Case Report

A Rare Case of Esophageal Leiomyoma

Dr. Vikram P. Mehta*, Dr. Prashant V. Mehta, Dr. Apoorva Chugh*****

*Assistant Professor, **Assistant Professor, ***Third Year Resident Doctor

Department of General Surgery, B.J. Medical College and Civil Hospital, Ahmedabad , Gujarat, India

Keywords : Esophageal leiomyoma, leiomyosarcoma, benign tumors of esophagus.

ABSTRACT

The majority of esophageal submucosal tumor is benign, of which leiomyoma is the most common type, accounting for about 90%.The development of endoscopic ultrasonography and thoracoscopic techniques has opened the door for the diagnosis and treatment of esophageal leiomyoma widely.[1,2]

Esophageal leiomyomas are the most common benign mesenchymal tumors of the esophagus. These tumors originate in the smooth muscle cells. The etiology and pathogenesis are unclear.[3,4]

They are rare lesions that constitute less than 1% of esophageal neoplasms. Approximately two-thirds of benign esophageal tumors are leiomyomas; the others are usually cysts and polyps.

We came across one such rare case in a 10 year old child diagnosed as esophageal leiomyoma for which Ivor Lewis Esophagectomy with gastric pullup was done.

CASE REPORT

A 10 year old boy presented with difficulty in swallowing solid food for past one month. He had no associated history of vomiting, fever, weight loss or appetite loss. On examination he was vitally stable and abdominal examination was unremarkable. All blood investigations were normal. Ultrasonography abdomen was unremarkable. HRCT Thorax showed dilated air and content filled esophagus in its entire extent from D1 – D12 vertebral level with maximum diameter of dilated esophagus measuring approximately 54mm at D8-D9 vertebral level. Dilated esophagus shows thickened , irregular wall with abrupt cutoff noted at D10 vertebral level. UGIscopy revealed dilated upper esophagus with some liquid residue. Luminal compromise due to external compression seen in lower esophagus from 30 to 36 cms from incisors with GE junction seen at 36 cms from incisors. Scope could be negotiated across it with some manouvering. Extrinsic bulge seen in cardia of stomach. CECT Abdomen and Thorax revealed possibility of primary malignant mass involving mid and lower third of esophagus and OG junction most likely leiomyosarcoma. EUS guided biopsy showed a submucosal gastro-esophageal lesion likely spindle cell neoplasm-leiomyoma.

Ivor Lewis Esophagectomy was done. Right posterolateral thoracotomy done and tumor involving lower third of esophagus removed. Stomach tube constructed based on right gastric and right gastroepiploic vessels. Feeding jejunostomy (FJ) done.

Patient was started on FJ feeding on postoperative day 3. Patient was started on oral feeds on postoperative day 8. Postoperative period was uneventful and patient was discharged on postop day 15. Histopathological examination which showed benign spindle cell tumor-Intramural leiomyomatosis longitudinally involving distal esophagus wall, GE junction and part of gastric wall. Distal cut end (gastric cut end) is free of tumor. Proximal cut end (esophageal cut end) shows involvement by leiomyomatosis.

Patient came for follow up after 15 days and was symptomatically improved. Patient was followed up every month.

DISCUSSION

Leiomyomas are the most common benign mesenchymal tumors of the esophagus. There is a higher incidence of leiomyoma of the esophagus in men when compared to women ratio of 2:1. The most frequent occurrence is usually between the ages of 20 to 50 years. The most typical location of these tumors is in the lower two- third of the esophagus. The occurrence of these tumors in the upper one-third of the esophagus accounts for only 10% of all leiomyomas of the esophagus. Majority of the tumors are detected when they are less than 5 cm in size. Rarely they grow to be larger than 10 cm and then, they

Correspondence Address : Dr. Apoorva Chugh
Phase 1, PG hostel, Room A 304 B. J. Medical College, Asarwa-380016, Ahmedabad, Gujarat, India • Email : apoorva.chugh29 @gmail.com

are called giant leiomyoma of the esophagus. These occur as single or multiple tumors. Leiomyomas of the esophagus rarely cause symptoms when they are smaller than 5 cm in diameter. When these tumors grow larger , they become more symptomatic in patients. The most common symptoms are dysphagia , chest pain, vague retrosternal discomfort, heartburn, and occasional regurgitation. Rarely they can cause gastrointestinal bleeding when the tumor erodes through the mucosa. There appears to be no consistent pattern of symptoms according to the anatomical location of the tumor.[5,,6] Giant leiomyomas of the esophagus may present as a mediastinal mass.

Leiomyoma of the esophagus is known to be a slow growing tumor with low malignant potential. The most frequent site of occurrence of this tumor is the lower two-thirds of the esophagus, and its distribution reflects the relative amount of smooth muscle cells present along the wall of the esophagus. When the leiomyomas situated in the distal esophagus reach a large size, they can press on the cardia of the stomach.

On histopathological examination, the esophageal leiomyomas appear as circumscribed lesions composed of intersecting fascicles of bland spindle cells with abundant cytoplasm. These well differentiated smooth muscle cells which are of the spindle type are arranged as braids. This tumor is thought to have a low malignant potential.

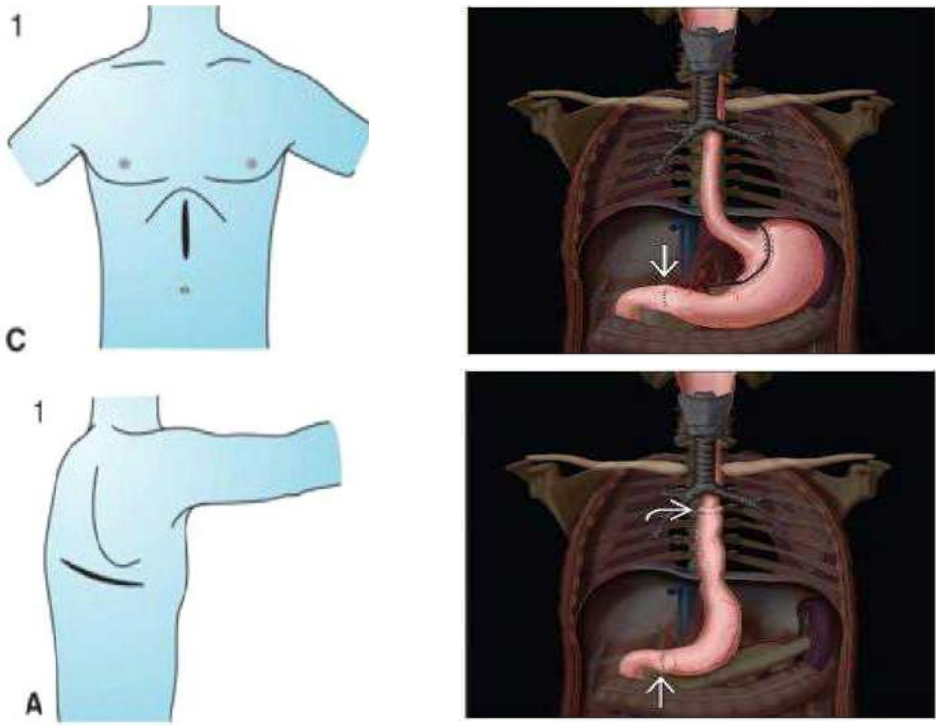
Often a diagnosis of esophageal leiomyoma is made as an incidental finding during routine investigation or screening for upper gastrointestinal (GI) pathology. In a Barium contrast series of the upper GI tract, an esophageal leiomyoma will be seen as a filling defect projecting into the lumen of the esophagus.

When an upper GI endoscopy is performed , these tumors can be identified as relatively mobile submucosal swellings protruding into the lumen of the esophagus, with normal looking mucosa covering the swelling. Currently Endoscopic ultrasonography (EUS) has become a critical investigation for diagnosis of esophageal leiomyoma.

Diagnosis of esophageal leiomyomas requires both endoscopic and radiologic examinations. Treatment depends on tumor size and location.

Once the clinical diagnosis of leiomyoma is established, tumor size and location are important, but also the patient's symptoms, general condition, and comorbidities should be taken into account. The surgical indications for tumor removal include unremitting symptoms, increased tumor size, mucosal ulceration, histopathologic diagnosis, and facilitation of other surgical procedures. A leiomyoma should be removed when diagnosed even

IVOR LEWIS ESOPHAGECTOMY



when asymptomatic, because there is always the possibility, rarely though, of malignant transformation.

Endoscopic approaches appear possible in case of small pedunculated tumors of 2-4 cm originating from the muscularis mucosae. Usually endoscopic mucosal resection (EMR) or endoscopic submucosal dissection (ESD) are done. Symptomatic small leiomyomas <5 cm can be enucleated either by open surgery or by means of video-assisted thoracoscopy (VATS). Transthoracic extramucosal blunt enucleation via a left- or right-sided thoracotomy is the most common procedure for small- to midsized esophageal leiomyoma, which is easier, faster, and safer compared to resection . Low tumors and tumors of the esophago-gastric junction can be approached via upper midline laparotomy. After enucleation, the muscular wall should be closed to avoid diverticular-like mucosal bulging and for the preservation of the muscular propulsing activity. The muscular wall should be repaired with pedunculated pleural film, diaphragm valve, or omentum, lung, pericardium. For giant esophageal leiomyoma esophageal resection and reconstruction is preferred. To perform partial or subtotal resection of the esophagus, and esophagogastric anastomosis, the results are satisfactory. Minimally invasive approaches or video - assisted thoracoscopic surgery (VATS) have progressively gained acceptance in the last few years.

REFERENCES

1. Inderhees S, Tank J, Stein HJ, Dubecz A. [Leiomyoma of the esophagus : A further indication for robotic surgery?] Chirurg. 2019 Feb;90(2):125-130.
2. Zhu S, Lin J, Huang S. Successful en bloc endoscopic full-thickness resection of a giant cervical esophageal leiomyoma originating from muscularis propria. J Cardiothorac Surg. 2019 Jan 21;14(1):16.
3. Tu S, Huang S, Li G, Tang X, Qing H, Gao Q, Fu J, Du G, Gong W. Submucosal Tunnel Endoscopic Resection for Esophageal Submucosal Tumors: A Multicenter Study. Gastroenterol Res Pract. 2018;2018:2149564.
4. Chiu PWY, Yip HC, Teoh AYB, Wong VWY, Chan SM, Wong SKH, Ng EKW. Per oral endoscopic tumor (POET) resection for treatment of upper gastrointestinal subepithelial tumors. Surg Endosc. 2019 Apr;33(4):1326-1333.
5. Peng W, Tang X, Fu X. Submucosal tunneling endoscopic resection of a large esophageal leiomyoma using endoscopy with near-focus mode. Dig Endosc. 2018 Sep;30(5):680
6. Tomulescu V, Stanescu C, Blajut C, Barbulescu L, Droc G, Herlea V, Popescu I. Robotic Approach in Benign and Malignant Esophageal Tumors; A Preliminary Seven Case Series. Chirurgia (Bucur). 2018 Mar-Apr;113(2):202-209.