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Case Report

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Esophageal Leiomyosarcoma: Insights from Three Cases and Literature Review

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Abstract

Esophageal leiomyosarcoma, constituting a mere 0.5% of esophageal sarcomas, stands out as an exceedingly rare form of esophageal tumor[1]. This paper delves into distinct instances of esophageal leiomyosarcoma in three patients, each presenting with unique clinical challenges. The spectrum of presentation, underpinned by radiological assessment, confirmed through histopathological evaluation highlights the imperative of an interdisciplinary approach in identifying and treating this rare condition. The ensuing discussion sheds light on diagnostic nuances, therapeutic strategies, and the indispensable value of radiological imaging in evaluating patients of leiomyosarcoma with rare metastatic involvement to the brain. This contribution aims to augment the existing compendium of knowledge on esophageal leiomyosarcoma, advocating for heightened clinical vigilance and a nuanced understanding of its management.

Keywords: Leiomyosarcoma; Esophageal Carcinoma; Dysphagia; sarcoma; spindle cells; submucosal tumors.

Introduction

Esophageal carcinoma is primarily diagnosed as squamous cell carcinoma or adenocarcinoma, which account for more than 95% of cases. However, esophageal sarcomas such as carcinosarcoma and leiomyosarcoma are uncommon, affecting only 0.1-1.5% of cases. The most common sarcoma is carcinosarcoma, followed by leiomyosarcoma. Carcinosarcoma was first defined by Virchow in 1865 as a tumour with mixed epithelial and spindle cell characteristics, with a better prognosis than squamous cell carcinoma of the oesophagus due to its mixed nature[2].

Esophageal leiomyosarcoma is a pure sarcoma of mesenchymal origin that often occurs in the middle and lower oesophagus, where smooth muscle is abundant, with esophageal leiomyosarcoma in the upper esophagus being an extremely rare occurrence[3]. Goodner et al. found only seven cases of leiomyosarcoma among 1456 (0.5%) patients with esophageal cancer[4]. Rocco et al. discovered just 19 cases of esophageal cancer among 6359 patients (0.3%) at the Mayo Clinic over a 76-year span[4]. Such information helps to explain the rarity of this esophageal tumour. Approximately half of all leiomyosarcoma cases occur between the sixth and seventh decades of life. There is slightly male preponderance(1.55:1,male:female)[4]. Leiomyosarcomas consist of interlacing whorls of spindle cells with pleomorphism. Sarcomatous transformation in a previously existing leiomyoma is uncommon, with only four examples reported thus far. Histopathology and immunohistochemistry are used to diagnose leiomyosarcoma, which is distinguished from leiomyoma by tumour size (>5 cm), cellular atypia, necrosis, mitosis (>2/10 HPF), and

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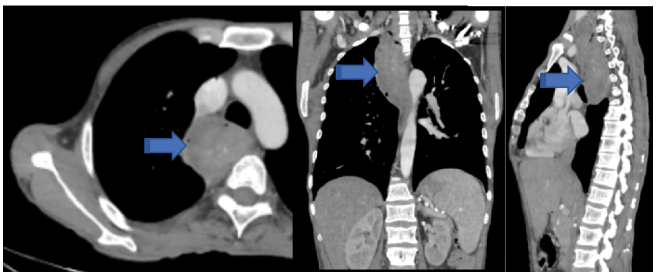
spindle cell arrangement[5]. We hereby discuss three separate cases of esophageal leiomyosarcoma in three different patients, each with unique clinical presentation.

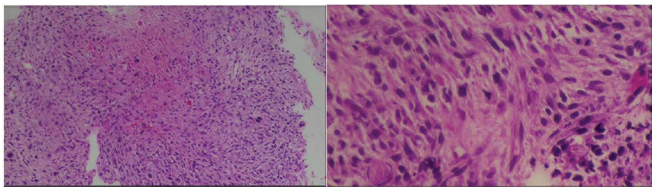
Case Study

Case 1

A 45-year-old male's battle with dysphagia, fever, and vomiting culminated in the discovery of a significant esophageal mass through contrast enhanced computed tomography (CECT) and upper gastrointestinal endoscopy, leading to thoraco-abdominal esophagogastrectomy. Despite a successful operation, the patient's journey was abruptly halted by unforeseen complications.



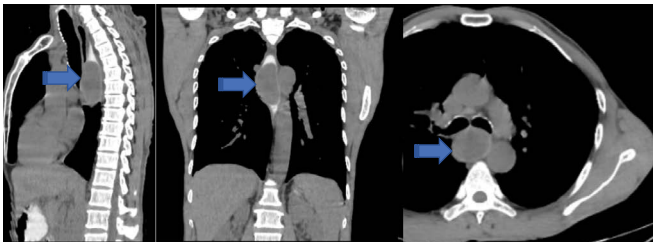
On CECT abdomen, an expansile inhomogeneous enhancing intraluminal esophageal soft tissue mass lesion (arrow head) involving the lower cervical esophagus, upper and mid thoracic esophagus is noted extending from lower border of C7 vertebral body upto upper border of D8 vertebral body causing near complete obliteration of the lumen. The lesion was seen compressing and displacing the adjacent parenchyma of right lung laterally.



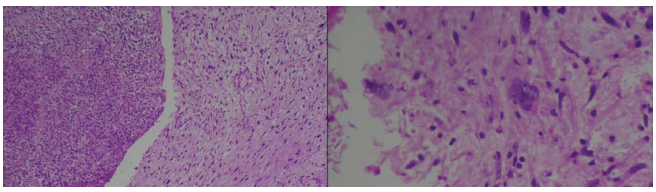
On histopathological examination, a tumor composed of spindle shaped cells with plump, blunt ended hyperchromatic nuclei and moderate pale to brightly eosinophilic fibrillary cytoplasm are seen. The cells are set in long intersecting fascicles parallel and perpendicular to the plane of the section. Mitotic figures, including atypical ones, are present.

Case 2

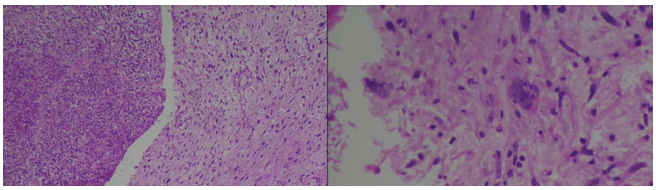
The ordeal of a 57-year-old male, marked by prolonged dysphagia, was met with a similar diagnostic and treatment pathway. This patient also underwent upper gastrointestinal endoscopy followed by a CECT thorax. As of the time of writing this paper, the patient had been free of recurrence for 15 months. His resilience through surgery and an extensive radiotherapy and chemotherapy regimen has set a course towards recovery, navigating the challenges of postoperative life.



CECT abdomen revealed a well-defined hypodense, soft tissue attenuating ovoid intraluminal mass lesion (arrow head) showing heterogenous post contrast enhancement is seen in the thoracic esophagus with lower margin being approximtely ~ 12 cm from the gastro- esophageal junction. The mass is almost completely occluding the lumen of the oesophagus & causing its expansion at the same level and anteriorly causing mild indentation over the trachea.



On histopathological evaluation, spindle cells with deep hyperchromatic nuclei were seen.



Case 3

A 48-year-old female, initially presenting with neurological symptoms like headache and altered metal status, unveiled the rarity of esophageal leiomyosarcoma's presentation. The initial misdiagnosis as high-grade glioma highlights the diagnostic challenges and the critical role of imaging in such atypical presentations. In this case, the absence of dysphagia/odynopahgia, weight loss and other typical signs was highly unusual and did not initially suggest esophageal leiomyosarcoma.

The patient underwent contrast enhanced magnetic resonance imaging of Brain which revealed a well defined altered signal intensity mass lesion with heterogeneous post contrast enhancement and surrounding white matter T2/ FLAIR hyperintensity in the right parietal lobe appearing hypointense on T1, predominantly hyperintense on T2/ T2 FLAIR, showing areas of restriction on DWI with corresponding defect on ADC and small foci of blooming on

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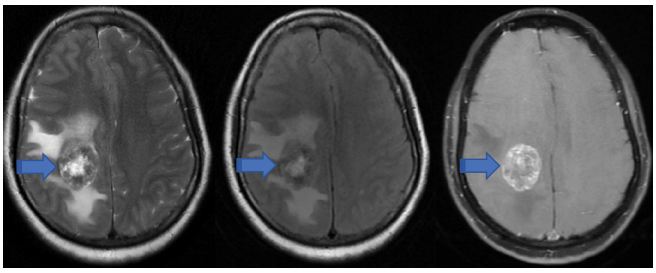
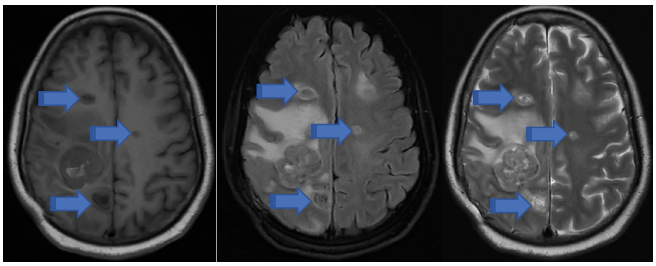
SWI. There was also mass effect in the form of effacement of adjacent sulco-cisternal spaces and right lateral ventricle with a midline shift of ~ 8.6 mm towards left side. A diagnosis of high grade glioma was suggested.



The patient subsequently underwent a follow-up CEMRI Brain on 4 months later which revealed multiple areas of altered signal intensity mass lesion with surrounding edema and mass effect consistent in bilateral cerebral hemispheres. A possibility of metastases with unknown primary was considered. The patient expired on 6th October, 2023 due to unknown reasons. This paper highlights the need for thorough diagnostic evaluation before concluding primary disease, especially when therapeutic decisions are affected.



Discussion

Esophageal leiomyosarcomas are extremely rare sarcomas of the esophagus with only few cases reported thus far. Rainer and Braus divided leiomyosarcomas into two types: intraluminal polypoidal (60%), and infiltrative invasive (40%)[5]. Individuals with polypoid and intramural tumours, intrathoracic tumours, and well differentiated tumours have a better prognosis than those with infiltrating lesions, cervical tumours, and poorly differentiated tumours[4]. Long-term survival rates for leiomyosarcomas is higher than those for esophageal squamous cell carcinoma. Esophageal leiomyosarcoma associated with squamous cell carcinoma is extremely rare, with only a few examples reported since Ovens et al. first characterised it in 1951[6]. Diagnosis can be difficult, particularly when differentiating the main tumor from metastases. Furthermore, intermutability or metaplasia between mesenchymal and epithelial tissues may occur, confounding the diagnostic process[6].

Leiomyosarcomas are distinguished by their huge size, slow growth, and limited penetration into adjacent structures, which frequently leads to late recurrence. However, in certain cases, such as the one reported by Koga et al., despite favourable characteristics, rapid tumour development and poor outcomes occurred, presumably due to the influence of squamous cell carcinoma on leiomyosarcoma growth via cytokines or growth factors[7]. Dysphagia and painful swallowing are the most common presenting symptoms in 78% of patients, making cases without them extremely rare. Other common symptoms include weight loss (54%), chest pain (36%), and regurgitation. In rare cases, respiratory symptoms caused by tracheo-bronchial tree compression, epigastric pain, vomiting, gastrointestinal bleeding due to ulceration, and anaemia may be present.

Leiomyosarcomas may spread locally to the lungs, pleura, pericardium, diaphragm, and stomach, or distantly to the liver, lungs, and bone. The brain and inferior vena cava are uncommon locations for distant metastases. Overall, one-third of leiomyosarcomas are known to have metastasized at the time of presentation; the pathological type, size, and location of the tumour are not predictive of metastases[4]. A barium swallow can reveal expansile intraluminal masses in polypoidal tumours, as well as irregular areas of luminal constriction in patients with infiltrative tumors. These are classified as large intramural masses with ulceration and/or tracking[4]. Endoscopic examinations typically reveal polypoid and exophytic masses, with ulcerative lesions and malignant strictures being less common findings. Endoscopy has limits in diagnosing esophageal leiomyosarcomas, particularly those with an intact mucosa covering intramural and exophytic tumours[8]. Endoscopic ultrasonography, on the other hand, can reveal typical features such as a hyperechoic mass, which can help with the diagnosis. Computerised tomography (CT) images reveal heterogeneous lesions with prominent exophytic components and low-density core regions free of surrounding structures. Paraesophageal nodes could be involved. Extraluminal gas or contrast material within the tumour may be detected if the lesion interacts with the oesophageal lumen or if the necrotic section of the tumour cavitates[4]. Magnetic resonance imaging (MRI) can detect an isointense mass in T1-weighted images and a hyperintense mass in T2-weighted images[4]. The use of FDG-PET-CT in identifying leiomyosarcoma has been described, with these tumours exhibiting substantial FDG absorption[7].

Historically, surgical resection was the primary treatment for malignant submucosal tumors (SMTs), but advances in endoscopic methods have made minimally invasive endoscopic resection more desirable. Endoscopic ultrasonography (EUS) is critical for evaluating SMTs since it provides information on size, layer of origin, echo pattern, and interior features, however the accuracy varies between observers. Biopsy is advised for SMTs larger than 2 cm and can be performed using several endoscopic procedures, including fine-needle aspiration or Tru-Cut biopsy, with varying diagnostic

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accuracy. Endoscopic submucosal surgery, which includes endoscopic submucosal dissection (ESD), endoscopic unroofing, and other procedures, has allowed for the identification and treatment of esophageal SMTs. Endoscopic correct muscle excision using an over- the-scope clip (OTSC) and interdisciplinary treatments combining endoscopy and thoracoscopy have showed promise in the management of larger SMTs[9]. Endoscopic stenting, particularly with self-expandable metallic stents (SEMS), is the norm for treating malignant dysphagia and esophageal-respiratory fistulas. Covered SEMS are favoured for reducing tumour growth[5]. Hatch et al. discovered in the largest ever study of smooth muscle tumours of the oesophagus that mortality associated with surgery for oesophageal leiomyosarcoma is substantial in the immediate postoperative period due to pulmonary and anastomotic complications[3].

This may underline the need of treating esophageal leiomyosarcomas with a less intrusive surgical method, even when the tumour is larger. Despite the fact that these tumours can be up to 16 cm in size and frequently present as mediastinal masses, a case series by Zhang et al. and others indicates that many have expansive growth with little neighbouring organ infiltration, supporting the use of less invasive procedures such as thoracoscopic mobilisation for esophageal leiomyosarcoma. This approach, as demonstrated in a patient with a 17 x 8 cm (length x width) tumour by Manipadam et al. and others, led to easier postoperative recovery and fewer complications compared to open surgery, underlining the potential benefits of minimally invasive techniques for managing these challenging tumours[3].

There is no standardized treatment procedure for esophageal sarcomas. Surgical removal, pioneered by Harrington in 1945, has yielded favorable results, prompting many to urge for esophagectomy as routine treatment[10]. It is believed that the presence of metastatic disease at presentation does not rule out surgical resection, as good palliation and even long-term survival rates have been reported. The resection of metachronous metastases in the liver has also been described. The Mayo Clinic reported 5-year survival rates of around 80%, with little further benefit from adjuvant chemotherapy or radiotherapy[10]. A recent meta-analysis revealed one-, three-, and five-year survival rates of 60.3%, 42.8%, and 32.1%, respectively, with disease- related fatalities frequently caused by local recurrence or distant metastases to the lungs or liver[5].

Radiotherapy may aid patients who are not candidates for surgery, and long-term survival has been recorded with local excisional surgery, indicating that endoscopic resection is a viable choice for intraluminal polypoid leiomyosarcomas[8]. The rarity of these tumours makes it difficult to provide definite therapeutic guidelines; nonetheless, better reporting of these unique tumours would benefit in understanding their behaviour and optimising treatment regimens.

This section critically examines the complexities surrounding esophageal leiomyosarcoma, emphasizing the pivotal role of radio-imaging in diagnosis. Despite its rarity, the condition's variable presentation necessitates a high degree of suspicion and a comprehensive diagnostic approach. The discussion further explores the therapeutic avenues undertaken, including the paramount importance of multi- disciplinary approach in metastatic scenarios and the potential benefits of adjuvant radiotherapy[7] and chemotherapy.

Conclusion

Our findings underscore the imperative of recognizing esophageal leiomyosarcoma's potential for atypical presentation and the crucial role of a multidisciplinary approach in its management. These cases serve as a testament to the challenges and complexities inherent in diagnosing and treating rare esophageal tumors, advocating for an amalgamation of clinical vigilance, advanced diagnostic strategies, and personalized therapeutic regimens to navigate this labyrinthine condition.

By drawing from these cases, we aim to elucidate the condition's clinical manifestations, radiological features, and histopathological characteristics, thereby contributing to the scarce literature and enhancing disease recognition and management strategies.

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