Esophageal leiomyomas presenting as a mediastinal mass

Léiomyomes oesophagiens se présentant sous la forme d'une masse médiastinale

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RÉSUMÉ

Les tumeurs bénignes de l'œsophage sont très rares, les léiomyomes en représentent le type histologique le plus fréquent. La plupart de ces léiomyomes sont de petite taille et asymptomatiques, sans risque de dégénérescence maligne. Ils sont généralement découverts fortuitement lors d'une endoscopie digestive. Parfois, ils peuvent se manifester cliniquement par une dysphagie, une hématémèse ou d'autres signes cliniques. Ils peuvent mimer un cancer de l'œsophage, qui est plus fréquent, ou certaines tumeurs médiastinales. Le diagnostic peut être orienté par le transit oeso-gastro-duodénal ou par d'autres explorations radiologiques, mais seul un examen histopathologique permet de confirmer le diagnostic. Nous rapportons le cas d'un homme de 50 ans, non-fumeur, se plaignant d'une dysphagie et de douleurs épigastriques, le tout évoluant dans un contexte d'altération de l'état général. La présentation clinique et radiologique étaient celles d'une tumeur médiastinale. Une chirurgie diagnostique et thérapeutique a été pratiquée et l'examen histopathologique de la pièce de résection a conclu à deux léiomyomes de l'œsophage.

Mots-clés : dysphagie, léiomyomes, masse médiastinale

SUMMARY

Although benign tumors of the esophagus are very rare, the leiomyomas are frequently recorded. Most of them are small, asymptomatic and without risk of malignant degeneration. These benign tumors are usually discovered fortuitously on endoscopy. Sometimes, they may manifest clinically by dysphagia, hematemesis or other signs. They may mimic the esophageal cancer, which is more frequent, or some mediastinal tumors. The diagnosis can be oriented by the barium swallow esophagogram or other imaging methods, yet, only the histological examination gives the confirmation of the diagnosis. We report the case of a 50-year-old man, non-smoker, complaining of dysphagia, epigastric pain and deterioration of general condition. The clinical and radiological presentation mimicked a mediastinal tumor. Surgery was performed, and histological examination concluded to two leiomyomas of the esophagus.

Key words: dysphagia, leiomyomas, mediastinal mass

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INTRODUCTION

Leiomyomas are the most common benign tumors of the esophagus. They represent 10% of all gastrointestinal leiomyomas. They are multiple in 3 to 10% of patients. Leiomyomas usually occur in young adult (patients are aged between 20 and 50 years) [1]. They appear in 80% of cases in the middle to lower third of the esophagus [2]. They become symptomatic if their diameter exceeds 2 centimeters or if they are in proximity of the pharyngo-esophageal junction [3]. On radiological exploration, a leiomyoma may appear as a mediastinal mass [4]. Moreover, computed tomography (CT) is not powerful enough to differentiate benign lesion of the esophagus from the malignant one and is not able to individualize the content of the tumor mass [5]. So, despite the investigation's technological improvements, definitive diagnosis may only be established histologically after surgery [6]. We report a case of esophageal leiomyomas presenting as a visceral compartment mass, according to the new definition of mediastinal compartments [7]. After different investigations, surgery was performed. We therefore emphasize the place of the definitive anatomo-pathological study in the diagnosis of leiomyomas if esophageal endoscopic ultrasonography is not available.

CASE REPORT

A 50-year-old man, diabetic, non-smoker, complained of epigastric pain, dysphagia and impaired general condition with weight loss. He had no other symptoms like fever, chest pain or hematemesis. Clinical examination and blood tests were normal. The chest radiography showed an enlargement of the middle mediastinum (Figure 1). An upper gastrointestinal endoscopy was performed to explore his dysphagia and revealed an extrinsic compression of the esophagus starting from 26 cm of the anterior incisor teeth extending to 32 cm of the anterior incisor teeth with normal mucosa. The chest CT showed a tissular mass of the visceral compartment of the mediastinum measuring 80x32x16 mm. This mass was well limited, homogeneous, oblong, slightly raised after injection of iodinated contrast agents (Figure 2). This lesion reduced the esophageal lumen and was in contact with the descending thoracic aorta, the posterior wall of the left main bronchus and the carina without signs of infiltration (Figure 3). There was no associated lymphadenopathy and no pulmonary nodule. The flexible bronchoscopy was normal. A first diagnostic thoracoscopy was decided. It showed several centimeter lymph nodes in sub carina region. The frozen section concluded an inflammatory tissue without malignancy signs. Conversion to postero-lateral thoracotomy was decided. It showed a mass of the esophageal muscle that was enucleated progressively. The frozen section and the final pathologic examination of the mass concluded to two esophageal leiomyomas measuring respectively 5.6x3x2 cm and 1.3x1 cm. The esophagography on the fourth postoperative day showed a persistent image on the postero-lateral left wall with no extravasation of barium or stenosis (Figure 4). The postoperative recoveries were simple, and the hospitalization did not exceed one week. The upper gastrointestinal endoscopy's checkup showed normal esophagus. Four years after surgery, no recurrence or other complications had been detected.

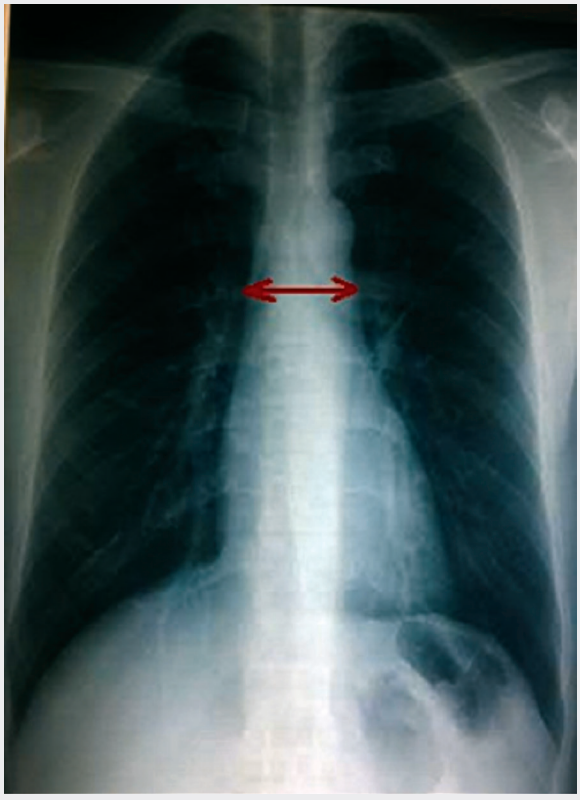

Figure 1. Enlargement of the middle mediastinum on the chest radiography (red arrow) with no associated pulmonary parenchymal abnormalities.

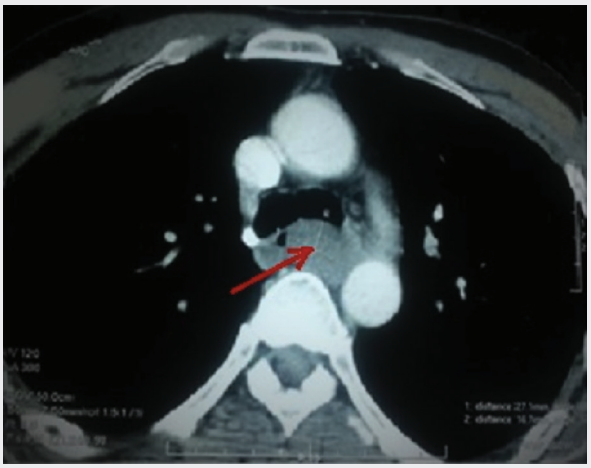

Figure 2. A cross section of contrast enhanced CT scan showing a tissue lesion of visceral compartment (red arrow), measuring 80X32X16 mm, well limited without signs of infiltration or associated lymphadenopathy.

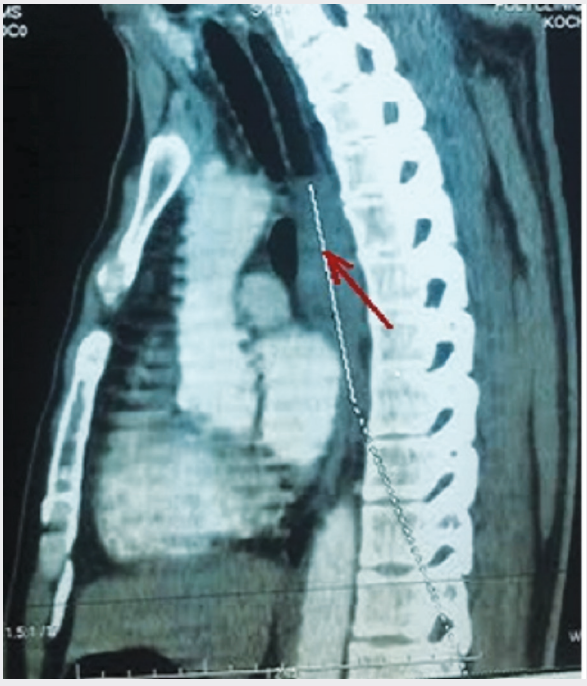

Figure 3. A longitudinal section of contrast enhanced CT scan showing a tissue lesion of visceral mediastinum (red arrow), in contact with the descending thoracic aorta, the posterior wall of the left main bronchus and the carina without signs of infiltration.

Figure 4. Postoperative barium swallow esophagogram showing a persistent image on the postero-lateral left wall of the esophagus (red arrow).

DISCUSSION

The case we are reporting emphasize the difficulties in the exploration of mediastinal masses especially in the visceral compartment. It also demonstrated that the chest CT scan is not performant in the description and the analysis of esophageal lesion and may misdiagnose the real number of lesions. If the esophageal endoscopic ultrasonography is not available, the final diagnosis will only be done after histological examination.

Leiomyomas are most commonly localized in the esophagus and constitute 10% of all gastrointestinal leiomyomas. Benign esophageal tumors rarely cause dysphagia and the lack of symptoms is due to their slow growth combined with the distensible character of the esophagus [3]. However, some cases of leiomyoma with extra-esophageal lesions which cause compression on mediastinal structures, such as bronchi and superior vena cava, have been reported [9]. In a review of 838 cases,

leiomyomas were intramural in 97% of cases, intraluminal in 1% of cases and extra-esophageal in 2% of cases [2]. In another study, leiomyomas were developed in mediastinum in 5% of cases [10]. In our case, there was two leiomyomas responsible of dysphagia which can be explicated by their size (80x32x16 mm).

Chest radiography is not sensitive and specific enough for the diagnosis of leiomyomas [11]. Indeed, the mass must reach a significant diameter before it becomes visible on chest radiography [12]. The chest CT has an estimated sensitivity of 91% for the evaluation of these leiomyomas. It helps to evaluate the invasion of nearby structures and the presence of extrinsic compression [13]. Thus, it allows excluding some other mediastinal masses that could lead to similar clinical presentations. Nevertheless, it is limited in the evaluation of intramural masses and especially in the description of the content of these masses [14]. For better study of mediastinal masses in contact with the esophagus, esophageal endoscopic ultrasonography should be performed when available [15]. It can help in the diagnosis and treatment strategy choice of esophageal leiomyoma [16]. However, in its unavailability, surgery will be needed for histological diagnosis and to rule out malignancy.

Nonetheless, when leiomyomas are diagnosed on esophageal endoscopic ultrasonography, they will be candidates for surgery if they are symptomatic or larger than 5 cm [17]. However, for small and asymptomatic tumors, regular (annual) endoscopic or / and radiological monitoring is essential because the diagnosis of leiomyosarcomas or stromal tumors remains not excluded. Small tumors with a pedunculated or a well-defined sessile insertion may benefit from endoscopic resection [18]. The surgery consists of the enucleation of the tumor traditionally by thoracotomy but since 1992 thoracoscopy has been much more used [19]. Multiple surgical techniques are described in the literature: open resection, endoscopic enucleation or minimally invasive surgical instrumentation using either thoracoscopy or laparoscopy [20-25]. However, there is no standardized surgical management and the choice of the technique depends on the experience of the surgeon and the location and the number of the lesions. In USA, surgical enucleation using the robotic daVinci Surgical System was used in 2001 and had excellent outcomes [17]. Finally, definitive pathological examination is the only method which confirms the diagnosis of benignity of these lesions and after surgery, it has good outcomes without risk of relapse.

CONCLUSION

Esophageal leiomyomas are rare benign tumors and asymptomatic in the majority of cases. However, sometimes, they could be responsible of symptoms evoking a mediastinal mass and the diagnosis can be difficult, especially with the low sensitivity of computed tomography in the exploration of the parietal tumors. Therefore, the esophageal endoscopic ultrasonography should be indicated whenever an esophageal leiomyoma is suspected to improve radiological diagnosis and to perform a non-invasive surgical treatment.

There is no conflict of interest for this case

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