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Esophageal Liposarcoma

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Abstract :
The Liposarcoma Of The Esophagus Or The Oropharyngeal Sphere Remains Very Rare,
We Present Here A Primary Esophageal Liposarcoma Discovered In A 70-Year-Old Male Who Consulted For A Cervical Dysphagia And Whose Complementary Examinations Were In Favor Of A GIST. A Laparoscopic Lateral Esophagotomy, Of More Than 10 Cm Was Performed To Extract The Intraesophageal Mass. The Histological Analysis Shows A Well-Differentiated Liposarcoma.
In This Case Report, The Diagnostic And Therapeutic Challenges Are Discussed.
Keywords: *Dysphagia, Esophagus; Liposarcoma, Esophagotomy*

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I. Introduction:

A primary esophageal liposarcoma is very rare. In the literature we have since 1983, only 42 cases for all esophagus and oropharyngeal sphere liposarcoma.

The atypical clinical presentation, and the complementary examinations, led to an initial diagnosis of mesenchymatous tumor, in our case to a gastrointestinal stromal tumor.
the diagnosis remained elusive until surgery and histological analysis had been completed.

II. Case report

We present the case of Mr. L aged 70, who had consulted for progressive lower cervical dysphagia, developed over 4 years, initially only for solid then with intermittent dysphagia for liquids. This dysphagia was associated with an intermittent dyspnea and weight loss. There was any notion of odynophagia, regurgitation, heartburn, or other functional sign.

His history included a resection of a Zenker diverticulum 30 years earlier. Dysphagia developed gradually over 4 years and chronic smoking quitted.

An esophago-gastro-duodenal fibroscopy had revealed a bulging of the mucosa partially obstructing the esophageal lumen with a regular, smooth surface, repressible, non-pulsatile formation.

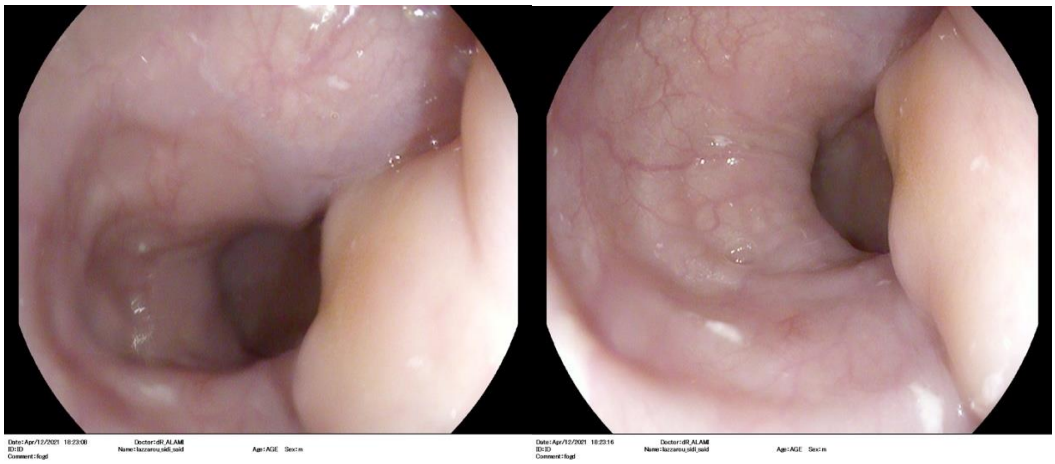

Figure 1 : endoscopic view of the oesophageal mass

Subsequently, an esophago-gastro-duodenal transit showed: The presence of diffuse dilation of the esophagus without significant parietal abnormality and flattening and dilation of the thoracic esophagus with a permeable cardia.

On the cervico-thoracic CT, we found a parietal process straddling the cervical and thoracic portion of the esophagus, fusiform starting 2-3cm from Killian's mouth and ending just below the carina at a height of nearly 15cm. This formation was hypodense with a small fatty contingent without enhancement after injection of contrast product. It does not exert any compression on the surrounding structures, on the trachea and carina. There was a regular appearance of the outer edges. The rest of the CT examination was unremarkable.

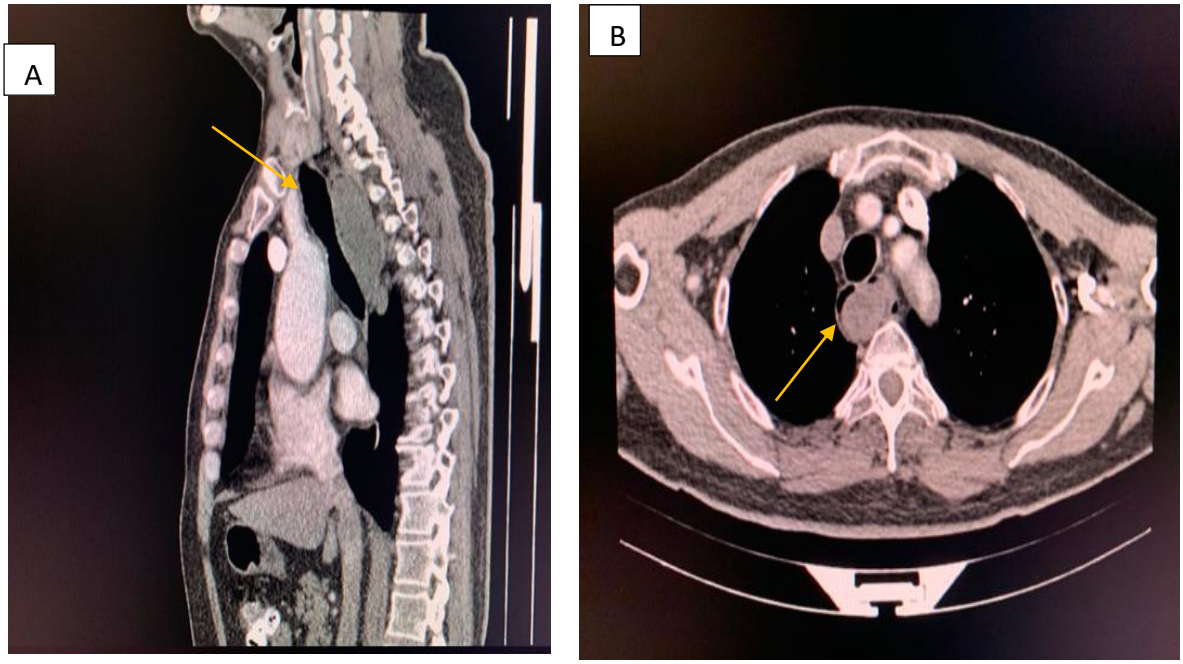

Figure 2: CT scan sagittal section (A) and axial section (B) showing the mass

The first diagnosis we think about it is a stromal tumor. The planned operation was a laparoscopic enucleation. However, the patient was informed about the possibility of an esophageal resection.

The thoracic approach was made by video surgery, the patient in prone position: with 3 trocars in the 5th, 7th and 9th right intercostal spaces.

The procedure required a dissection of the esophagus up to the top of the thorax. The myotomy had not found any tumor under the mucous membrane and it seemed rather intraluminal. A lateral esophagotomy of more than 10 cm was performed to extract the intraesophageal mass: corresponding to a long tumor covered with mucosa measuring 15 cm. The tumor was completely resected after application of an endoGIA© 60 forceps on the implantation base. The esophagotomy was closed in 2 layers.

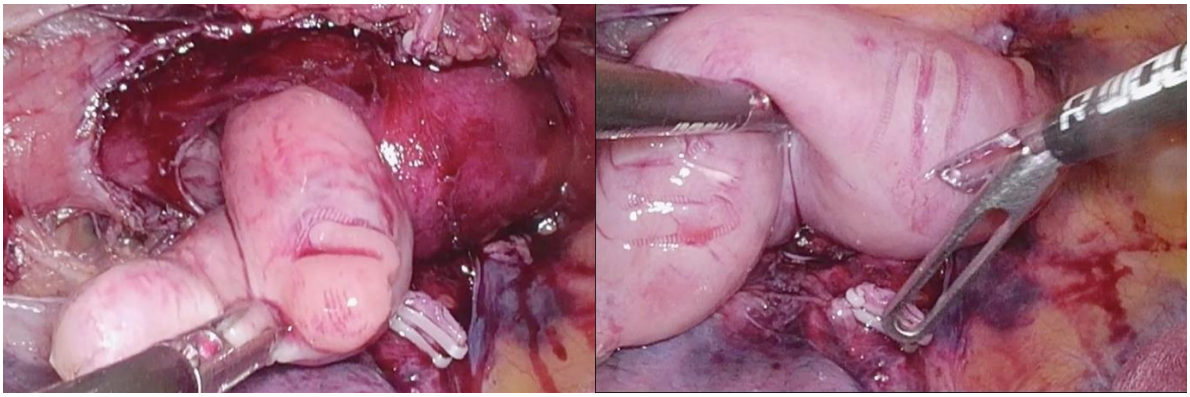

Figure 3: peroperative view of the mass

The patient had to be hospitalized in intensive care for 48 hours due to postoperative dyspnea. The evolution was then favorable without complications.

The macroscopic aspect of the mass revealed: an oblong formation measuring 12*6*2.5 cm. When cut, it was greasy, multinodular, measuring 0.5 and 3.5 cm and encapsulated.

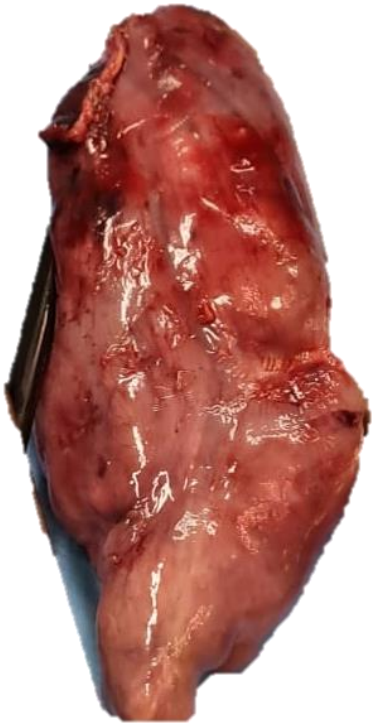

Figure 4: macroscopic view of the mass

Histological examination revealed mesenchymal proliferation with a majority lipomatous component suggesting a well-differentiated liposarcoma - lipoma like.

The immunohistochemical study showed: the absence of anti-MDM2, anti-CD117, anti-AML and anti-CD34 antibodies.

The search for MDM2 amplification was positive

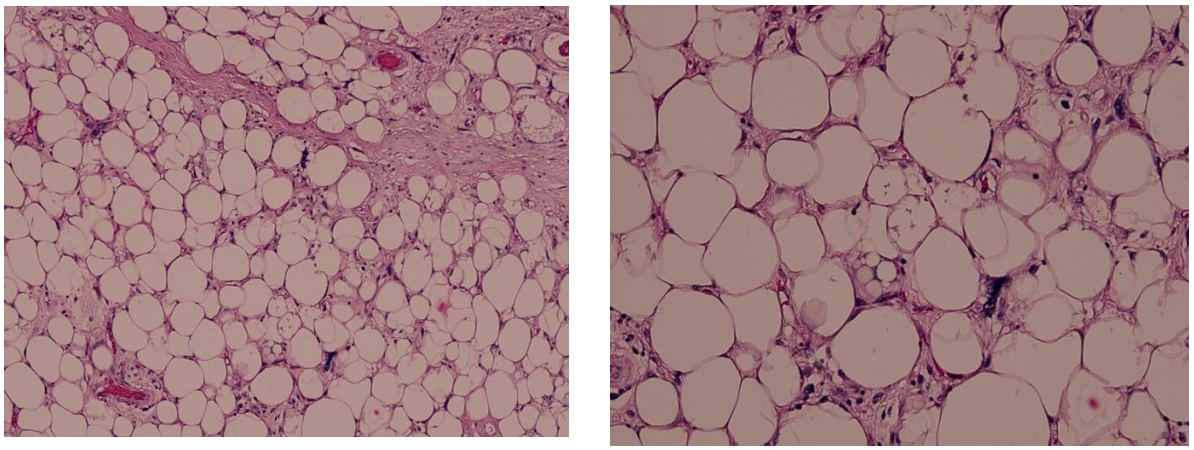

Figure 5: histological examination showing lipomatous proliferation

In conclusion, the diagnosis of well-differentiated liposarcoma was retained.

III. Discussion :

Liposarcoma is the most common sarcoma in adults, however it constitutes only 0.1 to 1.5% of all malignant esophageal masses. Its preferential location is in the retroperitoneum and the deep soft tissues of the lower limbs and the trunk. In the digestive tract it is usually found in both the colon and the distal ileum. Moreover, its location in the esophagus or the oropharyngeal sphere remains very rare. Only 42 cases have been reported in the literature [9,2].

Eighty percent of esophageal liposarcomas were in the cervical part of the esophagus. Most of the lesions cited were polypoid in nature (78% of cases) [9].

In the esophagus, the well-differentiated liposarcoma presents similar symptomatology, macroscopic and microscopic characteristics, and clinical course to those described in the giant fibrovascular polyp of the esophagus which was considered a benign lesion.

Well-differentiated liposarcoma is the form most frequently found in the literature, which is also found in our patient, whose main differential diagnosis was the giant fibrovascular polyp of the esophagus, the existence of which has recently been called into question: a series of 13 cases of giant fibrovascular polyps of the esophagus were reviewed.

This retrospective study reclassified six of these lesions as liposarcomas: 3 well-differentiated liposarcomas and 3 dedifferentiated liposarcomas, given the demonstration of amplification of the MDM2 gene by FISH. (4)

Given its mesenchymal nature, it can also be confused with other tumors originating from the mucosa or submucosal layer such as GIST or lipomas (11).

In our patient, we find an operated Zenker's diverticulum as an antecedent, however in the other cases described we do not find any particular antecedents. Also the average age of discovery is around 60 years with a male predominance.

Our patient complained of progressively worsening dysphagia: we indeed find the notion of dysphagia in the vast majority of reported cases. The other most frequently found symptoms were nausea, cervical constriction, regurgitation, odynophagia, and dyspnoea.

Some patients may present with unusual symptoms: chest discomfort, cough, shortness of breath, which makes the diagnosis more difficult [1].

Furthermore, the endoscopic examination was in favor of a submucosal lesion, and the cervicothoracic CT scan directed us towards the diagnosis of stromal tumor (GIST).

The diagnosis of liposarcoma could only be made after surgical resection and histological analysis with amplification of the MDM2 gene. Moreover, in the majority of reported cases, the diagnosis could only be made after surgical excision. In a few rare cases the diagnosis could be made on biopsies by echoendoscopy.

The standard treatment frequently performed is surgical resection: total or partial esophagectomy. Total or partial esophagectomy was performed in 85% of cases. Endoscopic treatment including simple polypectomy is performed in 8.6% while endoscopic submucosal dissection in 5.7% of cases.(9)

In our case we opted for a minimally invasive approach, surgical resection by video surgery with thoracic approach in prone position, which allowed us a conservative treatment with resection of the entire tumor. No adjuvant treatment is recommended after discussion in the multidisciplinary consultation meeting.

Despite the limited data collected for this tumour, the main recommendations are surgical resection with close monitoring.

In our patient, the follow-up was done by a clinical examination every 3 months and a thoracic scannographic examination every 6 months for 2 years which showed no abnormality, no recurrence. This follow up will continue every 6 month for a year, then every year for up to 5 years of evolution.

We do not find much information on the evolution of these liposarcoma resected or not in the literature.

In Graham's study, we find two patients who died of their esophageal disease during the course, one of whom had liver metastases.

IV. Conclusion :

The case we have reported illustrates the diagnostic and pathological difficulty faced with this type of tumor, which remains rare and whose evolution is not sufficiently specified by the authors.

Currently the most widely used protocol is based on surgical treatment followed by monitoring. No adjuvant treatment is offered.

In general, any large, benign or malignant tumor may require a complete resection of the esophagus.

However, minimally invasive surgery techniques can still be used: organ preservation is to be preferred as long as possible.

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