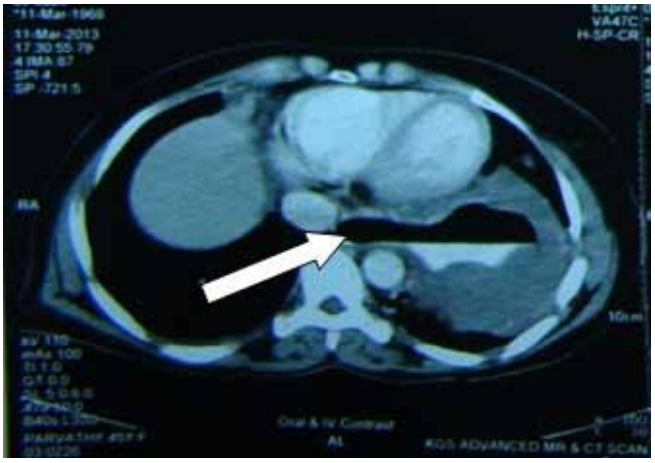

Fig.1: CECT Abdomen showed large mass in left lower thorax arising from distal esophagus. Centre of the mass was ulcerated and it was communicating with the esophageal lumen.

Discussion

Leiomyosarcoma of the esophagus are rare malignant smooth muscle tumors. These tumors are commonly seen after 6th decade, in lower 1/3rd of esophagus, with equal sex predilection [2]. Generally presenting complaints are dysphagia or odonophagia [3]. As per Rainer and Braus leiomyosarcomas are divided into intra luminal polypoidal (most common, 60%) or an infiltrative, invasive form (40%) [4]. In our first case polypodal growth was seen while second case was invasive type.

Preoperative workup includes upper GI endoscopy which shows an exophytic growth with intact mucosa, often looking like diverticulum due to cavity in the center of tumor which was classically observed in

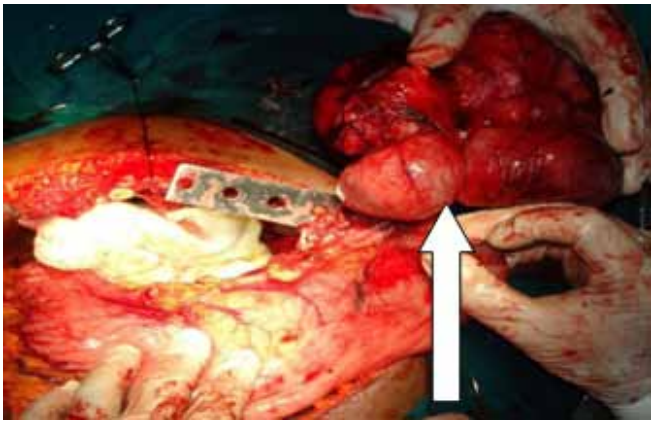

Fig.2: Intra operative photograph showing mass in lower end of the esophagus.

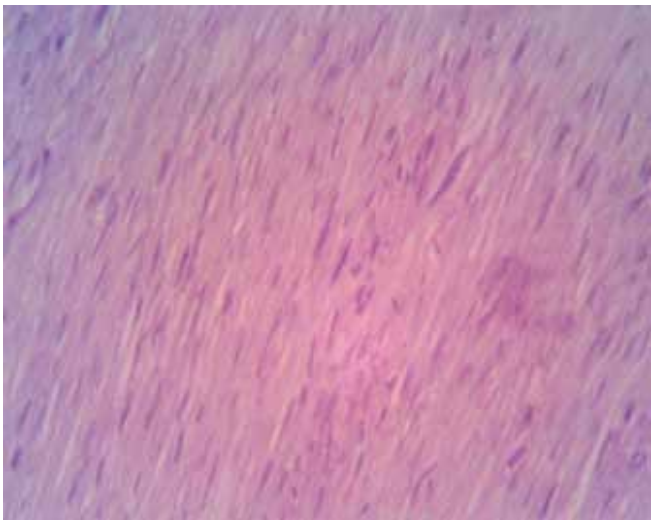

Fig.3: 40X microphotograph showing spindle cells arranged in long fascicles (H&E Stain).

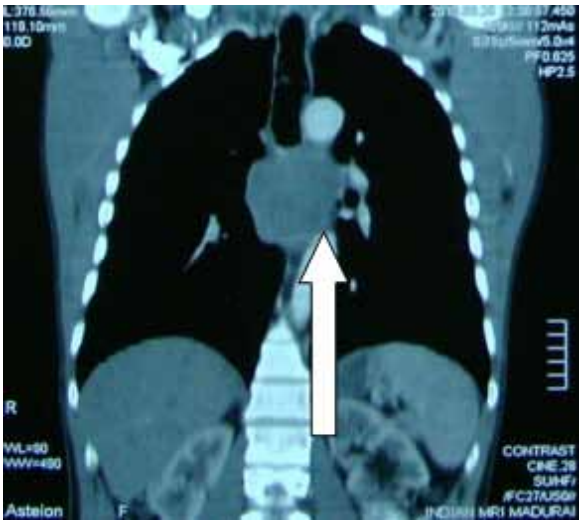

Fig.4: CECT abdomen showed a heterogenous mass in subcarinal esophagus causing luminal narrowing and proximal dilatation. Periesophageal extension with bilateral bronchial and aortic infiltration is also seen.

our first case. Endoscopic ultrasound and biopsies are generally noncontributory [5]. Barium swallow shows irregular intramural mass containing giant central ulcer communicating with esophageal lumen. CT chest shows heterogenous exophytic lesion with central areas of low density and extraluminal gas or contrast material within the tumor [6] as in first case. The best and confirmative diagnostic tool is histopathology and immunohistochemistry which differentiates leiomyosarcoma from leiomyoma by presence of size >5 cm, cellular atypia, presence of necrosis, no. of mitosis (>2/10 Hpf) and histologically tumor composed of spindle cells arranged in long fascicles. IHC will differentiate these from GIST by presence of CD34 and CD117 negativity, presence of desmin, vimentin, S100 positivity [7]. In our two cases biopsy showed features of malignant spindle cell tumor with significant mitotic activity. Immunohistochemistry showed strong positivity for SMA and desmin with negative CD 34 and CD 117. The significant survival advantage achieved with curative resection makes esophagectomy or esophago-gastrectomy the standard modalities of care [8]. The finding of metastatic disease at presentation however should not contraindicate surgical excision of primary mass. In view of significant palliative and long term survival, the other options can be considered especially if local invasion of aorta and bronchus have occurred. The literature review shows role of radiotherapy in inoperable cases, but role of chemotherapy is negligible. Endoscopic stenting is standard of care for palliation of malignant dysphagia and esophago-respiratory fistulae. Self-expandable metallic stents (SEMS) are effective, easy to insert with fewer complications. Covered SEMS are recommended owing to less tumor ingrowth. Though we could not find any evidence in literature regarding role of SEMS in leomyosarcoma, as it is malignant dysphagia, we tried this in second case which was inoperable. In combination with radiotherapy, probably it will palliate malignant dysphagia well. The 1, 3 and 5 year survival rates have been reported at 60.3%, 42.8% and 32.1%, respectively, in a recent meta-analysis [9]. Death due to disease is usually because of local recurrence or distant metastases to the lungs or liver.

Conclusion

We hereby report rare cases of esophageal leiomyosarcoma presenting with dysphagia and were confirmed with histopathology and IHC. Surgical resection is the standard of care in presence of even metastasis. Inoperable cases require tissue diagnosis by CT guided biopsy, and palliation by SEMS and radiotherapy.

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