

doi: <http://dx.doi.org/10.11606/issn.1679-9836.v97i3p00-00>

Esophagus leiomyoma with abnormal epidemiological characteristics: case report

Leiomioma de esôfago com características epidemiológicas anormais: relato de caso

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Pereira FB, Leão B, Leli LF, Lima MPL. Esophagus leiomyoma with abnormal epidemiological characteristics: case report / *Leiomioma de esôfago com características epidemiológicas anormais: relato de caso*. Rev Med (São Paulo). 2018 May-June;97(3):00-00.

ABSTRACT: *Introduction:* Despite its rarity, the leiomyoma of the esophagus is the most common non-epithelial benign tumor in this organ. *Case Report:* The case of a 73-year-old woman diagnosed, initially, with esophageal mass to be clarified, oligosynthetics, was reported. A thorax CT scan revealed the presence of a tumor in the posterior mediastinum, coming from the proximal wall of the esophagus with extraluminal growth. A high digestive endoscopy and a biopsy of the lesion were performed, but turned out to be inconclusive. We chose surgical management through a right thoracotomy, with the esophageal tumor enucleated and the esophageal wall reconstructed. The pathology and immunohistochemical confirmed the diagnosis of a esophagealleiomyoma. *Conclusion:* The case has atypical epidemiological characteristics, such as the age group, the gender and location of the leiomyoma of the esophagus. Moreover, the literature review emphasizes the rarity of this pathology, which is usually an incidental finding in imaging or endoscopic exams, being quite a challenge not only diagnostically, but also therapeutically.

Keywords: Leiomyoma; Esophagus; Neoplasms/diagnosis; Esophageal diseases; Thoracic surgery.

RESUMO: *Introdução:* Apesar da raridade, o leiomioma de esôfago é o tumor benigno não-epitelial mais frequentemente encontrado neste órgão. *Relato de Caso:* Relata-se o caso de uma paciente de 73 anos diagnosticada, inicialmente, com massa esofágica a esclarecer, oligossintomática. Realizou-se uma TC de tórax que evidenciou a presença de tumoração em mediastino posterior, proveniente da parede proximal do esôfago de crescimento extraluminal. Foi realizada uma endoscopia digestiva alta com biópsia da lesão, porém essa foi inconclusiva. Optou-se por conduta cirúrgica, através de uma toracotomia direita, sendo o tumor esofágico enucleado e a parede esofágica reconstruída. O anatomopatológico e imuno-histoquímico confirmaram o diagnóstico de leiomioma esofágico. *Conclusão:* O caso possui características epidemiológicas atípicas, sendo elas a faixa etária, o sexo e localização do leiomioma de esôfago. Além disso, a revisão bibliográfica enfatiza a raridade desta patologia, que geralmente é um achado incidental em exames de imagem ou endoscópicos, sendo um desafio diagnóstico e terapêutico.

Descritores: Leiomioma; Esôfago; Neoplasias/diagnóstico; Doenças do esôfago; Cirurgia torácica.

Scientific Paper presented at the poster session at the III International Congress of General Surgery FAMERP / FUNFARME, São José do Rio Preto, SP, 22-24/09/2016. Abstract published in the annals of the Congress. Scientific Work presented at the poster session of the IV Academic Medical Conference of the Faculty of Medicine of the University of Franca, Franca, SP, 10/21/2016.

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INTRODUCTION

Despite its rarity, the esophageal leiomyoma is the non-epithelial benign tumor most frequently found in this organ. It represents only 0.5-0.8% of esophageal tumors^{1,2}. The tumors originate from the muscularis propria layer and also, albeit rarely, from the muscularis mucosa. They occur more frequently in men than in women, and between the ages of 20 and 59, rarely occurring in children³.

The preferred location is in the middle and lower segments of the esophagus. Appearing in different sizes and hardened consistency; presents smooth, regular and, in some cases, multi-lobular surface; covered by normal-colored mucosa, with no continuity solution, sliding freely over the lesion, unless there is erosion or apical ulceration. The malignant transformation is rare, if it occurs.

They are generally asymptomatic, found incidentally in routine digestive endoscopies, or until they reach a larger size, when symptoms such as dysphagia, retrosternal pain, heartburn, coughing, odynophagia, weight loss and digestive bleeding occur⁴.

The endoscopic diagnosis is presumptive, since it is a lesion of the stomach wall, not of endoscopic diagnosis and, therefore, limited by the method, not by the professional. Hardly ever can a histological diagnosis be confirmed in such case, since the biopsies do not reach the layer of origin of the lesions, except in cases where ulceration exists and the muscular tissue is exposed¹. In virtue of its myogenic nature, the election method for confirmation of diagnosis is the endoscopic ultrasonography (EUS). One can easily locate the muscularis propria (fourth layer) of the esophagus where the leiomyoma most frequently originates, and presents itself as a hypoechogenic and homogeneous lesion. When there is doubt over the benign nature of the myogenic lesion, the histological diagnosis of the lesion may be done through echo-guided biopsies^{5,6}.

In general, the leiomyoma does not need resection. This treatment is indicated in symptomatic cases instead, or when there is doubt about the nature of the lesion, in which case the enucleation of the lesion is done through thoracotomy or videothoracoscopy^{6,7}. The endoscopic resection is of difficult execution and with risk of perforation and hemorrhage, reserved only for when the origin is the muscularis mucosa (second layer of the esophagus), well-documented by endoscopic ultrasonography and sometimes, with echo-guided resection^{2,8}.

CASE REPORT

N.R.B, 73 year-old, Caucasian female patient, born and residing in Franca, investigated for community-acquired pneumonia, enlargement of mediastinum was incidentally found in thorax x-ray. Patient procured pneumologist for investigation of radiological finding, a thorax TC and later an upper digestive endoscopy (UDE) were solicited, to investigate the possibility of mediastinal tumor. As symptomatology, the patient presented occasional complaint of dysphasia for solid foods and episodes of choking, as well as previous treatment for gastroesophageal reflux disease (GERD).

The computerized tomography evidenced an expansive lesion with a predominant component of soft tissue with calcifications compressing the esophageal lumen in her upper thoracic third, measuring approximately 2.5cm by 2.5cm by 3.5 cm. Mediastinal linfadenomegalia was not detected.

The endoscopic examination revealed esophagus with normal mucosa, with extrinsic compression of the proximal esophagus, with an extension of approximately 7.0 cm, non-adherent to the esophageal mucosa. The biopsy of the proximal esophagus was carried out, and through microscopic examination, fragments of esophageal mucosa of habitual thickness were detected, and in the chorion, lymphoplasmocitary inflammatory infiltrate of irregular distribution.

The thoracic surgeon and patient's treatment decision was for the resection of the tumor. A posterolateral right thoracotomy was performed between the 5th and 6th intercostal spaces. When entering the thoracic cavity, a solid tumoration was observed from the proximal esophagus wall at the level of the azygos vein. After the ligation of the azygos vein, a thorough dissection of the esophagus was executed, proceeded by a total enucleation of the lesion without opening the esophageal mucosa. After the hemostasis revision, a suture of the esophageal wall was done with no complications.

The piece was sent to the Pathological Anatomy Service, where the esophageal leiomyoma diagnosis was confirmed, through anatomopathological and immunohistochemical profile.

In the post-operative the patient developed nosocomial pneumonia. Antibiotic therapy was prescribed. The patient was discharged after 10 days of hospitalization and was oriented to return in ambulatory consultation to follow-up the case.

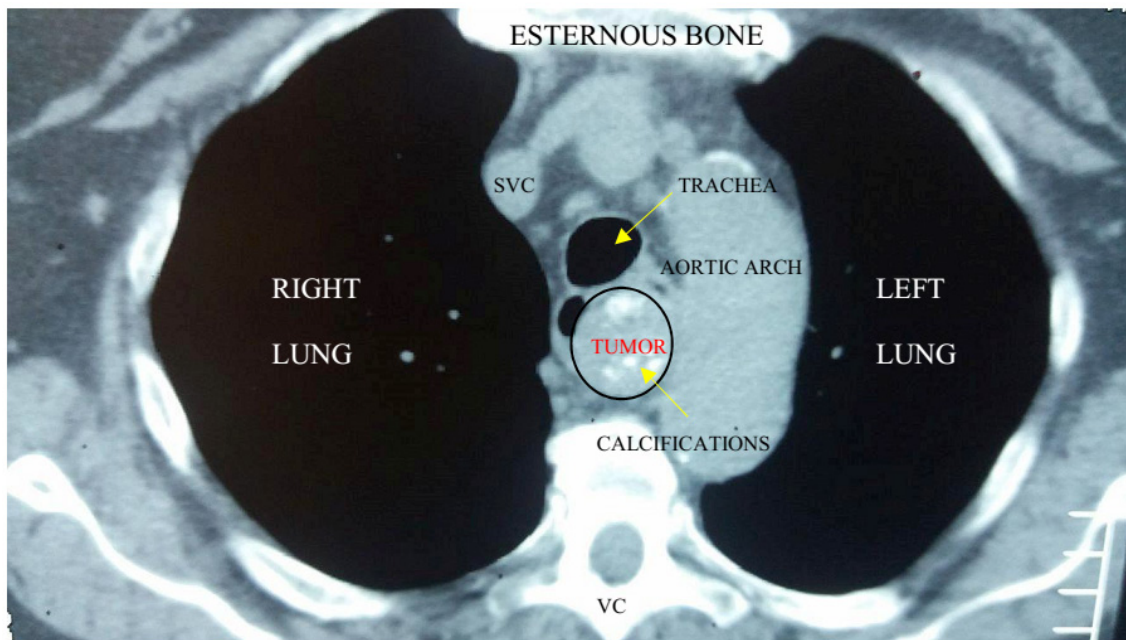


Figure 1. Cut Axial CT showing tumor composed soft tissue with calcifications, compressing the esophageal light



Figure 2. Endoscopic view of the extrinsic compression in the proximal esophagus

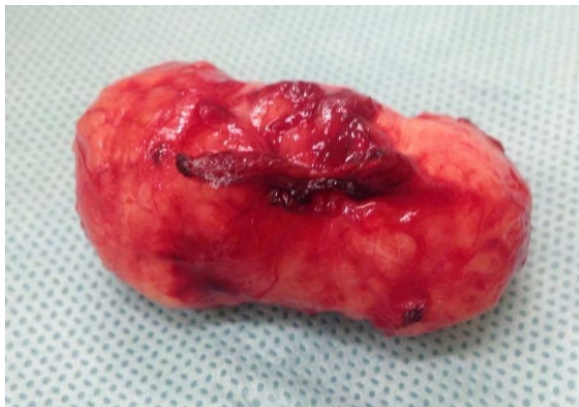


Figure 3. Surgical specimen resected with dimensions 4.5 cm by 2.5 cm by 2.0 cm

DISCUSSION

Esophageal leiomyoma is found, in more than 80% of cases, in the middle and lower thirds of the esophagus⁹, and in the case discussed, the leiomyoma was found in the proximal third of the esophagus, diverging from the majority of cases.

The patient was 73 years old in the occasion of the diagnosis, in discordance with the literature, since this esophageal tumor is more common between the ages of 20 and 59 and occurs more frequently in men, with relative frequency of 2:1 to 5:1⁶.

The esophageal leiomyoma is usually an incidental finding, since initial development is asymptomatic⁶. The patient mentioned presenting occasional dysphagia and/or odynophagia to solids and frequent choking, however the mediastinal mass diagnosis was done only after having done a thoracic x-ray which evidenced a mediastinal enlargement to be clarified, later investigated by thoracic TC.

It is a fact that the esophageal leiomyoma diagnosis is initially challenging, since the initial symptoms are nonspecific and may be confused with Gastroesophageic Reflux Illness (DRGE). The imaging resources are very useful to guide the diagnosis, however they do not possess good specificity for diagnosis, since other esophagic benign tumors can present similar images, like fibroma, lipoma, neurolemomma, hemangioma, lymphangioma, squamous papilloma, fibrovascular polyp and granular cell myoblastoma; as well as malignancies, such as carcinomas and gastrointestinal stromal tumors.

In the thorax radiography, it may present itself as mediastinal mass. In the endoscopy in general it causes discrete submucosal bulge as it occurred in the case of the

patient, or lumen reduction in the barium examinations⁷. The tomographical image may show intramural or excentric homogeneous lesion of soft tissue, which is rarely circumferential, while in the patients' case the image is circumferential as evidenced in Figure 1. The mucosa membrane is frequently normal and the magnetic nuclear resonance can reveal images in T2 with capture with no anormalities⁹.

The echoendoscopy (puncture) is used to define the limits of the tumor and shows five layers of the esophagus: 1) hyperechogenic, superficial mucosa; 2) hypoechogenic, mucosa and muscularis mucosa; 3) hyperechogenic, submucosa; 4) hypoechogenic, muscularis propria; 5) hyperechogenic, adventitious⁷.

The definitive diagnosis requires histopathological analysis of the tumor which is obtained through preoperative biopsy or of the resulting piece of the surgical act. Nevertheless, biopsies should be avoided as much as possible; they may be inconclusive by scarcity of material of the submucosa or generate inflammatory reaction which interferes in the operation. The difficulties in relation to the anatomopathological examination are exemplified in the present case, since the biopsy done in the first upper digestive endoscopy did not provide sufficient material to clarify the histological nature of the lesion⁵.

The treatment may be surgical or expectant. The excision of symptomatic leiomyomas, or of those greater than 5 cm, is generally recommended. On the contrary, in the asymptomatic or of those smaller than 5cm, an option given is of an expectant conduct, with accompaniment through periodical esophagographies, since these tumors have a very characteristic radiological appearance, generally a very low growth rate, and the risk of malignant degeneration is very low, but the surgical conduct was the option chosen by the patient.

However, because of the difficulty in differentiating it from the malignant form, many surgeons prefer the tumor resection, and posterior referral to the anatomical-pathological service⁹. This was the measure taken by our service, which opted for a torachotomy as opposed to a torachoscopy, because of the greater safety in the surgical approach open by the team in question. The surgical act was done by a thoracic surgeon and an oncological surgeon in which, according to the literature, the results are excellent, with no record of recurrences.

CONCLUSION

The case reported possesses epidemiological and topographical characteristics which differ from reported cases of esophageal leiomyoma.

Furthermore, through bibliographical review, particular attention is drawn to the rarity of the esophagus leiomyoma, which is considered a diagnostic challenge, being initially asymptomatic, and, in most cases, an

incidental finding in radiological or endoscopic tests. Therefore, the reported case promotes the recognition of a rare diagnosis, with low incidence, and stimulates the resolution of diagnostic challenges and the promotion of knowledge of the various ways of handling and resolving these cases.

Authors contributions: *Pereira FB* - author and editor of the article, bibliographic research, textual and image editing, organization of documentation and contact with Ethics Committee and Brazil Platform; *Lion BC* - Textual review; *Lelli LF* - Textual review and technical guidance; *Lima MP* - Textual review, technical guidance and documentation of ethics committee.

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Submitted for publication: November 15, 2017

Accepted in: February 26, 2018