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Case Report

Enucleation for a Giant Esophageal Leiomyoma in a Pediatric Patient with Preservation of the Native Esophagus. Case Report and Review of the Literature

Reem Alawna[1], Alaaeddin Hliqawi[1], Lana Hamad[1], Izzeddin A Bakri[2], Ahmad H Aliwisat[3], Ahmad Shaltaf[3], Hazem Ashhab[4] and Firas Abu Akar[5]*

[1]*Medical Research Club, Faculty of Medicine, Al-Quds University, Jerusalem, Palestine*

[2]*Department of Pathology, Makassed Charitable Society Hospital, Jerusalem, Palestine*

[3]*Department of Pediatric Surgery, Makassed Charitable Society Hospital, East Jerusalem, Palestine*

[4]*Gastroenterology Department, Al-Ahli Hospital, Hebron, Palestine*

[5]*Cardiothoracic Surgery Department, Makassed Charitable Society Hospital, East Jerusalem, Palestine*

***Corresponding Author:** Firas Abu Akar, Cardiothoracic Surgery Department, Makassed Charitable Society Hospital, East Jerusalem, Palestine.

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Abstract

Background: Esophageal leiomyoma is a sporadic tumor in pediatrics; when it's a giant leiomyoma and involves more than 2/3rd of the esophagus, esophagectomy may be the only option for the patient. In this Case report, a giant leiomyoma in a child that was enucleated with preserving the patient's esophagus is reported.

Case Presentation: A 13 years old male patient presented with dysphagia since two years. Based on the history, physical examination, imaging and pathology the patient was diagnosed with giant esophageal leiomyoma [10*10*8 cm]. The patient underwent a right anterolateral thoracotomy for tumor enucleation. The postoperative course was complicated with a leak which was handled with endoluminal stenting. The patient is now free of symptoms with a well-healed wound.

Conclusion: Usually esophagectomy is performed in this size of leiomyoma. In our case, although the tumor occupied more than 2/3rd of the esophagus, the native esophagus was preserved successfully. Surgeons should always try their best to avoid esophagectomy in this kind of tumor, especially in this pediatric age group.

Keywords: *Case Report; Huge Leiomyoma; Surgical Enucleation; Preservation of the Native Esophagus; Pediatric Leiomyoma*

Introduction

Esophageal leiomyoma is a sporadic tumor in pediatrics; when it's a giant leiomyoma and involves more than 2/3rd of the esophagus, esophagectomy may be the only option for the patient.

Case Presentation

A 13-Year-old male patient with a history of intermittent dysphagia since two years. Gastro-colonoscopy showed: hiatus hernia with severe reflux esophagitis, EUS showed: submucosal mass along the distal ⅔ of the esophagus, FNA/FNB was obtained, and pathology was consistent with Leiomyoma. The patient was admitted to our clinic for surgical management. There were no findings on the general examination. Chest and abdominal CT scan with contrast showed a 10.5 x 5 x 17.5 cm mass lesion in the posterior mediastinum; air bubbles were noted within the central part of the dilatation of the upper esophagus, the lesion extended downward to the fundus of the stomach

(Figure 1a and 1b). The lesion represents an esophageal tumor with involvement of the gastro-esophageal junction, no other masses or lymphadenopathy.

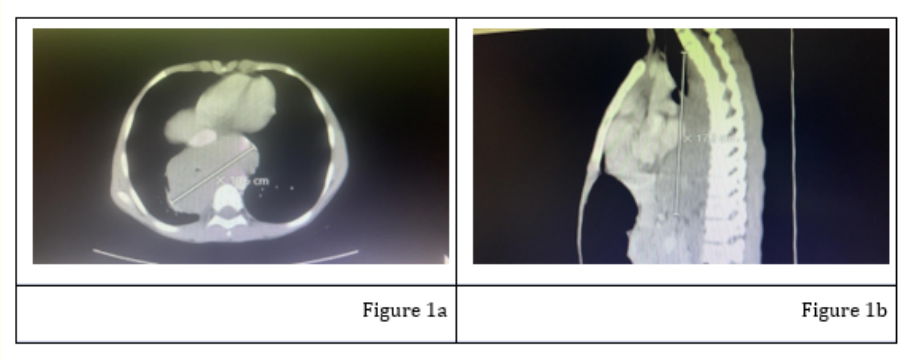

Figure 1: *(a) Axial view CT scan showing a posterior mediastinal mass arising from the esophagus with maximal diameter 10.6 cm. (b) Sagittal view of the mass showing its maximal length 17.2 cm.*

The preoperative endoscopic evaluation revealed esophageal dilatation with distal esophageal compression. The patient underwent a right anterolateral thoracotomy for esophageal mass enucleation. Despite its difficulty, the surgery was smooth, and the enucleation of the mass from the esophagus and the stomach was performed. This resulted in severe damage and many lacerations in the mucosal layer, which were Stitched using 4-0 vicryl absorbable sutures.

By the end of the resection, buttressing the area of the resection was done with parietal pleural flap (Figure 2). Two chest drains [28 Fr] were remained in the pleural cavity by the end of the procedure.

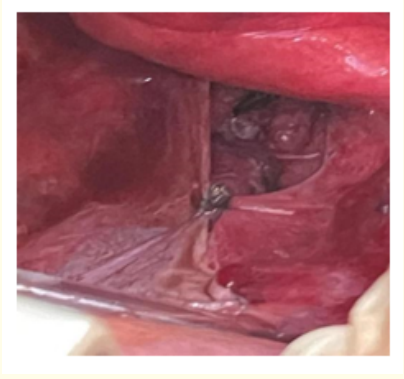

Figure 2: *Parietal pleural flap buttressing the area of the resection.*

During the early postoperative days, the patient maintained on total parenteral nutrition and empiric broad spectrum antibiotics and anti-fungal drugs. Chest drains showed high output drainage with high levels of amylase. Twelve days after the surgery, pleural fluid cul-

ture exposed a growth of gram-positive organisms treated with proper antibiotics. Gastrografin swallow study performed on day fifteen after the operation and showed minimal leak, and pleural ultrasound showed minimal pleural effusion. With good drainage, suitable antibiotics, and reasonable maintenance of the nutritional status, the patient's general situation remained stable and under control. On day 27 after the operation, the patient underwent endoscopic stenting to bypass the leak (Figure 3). 2 months post op the patient underwent stent removal-which was found in the stomach without any complication.

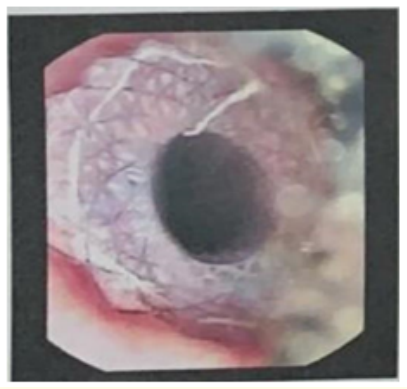

Figure 3: *Fully covered esophageal stent under direct vision.*

The final pathology report showed: Grossly specimen composed of multiple fragments of Pinkish gray white tissue with whorled cut surface measuring in aggregate 10 x 8 cm. Microscopic examination (H&E) showed intersecting fascicles of monotonous spindle cells with indistinct borders, eosinophilic cytoplasm, cigar shaped nuclei and small nucleoli. No atypia or necrosis seen. No mitoses could be identified (Figure 4). Immunohistochemistry study showed demonstrable positivity for Actin and H.caldesmon immunostains while negative for S100 and CD117(c-kit) markers (Figure 5).

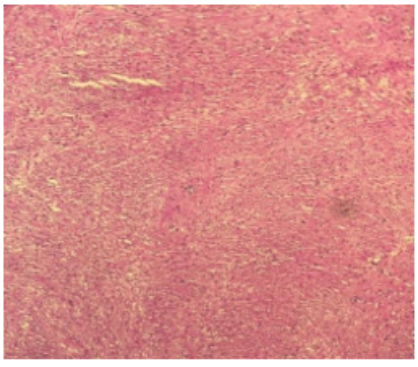

Figure 4: *H&E examination*

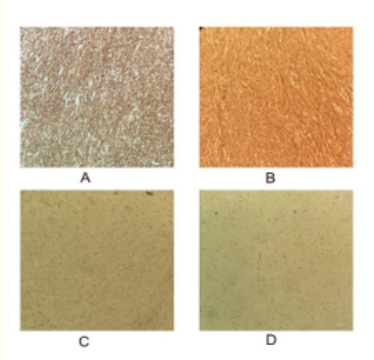

Figure 5: *Immunostains; a: 20X H.Caldesmon Immunostain positive. b: 20X Actin immunostain positive. c: 20X:S100 immunostain negative. d: 20X:CD 117 [C-kit] immunostain negative.*

Discussion

Pediatric esophageal leiomyoma is one of the rarest diagnosis, with the first case diagnosed in 1916 [1]. Now only around 40 cases were reported. Of all esophageal leiomyoma cases, pediatric leiomyoma represents 2.6%, with a mean age of 14 years, an age range of 4 to 20 years [2], yet it was reported as young as two years [3] and one year old [4,5]. It is 1.71 times more common in females [2], While the usual route for treating giant esophageal leiomyoma [GEL] is the open thoracotomy with tumor resection by total esophagectomy and gastric or intestinal pull-up [6-10].

New efforts are guided toward a less invasive and traumatizing approach. Here we describe successful treatment of a giant leiomyoma case measuring [10*10*8] cm with tumor enucleation and preservation of the native esophagus in a 13-year-old male patient.

In pediatric esophageal leiomyoma, the most common presenting symptom is dysphagia; other symptoms include retrosternal pain, vomiting, cough, dyspnea, weight loss, hematemesis, hiccups, choking, gastro-esophageal reflux, and bronchial asthma [6,11]. Bleeding may also occur when the mucosa over the tumor becomes ulcerated [11].

The frequency of the mass decreases when going proximal in the esophagus. In pediatrics, only one case reported had lesions in the upper esophagus, and it was associated with lesions in the middle part [12]. In general, it has been found that 53% of cases occur in the Lower and 43% middle thirds of the esophagus [11]. Inheritance was reported in 3 families in the same part of the esophagus [6,13-15]. The highest associated illness was with Alport's syndrome [2,16] other coexisting diseases included hiatal hernia, mucosal ulcerations, peptic ulcer, rectal prolapse, cholelithiasis, and genital anomalies such as hypertrophy or leiomyoma of the vulva and clitoris [2].

The coexistence of both esophageal and bronchial leiomyoma was described in two cases [3,4]. In both, the disease was at a very young age of 1,2 years [3,4].

Leiomyoma was found at the site of atresia repair in a 1-year-old girl with a history of long-gap esophageal atresia treated with recurrent bougienage dilation, including steroid injection [4].

The mean tumor diameter is 3.7 cm [range, 1 cm to 29 cm]. According to the literature, 22.5 × 10 × 7.5 cm is the most giant reported leiomyoma [6].

Tumor characteristics

In pediatrics, it presents in three types, localized leiomyoma 9%, multiple localized leiomyomata in less than 1%, and Diffuse leiomyomatosis 91% [2]. The tumor may be solitary in 91%, multiple in 9%, or calcified in 6% of cases [6,17]. Calcification of esophageal tumors, though rare, may be diagnostic of leiomyomas [18].

Diagnosis

Diagnosis of leiomyoma may be incidental and usually made on an esophagogram, chest X-ray, [CT], and [MR] scan. Recently, esophageal ultrasound [EUS] has been widely used for diagnosis.

The obstacle with the conventional endoscopy is that it cannot distinguish between esophageal leiomyoma and other submucosal lesions or external pressure neoplasms [11]. EUS-FNA has proven to be a safe and effective technique to diagnose these lesions, mainly when the immunohistochemical analysis is performed [19]. It reveals the five-layered structure of the esophageal wall and thereby confirms the nature, size, number, and origin of the lesions [20].

However, endoscopic biopsies should be discouraged unless there is some diagnostic uncertainty that would make resection unwarranted. Experts suggest that a preoperative biopsy may make the enucleation of the tumor harder due to the formation of post-biopsy mucosal adhesions [20,21].

On histological exam, leiomyoma stains positive for desmin and actin, in contrast to gastrointestinal stromal tumor GIST, which stains with CD34 negative and CD 117 [c-kit] negative [18].

Malignant transformation

In a comprehensive review of more than 800 cases reported in the world literature, only [0.2%] documented malignant transformation from leiomyoma to leiomyosarcoma [19].

Indication for surgery

Studies that did not focus on the pediatric age group may counter; Hennessy., *et al*. [22], stated that leiomyoma should be removed when diagnosed, even if asymptomatic as malignancy cannot otherwise be excluded are likely to develop if treatment is delayed. On the other hand, according to Pearson., *et al*. [23], although malignancy can only be ruled out by excision and histologic examination, it seems unreasonable to remove all small asymptomatic leiomyomas. However, in our literature review, we did not detect a reported case of pediatric esophageal leiomyoma that underwent a malignant transformation. Nonetheless, Surgical excision is the mainstay of treatment and recommended for symptomatic leiomyomas and those greater than 5 cm [11].

Treatment

The main treatment is surgical resection of the tumor via the right thoracotomy approach, being the most commonly performed [66%]. The traditional surgical approach for esophageal leiomyoma is usually mentioned as "small surgery, large incision" because it involves open thoracotomy [7,11]. Other approaches are Left thoracotomy, Thoracoabdominal, Midline laparotomy, VATS [Video-assisted thoracic surgery], and transverse cervical techniques, which are less commonly performed [11].

A report by Ozdil and his colleagues [24] described the treatment of esophageal leiomyoma with endoscopic percutaneous injection of ethanol. However, most of the esophageal leiomyomas treated during this manner were smaller than 10 cm.

Resection for giant esophageal leiomyomas [GEL] is typically accomplished with esophagectomy and gastric pull-up through left posterolateral thoracotomy. A study made by Bang-Chang Cheng., *et al*. on eight cases of giant esophageal leiomyoma [GEL]. Only one case was treated with transthoracic extra mucosal enucleation and buttressing the muscular defect with an omental flap. Three cases were treated with esophagectomy and esophagogastrostomy. Four cases were managed with total esophagectomy and esophageal replacement with the colon [6] subtotal esophagectomy with gastric advancement and cervical esophagogastric anastomosis for an 8 × 8 × 12 cm mass occupying the upper and middle thoracic esophagus was performed through the right thoracotomy approach [8].

If the tumor is densely adhesive to the mucosa, or if extensive mucosal damage occurs accidentally during enucleation, esophagectomy is preferred [25].

Giant leiomyoma of the esophagus is traditionally treated by open thoracotomy, which may cause considerable operative trauma and negatively affects respiratory function and resumption of a traditional diet and inevitable anastomotic complications, all of which can worsen the prognosis [7,26,27].

Because of the large tumor size and unclear boundaries with surrounding tissues, all the reported giant leiomyomas of the esophagus were managed by open thoracotomy or thoracoabdominal incision with gastroesophagectomy [7]. However, in recent years, some centers have gradually used minimally invasive surgeries to treat esophageal leiomyoma via thoracoscopy, laparoscopy, or Da Vinci robot-assisted thoracoscopy [28,27].

Other studies state that: If the lesion originates from the mucosal or submucosal layer, the preference is to do endoscopic therapy, such as endoscopic mucosal resection [EMR], and if it is originating from the muscularis propria, other endoscopic therapies, such as Endoscopic submucosal dissection ESD or the submucosal tunnelling endoscopic resection STER can be used [20].

The treatment of small esophageal leiomyoma [< 2 cm] originating from the muscularis mucosae with endoscopic aspiration lumpectomy has been proven to be safe and effective [29].

In addition to the well-known complications of any surgery such as infection and leakage, laceration of the esophageal mucosa is the most common intraoperative and postoperative complication associated with enucleation of esophageal leiomyoma [7]. The lacerated mucosa sites should immediately be repaired by suturing with absorbable sutures followed by interrupted suturing of the muscularis.

Gastric pull-up has some postoperative morbidity, including reflux esophagitis, stricture formation, dumping, diarrhea, reduced meal capacity, and weight loss [30]. The majority of postoperative complications may be related to the division of the vagal nerves and the subsequent loss of parasympathetic innervation to the foregut [31]. The rationale for using the colon is that there will be fewer complications related to acid reflux than when the stomach is used for esophageal replacement. Additional concern after enucleating a large leiomyoma is that the myotomy may functionally results in achalasia [32].

Prognosis and follow-up

The growth of esophageal leiomyoma is slow, and lesions usually remain stable during the follow-up period of 3 - 5 years. Even larger lesions had minor local and systemic clinical manifestations. Fortunately, no recurrence was observed after endoscopic or surgical resection during the follow-up [7].

Conclusion

As a general rule, esophagectomy is used to be performed in this size of leiomyoma with colon replacement, a procedure that carries many complications and long-life side effects. In our case, although the tumor occupied more than 2/3rd of the esophagus, the native

esophagus was preserved successfully. Surgeons should always try their best to avoid esophagectomy in this kind of tumor, especially in this pediatric age group. The patient has improved significantly and started to gain weight, further investigations and workup will be started now for his rectal thickness.

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