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Case Report

A Giant Hypopharyngeal - Oesophageal Fibrovascular Polyp

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Abstract

Fibrovascular polyp is a rare benign tumour of the hypopharynx and esophagus. It presents in a vague and non-specific way which can pose a diagnostic dilemma for many clinicians. Our objective in presenting this particular case is to introduce this extremely rare diagnosis and management of giant hypopharyngeal-esophageal fibrovascular polyp.

We present a 37-year-old man who was referred to the Ear, Nose and Throat (ENT) unit of Korle-Bu Teaching Hospital on account of a one-year history of foreign body sensation in the throat, dysphagia, discomfort in the throat and regurgitation of fleshy mass in the throat which leads to intermittent choking spells, asphyxiation and difficulty breathing. There was no cough, hemoptysis or weight loss.

He had been treated with several medications at several health facilities as pharyngitis and reflux disease with not much improvement until he was finally referred to our facility when symptoms of choking, asphyxiation and dyspnea were worsening.

On examination was a young healthy man with normal physical examination including ear, nose and throat. Flexible nasolaryngoscopy was revealed a suspicious lesion in the left vallecular- piriform fossa area but all other subsites were essentially normal. A computed tomography (CT) scan done was reported as a uvular mass which was inconsistent with our clinical suspicions. So, the CT films was interpreted by surgical team and revealed a polypoidal mass arising from the left piriform fossa and extending into the cervical esophagus. The other laboratory investigations done including FBC, sickling test, BUE Cr were essentially normal.

Diagnosis was made mainly based on clinical history and examination finding of the regurgitated fleshy mass in the throat as well as the CT scan films and suspicious flexible endoscopic findings.

The patient had an examination under anesthesia of his pharynx and esophagus using a rigid esophagoscope after which showed a huge polypoidal mass filling the left pyriform fossa and esophageal lumen. A lateral pharyngoscopy and excision of the hypopharyngeal polyp was done. Histopathology confirmed the diagnosis of angiolipoma.

Patient recovered fully on the ward and started normal diet after 10 days of nasogastric feeding. He is currently stable one month after surgery and is doing well almost one-month post operation.

When a young person presents with foreign body sensation in the throat with associated dysphagia, choking spells it should not be ignored and treated as reflux laryngitis, pharyngitis, or as psychogenic in nature especially when symptoms do not improve over a long period of time.

Our case emphases the need to take thorough history and take time to listen to patient for him or her to demonstrate all their symptoms and signs in order to make accurate diagnosis despite conflicting or uncertain findings on CT reports and endoscopy. It is also important to read the CT images ourselves as surgeons and to not only use reports which may contradict our clinical diagnosis or mislead us. It is important to read on any case that poses as a diagnostic challenge to point us in the right direction in our day to day case management.

Keywords: Hypopharynx; Giant; Polyp; Fibrovascular; Angiolipoma

Introduction

The presentation of hypopharyngeal or upper esophageal polyp can be vague and pose a real diagnostic challenge to most clinicians. Fibrovascular polyps of the esophagus are rare tumors. They usually arise close to cricopharyngeal muscle. Symptoms include dysphagia and regurgitation of the fleshy mass into the mouth, which can cause asphyxia [1]. Giant fibrovascular polyps of the esophagus and hypopharynx are rare benign esophageal tumors. They arise most commonly in the upper esophagus and may, rarely, originate in the hypopharynx. They can vary significantly in size. Even though they are benign, they may be lethal due to either bleeding or, rarely, asphyxiation if a large polyp is regurgitated [2].

We present a giant fibrovascular polyp in a 37- year-old man which had posed a diagnostic challenge for a year prior to referral for excision.

Case Summary

37-year-old healthy male who has no underling co-morbidities. He presented to our unit with a year's history of foreign body sensation in the throat with associated progressive dysphagia to solids, throat discomfort. He also complained of intermittent episodes of choking or asphyxiation associated with difficulty in breathing worse at night and regurgitation of a long fleshy polypoidal mass into the mouth which he could push back into his throat and swallow, using his fingers. He however denied any sore throat, hemoptysis, cough or aspiration, weight loss, hoarseness, stridor or neck swelling.

He had no chronic comorbidities except for benign lumps which had been excised from his breast on two occasions 6 and 10 years respectively prior to this presentation. He works as an administrator in a private company. He had no significant family history as well. He had a strong alcohol history of beer and spirits but denied any tobacco use. He is married with four children.

On examination, a well looking male healthy male was seen. Ear, nose and Oral and oropharyngeal examination were essentially normal as neck examination and systemic evaluation. The patient however volunteered to demonstrate the fleshy mass from his mouth which revealed a long pedunculated lesion which he could hold with his fingers. The base of the lesion was seen to be coming from the left side of the of tongue or vallecular region and it looked like a smooth mucosal polypoidal and irregular mass with some cystic component. The patient started complaining of shortness of breath and said he needed to push it back into the mouth which he did using his fingers and then swallowed it back. This dramatic demonstration was again repeated. Flexible nasolaryngoscopy was done which was suspicious of a left mucosal lesion involving left vallecular and piriform fossa region. At the time of endoscopy he did not regurgitate the mass.

Figure 1: Regurgitated poly on the oral cavity.

Figure 2: Patient holding the polyp.

Computed tomography scan was done which revealed a left piriform fossa -upper cervical esophageal polypoidal lesion.

Figure 3: CT Scan showing polyp in upper esophagus.

Figure 4: CT Scan showing polyp starting from the hypopharynx.

Initially rigid pharyngoscopy and esophagoscopy was done to confirm the lesion and the possibility of endoscopic excision which was abandoned on account of the size of the mass. There was a soft mobile intraluminal polypoidal mass with normal overlying mucosa. Trans-cervical excision of the upper esophageal lesion revealed a soft pedunculated 6cm by 10 cm multilobulated mass with a broad pedunculated attachment to the medial aspect of the left piriform fossa.

Operative procedure

under orotracheal intubation patient was cleaned and draped after which rigid pharyngoscopy and esophagoscopy was done and lesion assessed.

Figure 5: Endoscopic view of the polyp intra op.

Horizontal skin crease incision was made and deepened through the subcutaneous tissues and platysma layers.

Figure 6: Skin incision.

Figure 7: Polyp being excised trans cervical approach.

Figure 8: Defect in the hypopharynx after excision.

Subplatysmal flap was then raised superiorly and inferiorly. The medial wall of the sternocleidomastoid muscle was retracted laterally after which the superior constrictor muscles identified at the level of the left piriform fossa near the superior horn of the thyroid cartilage. A short rigid pharyngoscope was then passed into the hypopharynx and upper esophagus which confirmed the exact location of the hypopharyngeal mass. The superior constrictor muscles were then incised vertically at the left pyriform fossa region till the intra-luminal polypoidal mass was seen. The mass was then excised in toto and the hypopharynx was repaired in three layers with Vicryl 2 0 after cricomyotomy was performed and nasogastric tube had been passed. The wound was then closed in two layers; Vicryl 2 0 to subcutaneous layer and then Nylon 3 0 to skin after which a corrugated wound drain had been inserted. Wound dressing was then applied and anesthesia reversed.

Histopathology finding

Gross examination showed soft polypoidal mass with external surface mucosa with multiple polypoidal projections.

Figure 9: Gross specimen after excision.

Microscopic examination showed polypoidal tissue bits lined by nonkeratinized stratified squamous epithelium. Underlying subepithelial tissue show islands of mature adipose tissue intermixed with blood vessels of varying sizes and thickness with vascular ectasia. Moderate chronic lymphocytic infiltrate with formation of lymphoid aggregate are noted. No evidence of granuloma, atypia or malignancy is seen.

Progress report

Patient remained on admission for a period of 11 days. He was placed on nil per os and nasogastric tube feeding, analgesia, antibiotic and anti-reflux medications were commenced. On post-operative day one he complained of difficulty in swallowing even his own saliva, aspiration and later inability to lie down flat and difficulty in breathing for which he was given intravenous dexamethasone for twenty-four hours. He had the nasogastric tube repassed under endoscopic guidance on postoperative day four after several failed attempts after it was noticed to be kinked on the postoperative day one.

Patient's stay on the ward was unremarkable and he is now able to eat all types of food without any significant aspiration. His symptoms however improved over the period of his stay and he was discharged home 11 days after surgery. One month after surgery patient is tolerating all feeds and has no significant concerns. We plan following up every 3 to months over the next 2 years or more.

Discussion

Fibrovascular polyps are very rare benign neoplasms of the hypopharynx that can occur from any site in the pharynx as well as the upper esophagus. Though biologically non-cancerous, these giant fibrovascular polyps can have startling and even life-threatening presentations. A recent patient encounter has prompted us to review this rare condition.

Fibrovascular polyps of the esophagus and hypopharynx are the most common intraluminal tumor but are extremely rare [3], accounting for less than 1% of all hypopharyngeal and esophageal tumors [3]. They are also known as giant benign esophageal polyps, polypoid fibromas, giant fibrovascular polyps, myomas, fibro lipomas, pedunculated lipomas, fibroepithelial polyps, and lipomas of the hypopharynx [4]. In 1922, Vincent was the first to surgically excise a fibrovascular polyp, and by 1957, only 6 cases were reported from the Mayo Clinic [5].

Giant fibrovascular polyps are defined as polyps larger than 5 cm in maximum diameter which qualifies our case as one. To

date, there are just over 100 reported cases in the literature and the largest single series consists of 16 patients [6,7] but those involving the hypopharynx are extremely rare. They are slow growing, pedunculated tumor masses that often arise from the upper esophagus, near the level of the cricopharyngeal at the pharyngo-esophageal junction, and this area has also been termed as the Laimer-Haeckermann triangle (also known as the Laimer's triangle) [8]. Because of the mixture of stromal tissues, it is hypothesized that the fibrovascular polyp may be a hamartoma or an unusual type of inflammatory polyp. Another theory is that they arise secondary to mucosal injury [9], gastroesophageal reflex or ectopic gastric mucosa [10].

Our patient is a 37-year-old male and this is similar to what is found in literature. These are usually solitary lesions (rarely multiple) occurring predominantly in males in the third to fifth decades of life. In addition, however majority of the esophageal polyps are in the elderly in their 60s and 70s even though a case has been reported in an infant [4]. Most are large and pedunculated, with a stalk or pedicle attached to the region of the cricopharyngeal muscle [4]. Originating from any site in the pharynx or upper esophagus, they start as small sessile lesions which elongate due to peristalsis within the esophagus and the looseness of the submucosa [4]. The common origin for the mass was the cricopharyngeal part of the hypopharynx [3].

Fibro vascular polyps are composed of loose and dense fibrous tissue, adipose tissue and vascular structure and are covered by normal squamous epithelium. Depending on which histologic component predominates these lesion have been called, lipoma, fibroma, fibro lipoma, fibro myxoma, angiolipoma, fibroepithelial polyp [11]. Histological diagnosis of our patient was angiolipoma. Angiolipoma is one of the most common benign soft tissue tumors. Angiolipoma occurring at the hypopharynx-esophageal introitus is extremely rare in clinical practice. Angiolipoma is a distinct morphological variation of a lipoma, accounting for 5-7% of lipomas [12]. In a few cases, familial heredity was observed [13]. It commonly occurs in young men's arms and trunk, and it can also occur in the oral cavity and digestive tract. Familial angiolipomatosis has a predominantly autosomal-recessive inheritance, but some studies also show an autosomal dominant mode of inheritance [13].

Patients can remain asymptomatic for many years and symptoms occur when the polyp reaches a large size. Due to their size and mobility, they can be regurgitated and can cause asphyxiation or require emergent airway management [3]. Due to the indolent nature of these polyps and the potential space the esophagus provides, these fibrovascular polyps can grow up to considerable sizes without causing many symptoms till late, measuring as big as 26 cm in largest diameter [8]. Ease of diagnosis of fibrovascular polyps in the hypopharynx and esophagus depends on the location of the lesion and clinical presentation. When these tumors are situated further into the aerodigestive tract, however, the diagnosis may be challenging [3]. In the absence of an obvious regurgitation of the polyp during examination, diagnosis can be a challenge [14] and up to 30% of patient may die without a correct diagnosis [11].

Our patient's symptoms were classically noted to include dysphagia, a mass in the throat and regurgitation of the polyp into the mouth with its disappearance on swallowing [11,15]. Asphyxiation can result from impaction of the polyp in the glottis and is the most feared complication which was our patient's main concern [1].

In the literature, dysphagia was the most predominant complaint (present in 87%), followed by respiratory symptoms (25%) and regurgitation of the polyp into the pharynx or mouth (12%). The other reported non-specific symptoms included epigastric pain, odynophagia, non-exertional substernal chest pain, loss of weight, persistent cough and in some cases, gastrointestinal bleeding [14]. The main clinical manifestations of angiolipoma at the hypopharynx-esophageal introitus were dysphagia and intermittent chest pain. Our patient however denied any symptoms of intermittent chest pain.

Our diagnosis of this patient was made mainly by clinical judgement and radiological and endoscopic suspicion. Diagnosis can be made by a combination of clinical history and various investigations such as endoscopy and imaging studies like barium swallow studies, endoscopic ultrasonography (EUS), CT and magnetic resonance imaging (MRI). We did not conduct a barium study because of patient's worsening symptoms which needed urgent surgery. Barium studies are commonly used and can show up the characteristic appearance of a smooth intra-luminal sausage-shaped mass with bulbous tips, with varying degrees of lobulation

[16]. EUS can be a useful adjunct as it provides information on the size, origin of the stalk and vascularity of the polyp. In addition, EUS fine needle aspiration may provide a more diagnostic histological sample than a superficial biopsy from an endoscopic approach. This procedure is not offered in our facility. Ct report provided was not consistent with our clinical suspicion but on reading the imaging a suspicious polypoidal mass was seen in the left piriform sinus and extending into the esophagus. CT has been recommended by some; in addition to providing information on the components of the tumor, at an early arterial phase, especially in large polyps, feeding vessels can be visualized which can aid in surgical planning [2]. The two most common methods of diagnosis are barium swallow and esophagoscopy and both are usually necessary. Biopsy is not recommended because these polyps are quite vascular [17].

In this case we did not have presurgical diagnosis of angiolipoma however it is believed that the presurgical diagnosis and differential diagnosis of angiolipoma mainly rely on endoscopic manifestations, ultrasonic endoscopic changes, CT, and postoperative pathology. Because angiolipoma is rich in blood supplies and it is difficult to get the tissue from the biopsy at the mucosal surface, hence endoscopic biopsy is not recommended [17].

The mainstay of treatment once diagnosis is achieved is surgical excision in view of the potential risk of respiratory compromise, bleeding and the debilitating symptoms. Tumor excision is the only treatment for fibrovascular polyp. Sudden asphyxia may occur due to this tumor, and thus, surgical treatment is required as soon as possible after encountering this tumor. Other reasons for excision include the possibility of malignancy and symptomatic improvement [18]. This agrees with our decision to do urgent surgery for our patient. The surgical options include endoscopic resection, open surgery via transcervical, trans-thoracic or trans-abdominal approaches. This should be tailored on a case-to-case basis and a combination of the approaches should be considered in difficult cases [2]. However, it is important take note, at this point, the value and accuracy of pre-operative assessment of the base of the pedicle and the bulk of the tumor mass and clear resection of the base to prevent recurrences. There are 2 options: surgery or endoscopic resection, based on the size, the location of the stalk's base, its mobility, and the vessels that nourish it. The Fibrovascular Polyp less than 2 cm can be removed by endoscopic resection. But those larger than 5 cm usually require open surgical resection, because of the thick vascularized pedicle as was seen in our case [8]. In most cases, the left-sided cervical is the preferred approach similar to our case. The classical approach is cervical esophagotomy with ligation of base and excision of polyp [11] which was opted for our patient mainly based on size. Due to the thick vascularized pedicle, hemostasis is most safely achieved by open surgical techniques [8]. If the lesion is huge, a thoracotomy or a laparotomy may be necessary. In cases of recurrent giant Fibrovascular Polyp patient underwent several surgeries via a combination of endoscopic, trans-oral, trans-cervical, trans-thoracic and trans-abdominal approaches [2]. Recently, endoscopic resection with ultrasonic shears, a needle-type knife, and an electrosurgical snare has been performed [19]. The Scissor type knife facilitates grasping the pedicle and enables simultaneous excision and coagulation while maintaining a stable view by use of only a single device [20].

The more common differential diagnosis to consider includes leiomyomas, leiomyosarcomas, squamous papilloma's, lymphomas, spindle cell carcinomas and hemangiomas amongst others. It is also worth noting that atypical lipomatous tumor (ALT) and Fibrovascular Polyp symptoms are very similar, but ALT is a rare, low-grade malignant neoplasm [21]. Malignant transformation is rare but has been reported in esophageal polyps. The lipomatous components can undergo sarcomatous changes, the squamous mucosa can develop into squamous carcinomas and small polyps have developed into adenocarcinoma.

The recurrence of FP after resection within years is very rare, so patients should undergo CT and endoscopic surveillance for several years [2]. For this reason our patient will be followed up for several years to come.

Conclusion

Clinical examination may be normal in patients presenting with fibrovascular polyps and a high index of suspicion is needed in diagnosing this extremely rare condition. This is even more so in the scenario where there is no regurgitation of fleshy mass into the mouth. Our case emphases the need to take thorough history and take time to listen to patient to demonstrate all their symptoms and signs in order to make accurate diagnosis despite conflicting or uncertain findings on CT reports and endoscopy. It is also important to read the CT images ourselves as surgeons and to not only use reports which may contradict our clinical diagnosis. It is important to read on any case that poses as a diagnostic challenge to point us in the right direction in our day to day case management.

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Conflict of Interest

No conflict of interest to declare.

This manuscript is not under consideration for publication by any other journal.

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