

The results of surgical treatment of distal esophageal leiomyomatosis performed distal esophagectomy

Distal özofajektomi yapılan distal özofageal leiomyomatozislerde cerrahi tedavi sonuçları

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SUMMARY

Objective: Leiomyomas are the most common benign esophageal neoplasms. Surgical treatment is the therapy of choice for such tumors. In this study, we submitted results of 6 patients with distal esophageal leiomyomas who underwent surgical treatment during a 5-year period. The aim of the current study was to evaluate the long-term outcomes of patients with leiomyoma located at distal esophagus who had undergone distal esophagectomy and esophagogastrectomy.

Method: Demographic features, clinical and laboratory findings including symptoms, diagnosis, histopathology, and results of surgical treatment were examined.

Results: 6 patients (four women, two men; mean age 48.8y) were operated due to leiomyoma located at the lower esophagus. The most common symptoms were dysphagia and dyspepsia. Distal esophagectomy and esophagogastrectomy was performed to all patients with a median laparotomy and a right posterolateral thoracotomy. All patients were relieved from their symptoms without postoperative mortality, while pneumonia was occurred in one patient which is treated medically.

Conclusions: Esophageal leiomyoma is a benign tumor which becomes symptomatic in large sizes. Surgical resection is considered to be effective for distal esophageal leiomyomas, without complications.

Keywords: Esophagus, leiomyoma, distal esophagectomy

ÖZET

Amaç: Leiomyomalar en sık görülen benign özofageal neoplazilerdir. Cerrahi tedavi bu tümörlerde seçilen tedavi şeklidir. Bu çalışmada, 5 yıllık bir sürede distal özofageal leiomyoma nedeniyle cerrahi uygulanmış 6 hastanın tedavi sonuçlarını sunduk. Çalışmadaki amacımız distal özofagus yerleşimli leiomyoması olup distal özofajektomi ve özofagogastrectomi uygulanmış 6 hastanın uzun dönem sonuçlarını değerlendirmektir.

Yöntem: Demografik özellikler, semptomlar, tanı, histopatolojiyi içerecek şekilde klinik ve laboratuvar bulgular ve cerrahi tedavi sonuçları incelenmiştir.

Bulgular: Distal özofagus yerleşimli leiomyoma nedeniyle 6 hasta (4 kadın, 2 erkek; ortalama yaş 48.8 yıl) opere edilmiştir. Disfaji ve dispepsi en sık görülen semptomlardı. Tüm hastalara median laparotomi ve sağ posterolateral torakotomi ile distal özofajektomi ve özofagogastrectomi uygulanmıştır. Hiçbir hastada mortalite gelişmemiş olup tüm hastaların semptomlarında iyileşme olmuştur; bir hastada pnömoni gelişmiş olup medikal olarak tedavi edilmiştir.

Sonuç: Özofageal leiomyoma benign bir tümör olup büyük boyutlara ulaşınca semptomatik hale gelir. Distal özofageal leiomyomalarda cerrahi rezeksiyon etkili bir yöntemdir.

Anahtar sözcükler: Özofagus, leiomyoma, distal özofagus

INTRODUCTION

Benign tumors are uncommon pathologies with an estimated incidence of 1% in all esophageal neoplasms in the literature¹. Leiomyoma is the most common of benign tumours (67%-80%) of the esophagus^{1,2}. It arises from smooth muscle cells and the incidence is reported to be between 0.005% and 5.1% in large autopsy series^{3,4}. It is located mainly in the lower and middle thirds of the esophagus and, in most cases they are single lesions⁵. Leiomyoma can be diagnosed at any age while 90% of cases are between 20-69 years. The peak incidence is usually in the third to fifth decades⁶. The male-to-female ratio is approximately 2:1^{6,7}. Most of them are smaller than 5 cm in-diameter and leiomyomas larger than 10 cm are defined as giant leiomyomas^{2,4,5}. About half of the patients with leiomyoma are asymptomatic and malignant change is rare. The main symptoms are usually dysphagia and epigastric pain, but they are not specific for the disease. Endoscopic appearance is that of a mass covered by normal mucosa⁸.

The need to resect or not a leiomyoma is based on the symptoms it produces and on the differential diagnosis with Gastrointestinal Stromal Tumor (GIST)⁹. This report summarized the operative experience of distal esophageal leiomyoma performed resection in our department.

MATERIAL AND METHODS

The medical records of 6 patients with esophageal leiomyoma, operated between 2009 and 2014, were retrospectively analyzed. All patients underwent preoperative workup including upper gastrointestinal barium swallow, endoscopy, endoscopic ultrasonography (EUS) and dynamic computerized tomography (CT) scanning of the chest and abdomen.

Demographic features, clinical and laboratory findings including symptoms, diagnosis, histopathology, and results of surgical treatment were examined. Fine needle aspiration was not performed in any case.

RESULTS

There were six patients (two male and four

female; 27-70 years, mean age 48.8 yr); all patients were symptomatic whereas 4 patients suffered from dysphagia, two from dyspepsia and one from cough. All patients were subjected to the standard examinations, which demonstrated an esophageal tumor with features compatible with leiomyoma. The tumor is located at the distal third of the esophagus in all cases. In all cases, the masses were resected completely with a distal esophagectomy and esophagogastrostomy. Pyloroplasty was added to the procedure in all patients. In one patient, first of all, enucleation was applied but perforation was developed, then because of the wide mucosal defect the surgeon was decided to apply segmental resection. Frozen sections showed typical leiomyoma of the esophagus. In five patient lesions were single but in one patient there were six lesions in a short segment of esophagus; so we decided to resect effected segment of esophagus. The minimum size of tumor was 4cm, the maximum was 11 cm. The mean in-hospital staying was 9 days. Post-operative follow-up did not reveal any mortality related to the primary diagnosis; pneumonia was occurred only in one patient and was alleviated with medication. Mean follow up period was 41, 6 months (14-66 months). Four patients were relieved from their symptoms, after surgical removal of the tumor. In follow up period, LA-grade A esophagitis was established in the other two patients, but these patients relieved from their symptoms by using of proton-pump inhibitors. Clinical presentation, diagnostic findings and management of the patients are summarized in Table 1.

DISCUSSION

Although leiomyoma is an uncommon pathology, it is the most common benign tumor of esophagus. It may be present in all parts of esophagus with an incidence 60 % in the distal third, 30% in the middle, and 10% in the proximal esophagus¹⁰. In this study, all lesions were localized in the distal third of esophagus and three of them reached under the z-line.

These tumors are usually found in middle age and are almost twice as common in men⁷. Cystic or calcific degeneration may

occur in leiomyoma but malign transformation is rare¹¹. Leiomyomas grow slowly, and half of the patients are asymptomatic unless the tumor grows intramurally^{2, 12}. Approximately one-half of all tumors are smaller than 5 cm, but 5% may be larger than 10 cm⁷. Dysphagia with concomitant epigastric pain or retrosternal burning usually appears when the tumor's diameter becomes larger than the critical point of 4.5-5 cm¹³. In contrary to gastric leiomyomas, the bleeding of esophageal leiomyomas is rare because they do not ulcerate¹⁴. This presentation includes two men and four women with mean age of 48, 8 years. The most common symptom was dysphasia, which was present in four cases. Only one lesion's size was 4 cm, three of them were larger than 10 cm and in one patient multiple lesions were detected.

Leiomyoma has a radiological appearance characteristic of concave images and clear edges¹². Endoscopic ultrasonography should be combined with esophagogastros-copy to exclude esophageal cancer¹⁵. CT is useful for determining the tumor's size and its relationship with neighboring organs¹⁶. Differential diagnoses include foreign op-pressed disease of esophagus, malignant esophageal tumors such as squamous or adenomatous carcinomas or leiomyosarcoma and other benign tumors. There are controversies about the need of preopera-tive biopsy for diagnosis^{13, 14}. In cases of ulcerated mucosa and suspicion of cancer, endoscopic biopsy is prescribed¹⁷. We evaluated our patients with upper gastroin-testinal barium swallow, endoscopy, endo-scopic ultrasonography and dynamic CT scanning of the chest and abdomen. Our policy is not to recommend endoscopic biopsy, because if we decide to enucleate the lesion the tumor is easily adhesive to the mucosa and the mucosal damage oc-curs accidentally during enucleation. In addition, accurate histopathological diag-nosis could not be established with biopsy in many cases, especially for submucosal lesions.

Surgical treatment of leiomyomas is an-other debating issue. Some authors rec-ommend resection in symptomatic patients and observation in asymptomatic patients with lesion smaller than 5 cm¹⁸, while oth-

ers suggest resection even if it is asymp-tomatic due to possibility for transfor-mation to malignancy¹³. We suggest that a leiomyoma should be removed when the patient is symptomatic or the lesion is larger than 3 cm even though the patient is asymptomatic. Histological and immuno-histochemical examination is important to distinguish leiomyomas from GIST. Leiomyomas are not immunoreactive for c-kit, CD117 and CD34 in contrary of GIST^{4, 5, 16}.

There are many treatment options depend-ing on location and size of the tumor. En-doscopic mucosal resection or endoscopic submucosal dissection is reported to be effective in the treatment of small pedun-culated tumors of 2-4 cm originating from the muscularis mucosae^{14, 20}. Symptomatic small leiomyomas <5 cm can be enucleat-ed either by open surgery^{13, 18} or by means of video-assisted thoracoscopy (VATS)¹⁹. Myotomy of esophagus and extramucosal enucleation of the leiomyoma is the stand-ard and established procedure¹³⁻¹⁸. Leiomyomas at the proximal and middle third of esophagus can be resected via right thora-cotomy while transhiatal approach is effec-tive for the leiomyomas located at the distal third^{4, 10}.

Large tumors and tumors located at gas-troesophageal junction can be treated with surgical resection of esophagus; the choic-es of this method depend on several fac-tors^{7, 14}:

- 1) The esophageal leiomyoma is too large to allow a resection without damaging the mucosa,
- 2) Large leiomyomas locate in the distal esophagus and extend downward across the cardia,
- 3) For giant esophageal leiomyoma, it may have leiomyosarcoma-like transformation or with small leiomyosarcoma-like lesions and,
- 4) Multipl leiomyomas in a short segment of esophagus. To perform partial or subtotal resection of the esophagus, and esophagogastric anastomosis, the results are satis-factory.

This operation is typically performed with

esophagectomy and gastric pull-up through left or right posterolateral thoracotomy. Gastric pull-up procedure is associated with complications including reflux esophagitis, stricture formation, dumping, diarrhea, reduced meal capacity, and weight loss²¹. So, while some authors don't prefer pyloroplasty after resection²², others prefer esophageal replacement with colon for the leiomyomas located in the upper third of esophagus and if the mucosa is damaged by the lesion⁷. We suggest that the resection must be done for large tumors and multiple tumors in a short segment of esophagus. In our presentation, four patients had a leiomyoma larger than 5cm and three of them were giant leiomyoma. One patient had multiple lesions and one patient had a leiomyoma smaller than 5 cm but we couldn't differentiate from GIST by EUS, preoperatively. In all patients, the

lesions were resected using a thoracoabdominal approach, and a distal esophagectomy and esophagogastrostomy were performed. Pyloroplasty was added the procedure in all patients. All of the six patients recovered approvingly with no mortality and resumed normal diet after operation. In one patient, pneumonia was occurred and was treated with medication. In follow-up period, we established esophagitis in only two patients; their symptoms were alleviated by using of proton-pump inhibitors. In conclusion; once the clinical diagnosis of leiomyoma is established, the surgical management should be defined according to the patients' suffers and lesion characteristics. Currently, resection via thoracoabdominal approach is recommended for large and multiple tumors located at the distal-esophagus.

Table1: (EGS: esophagogastroscopy; EUS: endoscopic ultrasonography ; PPI: proton-pump inhibitors; LA-A esophagitis :classification of esophagitis based on LA Classification).

Sex	Age (year)	Symptoms	Diameter(cm)	Localization (EGS-EUS)	Treatment	Follow up period (month)	EGS findings and usage of PPI
F	27	Dysphagia, caught	10x5,5x5	32-40	Ivor-Lewis	14	LA-A esophagitis; PPI(+)
F	44	Dyspepsia	11x5x4,5	31-40	Ivor-Lewis	18	Normal; PPI(-)
M	64	Dysphagia	6 lesions, ranging from 3 mm to 3cm	30-38	Ivor-Lewis	46	Normal; PPI(-)
M	70	Dyspepsia	5,5x3,5x2	40-under z-line	Ivor-Lewis1	47	Normal; PPI(-)
F	44	Dysphagia	4x3x2	40-under z-line	Ivor-Lewis2	59	Normal; PPI(-)
F	44	Dysphagia	11x4x2,5	36-under z-line	Ivor-Lewis	66	LA-A esophagitis; PPI(+)

¹First of all enucleation was applied but perforation was developed; then we decided to apply resection.

²Couldn't differentiate from GIST before operation and lesion was located at the GEJ

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