CASE REPORT

Complete Excision of Rare Esophageal Tumor; Leiomyosarcoma

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ABSTRACT

Leiomyosarcoma are rare malignant esophageal tumors that remain asymptomatic unless size exceeds 5 cm. They grow at a slow rate and in very few cases have metastasized by the time of their presentation. Here, we present a case of a 65 years old male with progressive dysphagia and occasional dull chest pain for one and half years. Clinical examination was unremarkable. Computed tomography showed an 8 cm mass of the upper and middle thoracic esophagus with lateral wall thickness of 1.5 cm. Fat planes were intact with enlarged subcarinal nodes. Biopsy was suggestive of leiomyosarcoma and Mckeown esophagectomy was performed. Intraoperatively, mass was consistent with radiological findings, however it was adherent to surrounding structure with enlarged carinal nodes. Patient had an uneventful recovery period with histopathological evidence of complete recession of tumor.

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INTRODUCTION

Leiomyosarcomas are among the rare malignant smooth muscle tumors of the esophagus. They make up to 0.5% of all esophageal malignancies[1]. Considering the rarity of tumor, only 165 cases of leiomyosarcomas have been reported till date[2]. Patients are often affected at or after fifty years of age with a slight male preponderance having 2:1 of male to female ratio[3]. As a result of their slow growth rate and late metastasis, they have good survival rate. Diagnosis and management of leiomyosarcomas are challenging as they are rare and no well established guidelines are present[4].

CASE REPORT

A 65 years old male presented to outpatient department with history of progressive dysphagia for one and half year. Initially dysphagia was for solids which later progressed to liquids over twelve months. Patient also complained of occasional dull chest pain with a feeling of sticking sensation in the center of the chest. There was no relevant history of weight loss or regurgitation. Family members reported a change in his dietary habits and frequency of meal over the year.

Patient underwent flexible endoscopy that identified a polypoid mass extending into the lumen with overlying normal mucosa. Biopsy showed spindle cells with spindle shaped hyperchromic nuclei with mild to moderate nuclear pleomorphism, few mitotic figures, and h-Caldesmon positive.

Computed Tomography(CT) revealed a large mass starting from the upper and extending upto the middle thoracic esophagus. Mass was 8 cm in length with lateral wall thickness of 1.5 cm and enlarged carinal nodes (Figure 1). All metastatic work up was negative. After consultation with multidisciplinary team, surgery was advised as preoperative chemoradiotherapy had no role.

After optimization and obtaining informed consent, McKeown esophagectomy was carried out. Operative findings included a large bulky mass extending from lower one third of upper thoracic esophagus up to the junction of lower and middle esophagus. Esophagus was found adherent to the mediastinum and posterior membrane of the trachea. Stations 7 and 8 nodes (subcarinal and esophageal respectively) were visibly enlarged. Tumor was resected completely along with lymphadenectomy without iatrogenic injury to any

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underlying structure. Patient was managed postoperatively in the ward with early commencement of enteral feed via feeding jejunostomy. Nasogastric tube was removed on the third day followed by chest tube removal on day 5. The patient was allowed orally on the 10th postoperative day and discharged without any complication.

At follow up, the patient had no active complaints. Histopathology was consistent with leiomyosarcoma of esophagus showing spindle cells arranged in intersecting fascicles and intervening bundles. Mitotic activity of 2/10 Hpf was noticed. All nodes examined were non-reactive and the resected margins negative for malignancy. Radiological assessment post-surgery showed no evidence of anastomotic leak or metastasis.

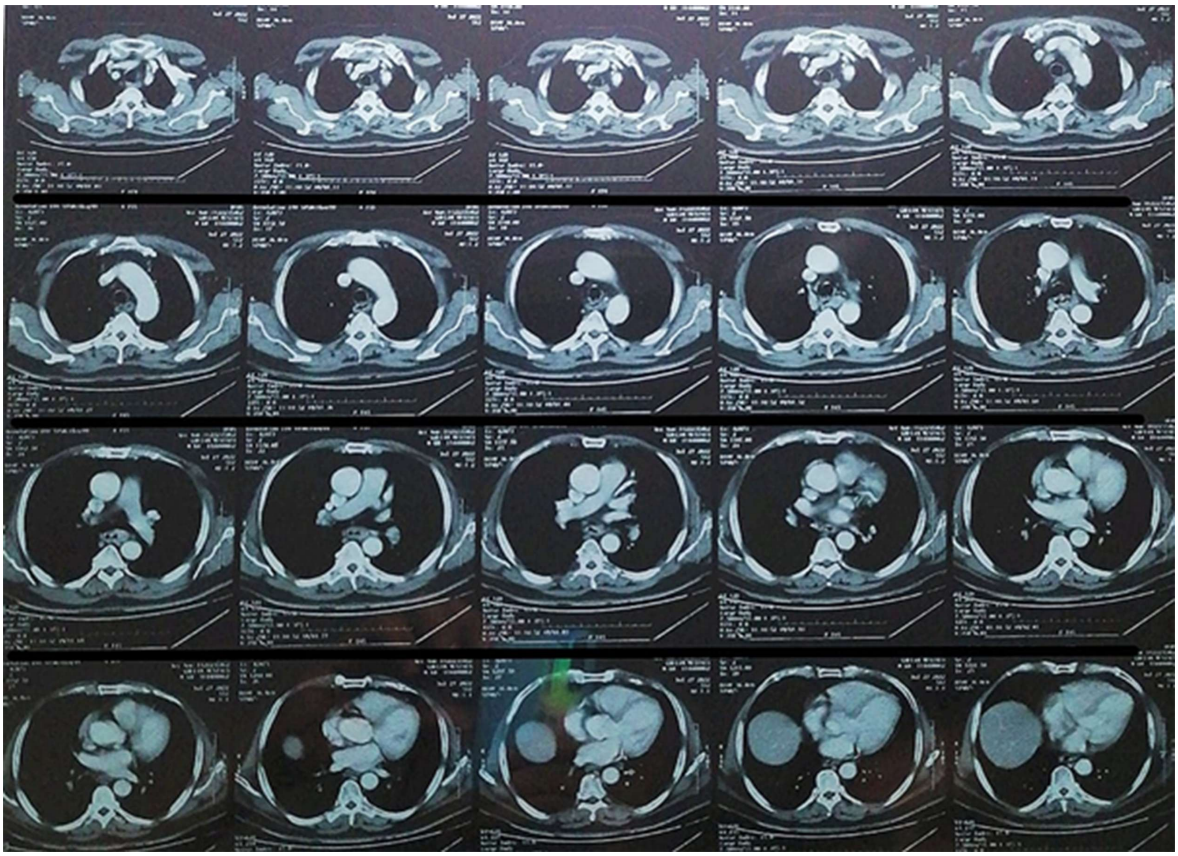

Figure 1: CT scan of leiomyosarcoma

DISCUSSION

Leiomyosarcomas are malignant smooth muscle tumors originating from mesenchymal cells of muscularis propria of the esophagus. Macroscopically, they appear polypoidal, remaining intramural, however in selective cases, they may be infiltrating in nature[3]. In our case, mass was intramural in location. The malignancy has a higher predilection for males in the sixth decade of life[4]. Leiomyosarcomas usually affect middle and lower esophagus as per literature but our patient presented with the involvement of upper and middle esophagus[3,6].

The commonest symptom at presentation is usually dysphagia which occurs in three-fourths of the patients[1]. Other symptoms include retrosternal pain, weight loss, and regurgitation. The tumor has indolent growth pattern due to which patients modify their dietary habits as seen in our case; ultimately one-third of them present with metastasis[3]. They metastasize to the surrounding structures such as lung, pleura, pericardium, diaphragm, and to distant sites as well, such as stomach, liver, and bones[1]. As per literature, 5-year survival is only 20% after metastasis[4].

The accurate diagnosis preoperatively is difficult as differentiation between leiomyoma and leiomyosarcoma is only possible by histopathological examination. Till date, preoperative differentiation of leiomyoma and sarcoma have been possible in few cases only[5]. Differential diagnosis includes spindle cell sarcoma and carcinosarcoma of the esophagus[6,7].

Transthoracic en-bloc esophagectomy with radical lymphadenectomy should be the best surgical option to achieve complete resection. Significant survival

advantages have been achieved with local excision surgery for well localized lesions[1,7]. Role of adjuvant chemotherapy and radiotherapy remains controversial[1,3]. Few authors recommended radiotherapy for treatment of metastatic tumors to prolong survival in cases exhibiting extensive or unresectable metastases[4,6]. In literature, survival rate over 5 to 10 years is 47% and 31% respectively[3].

CONCLUSION

Any patient presenting with prolonged dysphagia should be thoroughly investigated. Although rare but leiomyomas/sarcomas should be given due attention and resected in surgically fit patients for better histopathological assessment. Surgical resection via esophagectomy remains standard treatment. Although leiomyosarcoma exhibits poor sensitivity to radiation, the tumor may be effectively controlled by increasing the radiation dose appropriately.

Conflict of interest: Authors declare that there is no conflict of interest.

Authors' Contributions: SN prepared draft and literature search; TA contributed to the conceptualization and provided financial guidance; MM oversaw the case reviewed edited the draft; AA, RZ and MQ assisted with the review and literature search.

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