Endoscopy Assisted Resection of Huge Esophageal Hamartoma- Mimicking Second Tongue

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Abstract

Hamartoma of the esophagus is a rare lesion and number of cases reported in literature to date is limited. The majority of hamartomas are intraluminal tumors located in the upper third of the esophagus. We herein present a case of a 60 years old man with an intraluminal hamartoma located in the upper third of esophagus. He presented with dysphagia and a mass coming out as if he had a second tongue. He could replace it back. Endoscopic assisted resection of the mass was done after routine investigations. Biopsy revealed hamartoma and he was discharged without any uneventful episode.

KEYWORDS- polyps, Hamartomas, Pedunculated, endoscopy, polypectomy

INTRODUCTION

Large pedunculated polyps of the esophagus and hypopharynx are infrequently encountered in clinical practice. Benign tumors of the esophagus are rare, accounting for <1% of all esophageal neoplasms. Leiomyomas are the most common benign esophageal tumors, whereas hamartomas are the rarest benign tumors (1).

Harrington credited Sussius with first autopsy report of esophageal polyp in 1559(2). In 1763, Monroe reported one of the earliest treated cases which were accomplished by simple Tran's oral ligation (2).Vinson in 1992 was the first to describe, in the American literature, the successful surgical removal of a large esophageal polyp which was approached through left cervical incision (3).

CASE REPORT

60 years old male patient presented to surgery department at our institution with progressive dysphagia and gradual restriction of oral intake since 2 years. During all this time he has been having episodes of regurgitation of fleshy mass between his teeth, associated with respiratory distress, relieved when the patient manually reduced the mass into his esophagus back (Fig1). His physical examination was unremarkable. In general, patient was well preserved with no history of weight loss.

Patient underwent CECT of neck and thorax suggestive of large mixed (soft tissue and fat) density, pedunculated mass lesion arising from the anterior wall of esophagus and projecting into the lumen causing its dilatation and luminal narrowing. The lesion is in upper part of esophagus ending from C6-C7 to D7-D8 level and measures 11cm x25cm in axial plane. The caudal pole of the lesion is approximately 70 mm from esophageal hiatus, most likely to be leiomyoma.

The patient was operated under general anesthesia with endotracheal intubation. Incision was made in the neck with a semilunar incision and on opening the laryngopharynx, endotracheal tube was visualized and the ryles tube was withdrawn and endoscope was inserted. With the help of an endoscope the esophageal polyp was brought out into the wound it was fleshy big pedunculated mass had smooth and shiny surface with no ulceration (Fig.2). The polyp or mass was arising from the anterior wall of esophagus from the lower part of upper third of esophagus having an intraluminal extension almost reaching the lower end of esophagus, it was ligated at the base and excised completely (Fig.3). Post-operative period was uneventful and patient was fully relieved from his symptoms.

Microscopically the tumor was lined by hyperplastic squamous lining with several blood vessels of variable sizes filled with RBC, surrounded by fibro fatty collection of inflammatory cells at several places.

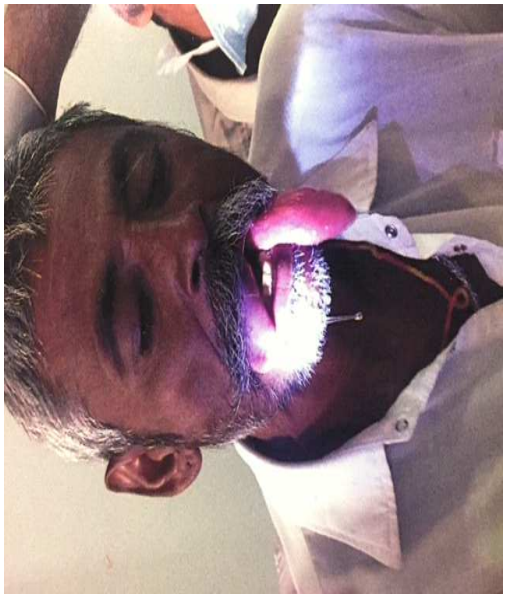

Figure 1

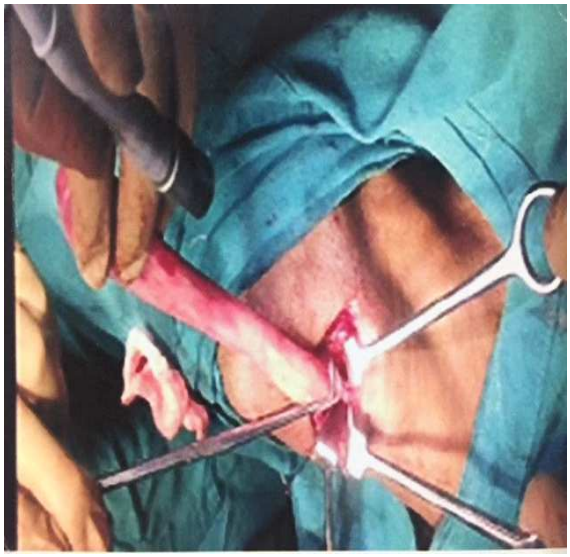

Figure 2

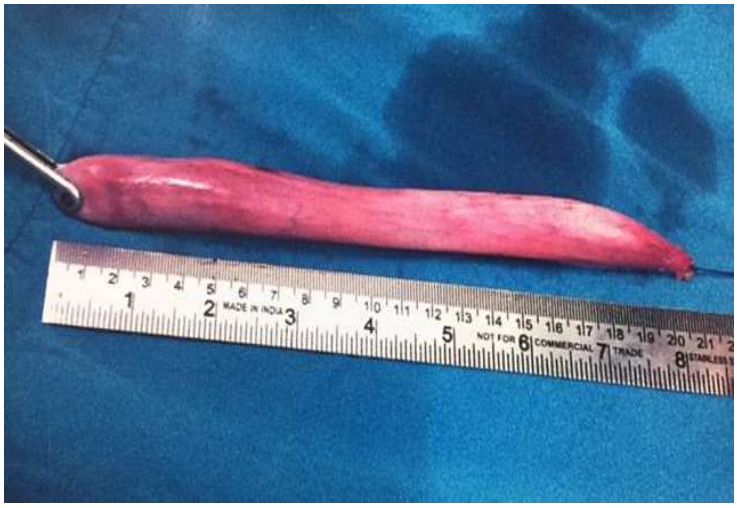

Figure 3

DISCUSSION

Hamartomas is defined as a non – neoplastic, uni- or multifocal developmental malformation, comprising a mixture of cytologically normal mature cells and tissues which are indigenous to the anatomic location, exhibiting a disorganized architectural pattern with predominance of one of its components(4). Hamartomas may occur in any organ, but are most commonly located in the lungs, liver, pancreas and spleen; however they are rare in the esophagus.

Benign tumor of the esophagus is distinctly uncommon. Schaffer et al(5) recorded 11 cases out of total 6001 autopsies; while Moersch et al(6) encountered 44 out of 7459 autopsies. Esophageal polyps are the most common intraluminal tumors although its occurrence is rare. Depending on their histological features they can be termed as fibromas, fibro lipoma,and fibro vascular polyps/lipoma. In presence of mixed architecture they are called hamartomas(7). Esophageal hamartomas are classified based on their location in relation to the esophageal wall as intramural, extramural or intraluminal(1).

Pedunculated esophageal hamartomas are not true neoplasms,but represent developmental lesions of obscured origin. Possibly they are derived from nest of embryonic tissue associated with the development of the foregut and tracheobronchial groove (8). It has been suggested that combination of remarkable looseness of the sub mucous tissue in the upper esophagus together with active peristaltic movements with molding result in the development of these long pedunculated polyps(9).

It appears that these hamartomatous polyps produce few symptoms until they reach an impressive size. Dysphagia was most common symptom which was found in our patient.

According to a large study done by Manuel Caceres et al on 110 reported cases majority of patient presented with dysphagia (62%). Second most common symptom was regurgitation of the mass into the mouth (38%) (10).

Some other complication includes sense of lump in the thorax, partial regurgitation of fleshy mass in the mouth and its disappearance upon swallowing. Asphyxia due to laryngeal impaction of a regurgitated polyp has been well described in the literature (11). Weight loss if present is related to dysphagia and not to the size and growth of the polyp. Hematemesis or occult GI bleeding may occur and is probably related to ulceration of the tumor.

An appropriate initial evaluation in the patients presenting with these symptoms should include elaborative history and physical examination followed by barium swallow and upper GI endoscopy.

According to Manuel et al study 84% of this polyp was found to originate in the esophagus. Of these 76% had their pedicle attached at or near the upper esophageal sphincter, 15% were attached in the upper esophagus, 3% in the middle esophagus and 6% in lower esophagus(10). Hamartomas are pathologically sub classified according to the relative abundance of a particular endogenous tissue component, and the variants described include chondroid, chondromesenchymal, angiomatous, lipomatous and leiomyomatoushamartomas (12).

Grossly, these polyps usually appear pink and smooth. They range in consistency from firm to rubbery. Occasionally they are multilobate and 19% have ulcer present on their most distal aspect (10).

Malignant transformation of these large polyps occurs infrequently. There are no tumor markers or imaging characteristics that allow a definitive preoperative diagnosis of esophageal hamartomas, and the majority of the cases are diagnosed following surgical resection or at autopsy. The diagnosis is based entirely on histopathological evaluation. The differential diagnosis includes choristoma and teratoma.

Because of potentially disastrous complications, surgical excision of benign hypopharangeal and esophageal polyps is strongly recommended. The possibility of progressive or complete obstruction of the esophagus or sudden obstruction of larynx warrants prompt excision once diagnosis is established.

This can be accomplished using a transoral, transcervical, or transthoracic approach,with strategy dependent on the location and size of the polyp.

Hypopharangealpolyps have been removed by laryngoscopic or endoscopic ligation (80%) or through a cervical incision (20%).

The surgical approach to these large polyps should be decided on a case by case basis. Endoscopic ligation appears to be reasonable alternative to operative exploration if it can be accomplished safely in the hands of an experienced endoscopist. Of primary concern is the potential for large polyps to cause airway compromise during endoscopic retrieval and need to obtain haemostasis at the transacted pedicle, which is frequently well vascularised. If surgical removal is warranted, a left cervical approach is appropriate if the base of polyp can be identified with certainty in the hypo pharynx or upper esophagus. Otherwise, a thoracotomy may be necessary. The transthoracic approach, using an esophagotomy through the wall opposite to the lesion, allows complete excision and mucosal approximation under direct vision.

CONCLUSION

Large pedunculated esophageal and hypopharangeal polyps are relatively uncommon. They tend to occur in older male patients and their discovery is usually preceded by a history of progressive dysphagia, regurgitation of the mass, or the sensation of having persistent lump in the throat. Although the regurgitated mass is swallowed, this event has led to fatal asphyxiation on several occasions. The pedicle most frequently originates from the posterior wall of the esophagus at or near the upper esophageal sphincter (UES). Negative contrast and endoscopic studies should be accepted with caution and repeated if necessary. A CT scan and MRI may also identify the lesion. These lesions are almost benign and the method of polypectomy (endoscopic versus surgical) is determined on a case by case basis. Malignant degeneration is unusual and following polypectomy, these lesions rarely recur.

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