



# An atypical lipomatous tumor mimicking a giant fibrovascular polyp of the hypopharynx

### A case report

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#### **Abstract**

**Rationale:** Giant fibrovascular polyps (GFVPs) found in the hypopharynx are exceedingly rare. These are benign tumors which are identified by CT or MRI and usually treated based on symptoms. Even more rarely, pathology may identify one of these masses as an atypical lipomatous tumor (ALT). This paper will present a case of an ALT of the hypopharynx that was originally classified as a GFVP, highlighting the difficulty in distinguishing between them and the importance of making the correct diagnosis.

**Patient Concerns:** An 84-year-old man presented to the emergency department with a 6-month history of a pedunculated hypopharyngeal growth, dysphagia, and intermittent dyspnea.

**Diagnoses:** The mass was characterized as a GFVP by barium swallow and MRI.

**Interventions:** The hypopharyngeal mass was resected for obstructive symptoms and to confirm the diagnosis. Final pathology found the mass to be more consistent with an atypical lipomatous tumor (ALT).

Outcomes: The patient's dysphagia and dyspnea resolved. He was free of recurrence at 22 months postoperative.

Lessons: Both GFVPs and ALTs are very rarely found in the hypopharynx but can be easily misclassified as one another. Imaging is useful to initially characterize the mass, but to definitively differentiate between them, pathological analysis is necessary. Although they are rare, it is important to consider both possibilities on the differential for hypopharyngeal masses. Further, accurate analysis is essential to distinguish between them because their definitive management and follow-up is different.

**Abbreviations:** ALT = atypical lipomatous tumor, FVP = fibrovascular polyp, GFVP = giant fibrovascular polyp.

Keywords: case report, giant fibrovascular polyp, hypopharynx, liposarcoma

#### 1. Introduction

Giant fibrovascular polyps (GFVPs) are rare benign tumors that can be found in the esophagus or, rarely, in the hypopharynx or larynx. <sup>[1,2]</sup> The differential diagnosis for a fibrovascular polyp (FVP) includes lipoma, hamartoma, inflammatory polyp, hemangioma, lymphangioma, schwannoma, carcinoid tumor, chemodectoma, <sup>[3]</sup> and achalasia. <sup>[2]</sup> FVPs can only be definitively diagnosed upon histopathological evaluation. <sup>[2]</sup> If the specimen does not fulfill the histopathological criteria for a FVP, then an alternative diagnosis should be considered.

Atypical lipomatous tumors (ALTs) are primary malignant tumors that can masquerade as a GFVP, but are rarely found in the hypopharynx or esophagus. [4] They are identified on histopathological analysis by the presence of adipocytes of variable size, atypical multivacuolated lipoblasts, and nuclear atypia in adipocytes and spindle cells. [4] The most important reason that these tumors need to be differentiated from FVPs is that they are malignant and must be excised. They also have the potential for recurrence so patients must be followed up for signs of local recurrence closely postoperatively. This article will present the case of a patient who presented with what was initially thought to be a GFVP, but was found to be an ALT upon histopathology. This is unique as FVPs of the hypopharynx are rare to begin with, and ALTs presenting as such as rarer still. It is important to differentiate them as there are considerable differences in their management.

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The authors report no conflicts of interest.

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#### 2. Case presentation

Timeline is shown in Table 1.

An 84-year-old man presented to the emergency department with a 6-month history of a progressive growth in the oral cavity. At the time of presentation he was experiencing some dysphagia and intermittent dyspnea. He did not report any pain, hemoptysis, numbness, or changes to his sense of taste. He also did not report any B-symptoms (fever, night sweats, or weight loss). On questioning, he disclosed that sometimes the mass could be seen and other times it was not visible. The patient's medical history was also significant for diabetes, hypertension, coronary artery disease, prior cardiac stenting and coronary artery bypass grafting, atrial fibrillation, gastroesophageal reflux disease, Parkinson disease, hyperlipidemia, and hypothyroidism.

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#### Table 1

#### Timeline of patient presentation, intervention and outcome.

Date	Action
~June 2012	Perceived onset of growth in the oral cavity.
January 5, 2013	Presentation to the emergency department with progression of growth, dysphagia, intermittent dyspnea.
January 30, 2013	Initial otolaryngology consultation. Pedunculated mass seen on physical examination and nasolaryngoscopy. MRI and barium swallow ordered. Plan for surgery.
February 27, 2013	Endoscopic surgical resection of mass.
February 28, 2013	Discharged from hospital stable on postoperative day 1.
March 2013	Pathology returns as atypical lipomatous tumor. Discussed at sarcoma rounds with radiation oncology and surgical teams. Decided no further treatment necessary.
April 2013 to December 2014	Follow-up appointments every 3 to 6 months. No return of symptoms. No evidence of recurrence on physical examination or on repeat MRI in December 2014.
January 2015	Patient died of cardiac arrest.



Figure 1. The mass pre-excision in the oral cavity.

On physical examination, there was a long pedunculated lesion to the left side of the oral cavity, next to (and distinct from) the tongue (Fig. 1: view of the endoscopic approach). The examination was unable to discern the site of attachment from the initial examination. The distal aspect of the lesion was slightly ulcerated. The neck was soft with no cervical lymphadenopathy. On nasopharyngoscopy, it was evident that the stalk of the lesion

extended into the larynx and likely originated from the left aryepiglottic fold. The vocal cords were mobile bilaterally and no other abnormal masses were seen from the larynx to the glottis.

A barium swallow was then ordered to further characterize the mass. The investigation showed a moderately sized FVP but no obstruction of the hypopharynx or esophagus. Investigations were continued with an MRI of the neck, which was unable to

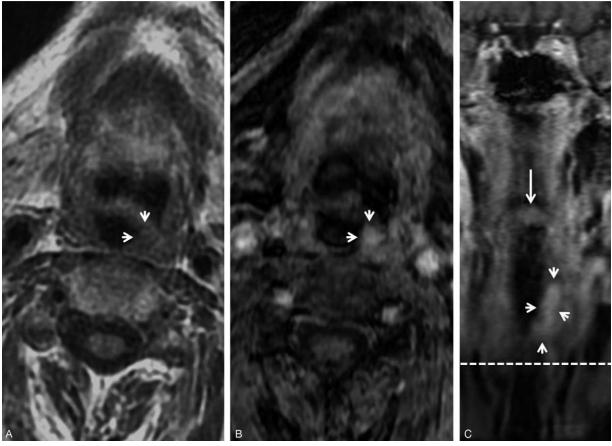


Figure 2. Axial T1 weighted MRI images pre (A) and post (B) intravenous gadolinium administration demonstrates the presence of a small enhancing lesion (white arrow heads) at the top of the left aryepiglottic fold. This is seen on the coronal post gadolinium fat saturated T1 weighted sequence (C) to be an elongated lesion cranial to the level of the glottis (white dotted line). The long white arrow indicates the uvula.

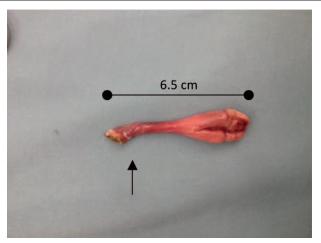


Figure 3. Mass after excision. Arrow points to the stalk end. Total length of mass is 6.5 cm.

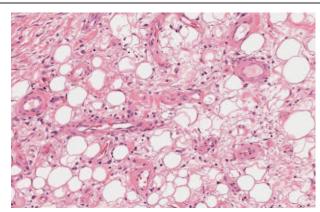


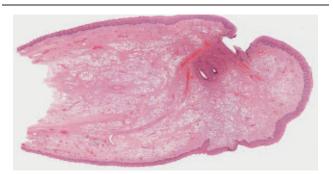
Figure 5. Pathological image of the specimen at  $20 \times$  magnification. The image demonstrates variability in adipocyte size, atypical stromal cells, and scattered lipoblasts, features characteristic of well-differentiated liposarcoma/ALT. ALT = atypical lipomatous tumor.

identify the mass or its site of origin. However, the MRI was able to pick up some questionable thickening and increased signal in the left hypopharynx (Fig. 2: MRI images). The mass was originally thought to be a giant FVP of the hypopharynx.

Because the patient was symptomatic, he consented to surgical resection of the mass. The lesion was found to be attached to the left medial piriform sinus mucosa. The surgeon removed the mass endoscopically through a Dedo laryngoscope using bipolar and monopolar cautery to remove the stalk but preserve the underlying mucosa (Fig. 3: mass after excision). No other pathology was found during laryngoscopy and rigid esophagoscopy.

Pathology demonstrated a polyp that was measured to be  $3.5 \times 1.5 \times 0.8$  cm with a stalk that was 3.0-cm long and 0.4-cm wide. It was solid, granular, smooth, and light tan in color. The polyp had a core of fibrovascular adipose tissue (Fig. 4: pathology  $0.8 \times$  magnification), but on further examination there was variability in the size of the adipocytes, a significant number of spindled to stellate stromal cells and many hyperchromatic cells. The findings were consistent with an ALT mimicking a GFVP (Fig. 5: pathology  $20 \times$  magnification). The resection margin was negative for lesional cells

Postoperatively, the patient symptoms of dysphagia or dyspnea resolved. He also did not report any other new symptoms, such as hoarseness, changes in voice, change in weight, heartburn, pain,



**Figure 4.** Pathological image of the specimen at  $0.8\times$  magnification demonstrating the polypoid nature of the mass. The squamous mucosa is intact and the polyp core is composed of fibrovascular adipose tissue. These findings indicate that a GFVP is a diagnostic consideration. GFVP = giant fibrovascular polyp.

otalgia, or neck masses. The patient could tolerate an oral diet and was sent home the day after surgery. There was no evidence of recurrence 22 months after the tumor removal, both by physical examination and by MRI, but the patient continued to be monitored for signs of tumor recurrence. The patient died 23 months post operatively of cardiac arrest with no evidence of tumor recurrence. Ethics approval was not required for this paper as it is a case report. Patient consent was not obtained as the patient died prior to the writing of this report.

#### 3. Discussion

This article presents the unique case of an 84-year-old man who presented with a hypopharyngeal mass that was initially thought to be a GFVP. On pathology, the mass was found to be a liposarcoma (ALT). This case was highly unusual because liposarcomas are very rarely found in the hypopharynx. It sheds light on the fact that although liposarcomas are not often found in this region, they should be kept on the differential when presented with pharyngeal lesions.

#### 3.1. Fibrovascular polyps: presentation and investigation

FVPs are rare, benign non-neoplastic masses that have been described mostly in case reports to date. [5] They are slow growing, pedunculated masses that usually arise from the upper third of the esophagus, close to the cricopharyngeus muscle. [6] These lesions appear virtually identical to ALTs and must be identified by histopathologic methods to give a definitive diagnosis. The origin of FVPs is unknown, but the currently accepted theory is that they develop as small polypoid lesions on the esophagus that elongate as a result of peristalsis and looseness of the submucosal tissue. [6,7]

FVPs present as a smooth, pedunculated, elongated mass. Clinical signs and symptoms can include dysphagia such as regurgitation of a fleshy mass into the pharynx or mouth, globus sensation, a sensation of food sticking in the upper chest, or odynophagia. Respiratory symptoms can include dyspnea, asphyxiation, choking, wheezing, or inspiratory stridor. Chest pain or constitutional symptoms such as weight loss can also be present.<sup>[8]</sup>

Several different modalities are used to visualize masses in the pharynx. The clinical examination is usually used first but can be limited by location and size of the mass or, as in this case, the fact that the mass may be swallowed and hidden periodically. Nasopharyngoscopy or esophagoscopy may help to improve visualization of the lesion. The majority of FVPs are diagnosed by a radiographic study of the esophagus using barium as a contrast medium. A barium swallow can show the length and size of the polyp as well as its location. I A positive investigation will show a crescent-shaped intraluminal filling defect in the case of a GFVP. However, too much radiocontrast can obscure the mass and make a GFVP look like a dilated esophagus suggestive of achalasia. This can lead to an incorrect diagnosis, significantly greater patient suffering in the long term, and the possibility of asphyxiation if the GFVP is not identified. Simple radiography is less useful in characterizing the lesion, but may show a lobulated mass in some patients (up to 50%).

Further investigations such as CT or MRI imaging are used to further characterize the polyp. CT scan can characterize the homogeneity or heterogeneity of the polyp's make-up by showing the difference between soft tissue and fat in the core of the polyp. MRI may be able to distinguish areas of hemorrhage or necrosis. [10] MRI, when available, is usually used to further define the location and characteristics of the mass prior to surgery. While MRI can be a valuable tool for characterizing the mass, this case has shown that MRI should not always be relied upon. This patient's MRI was unable to define the mass or its location. Prior to surgery, the only investigation that gave information about the polyp was the barium swallow, which has been previously described as the best diagnostic tool for this type of polyp. [9] That being said, others have been able to use MRI in prior cases with success so it should not be ruled out as a diagnostic tool.

## 3.2. Liposarcomas: presentation, classification, and treatment

Liposarcomas, or lipomatous tumors, are rarely found in the hypopharynx. However, when they do occur in this region they are usually of the well-differentiated or myxoid type, are found to have a lower grade, and carry a better prognosis than those found in the rest of the body. [11] It is unknown how liposarcomas of the head and neck region develop, but the currently accepted theory is that they arise de novo and do not differentiate from lipomas or other benign tumors. [12] However, case reports of benign tumors differentiating into liposarcomas have been reported. [13] Genetic factors may also play a role as patients with neurofibromatosis type 1 have been noted to have elevated rates of liposarcoma. [14] Liposarcomas are usually identified on barium swallow, where they present with a smooth-walled filling defect and a widened esophagus. [15] To distinguish a liposarcoma from a more benign tumor, like a GFVP, pathological analysis is necessary. Liposarcomas are classified into 4 types: well differentiated, myxoid, pleomorphic, and round cell. The liposarcoma in this case was a well-differentiated liposarcoma, which is the most common type. Three subtypes of well-differentiated liposarcomas are recognized: adipocytic, sclerosing, and inflammatory. The adipocytic subtype is characterized by mature adipocytes of variation in size, focal nuclear atypia, and hyperchromasia. [16] Lipoblasts may also be found, but do not make or exclude a diagnosis of adipocytic well-differentiated liposarcoma. [16] The sclerosing subtype is usually found in the retroperitoneum and is characterized by the presence of distinctive stromal cells, multivacuolated lipoblasts, and a fibrillary collagenous background. [16] The inflammatory subtype is characterized by the presence of an inflammatory infiltrate, usually with a predominantly B-cell population but occasionally with a predominantly T-cell population. [16]

The recommendation for management of liposarcomas is surgical excision. Clear margins are important to minimize the risk of recurrence, but are not always possible due to anatomical constraints. The use of adjuvant therapy may be indicated in patients with a positive margin or incomplete excision. [13] The patient described in this case was managed by a suspension laryngoscopy approach, but this is not always possible. Endoscopic resection is useful when, as in this case, there is evidence of a tumor peduncle or potential for curative resection. [17,18] Other techniques include excision by lateral pharyngotomy, microlaryngoscopy, or total laryngectomy. The choice of excision technique depends on the accessibility and location of the mass, as well as the ability to resect with wide margins, among other factors. [19] On review of cases in the literature, the majority of these masses has been resected by simple excision or lateral pharyngotomy. [18] There are reports of late recurrence of liposarcomas which means that long-term follow-up is necessary. The patient in this case was followed up until his death (from unrelated causes). A follow-up MRI was scheduled for 1.5 to 2 years postoperatively to monitor the surgical site, which showed no signs of recurrence.

#### 4. Conclusion

This report describes a unique case of a patient presenting with a liposarcoma of the hypopharynx that originally presented as a GFVP. This is unusual since both types of masses are rare, especially so when found in the hypopharynx. When investigating masses of the hypopharynx, histology is essential and liposarcomas should be kept on the differential diagnosis because, although rare, they change future management and require surgical excision and significant follow-up.

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