BENIGN AND MALIGNANT LIPOGENIC TUMORS OF THE RETROPHARYNGEAL SPACE

DK Chhetri

Division of Head and Neck Surgery,
62-132 CHS, UCLA Medical Center,
Los Angeles, CA 90095, USA

Correspondence to:
Dinesh K. Chhetri, M.D.
62-132 CHS, Div. Head & Neck
Surgery, Los Angeles, CA 90095USA
E-mail:dchhetri@mednet.ucla.edu

Lipogenic tumors of the retropharyngeal space are neoplasms that can grow to a considerable size before becoming symptomatic. Careful inspection of preoperative imaging will heighten suspicion for malignancy, which is critical in planning treatment. In this study we compare clinical, radiologic, and histologic findings of benign and malignant lipogenic tumors of the retropharyngeal space.

Key words: lipogenic tumor, retropharyngeal

INTRODUCTION:

Lipoma is a common benign mesenchymal soft tissue tumor, which may arise within any site of the body. It occurs in the head and neck region in about 13% of all cases. It typically presents as a soft, mobile, subcutaneous mass without significant symptoms and is treated with simple excision. Lipomas of the retropharyngeal space are rare. Yoshihara et al. identified twenty-six cases previously reported in the literature.[1] There are many histopathologic subtypes, but microscopically are conventionally seen as a thinly encapsulated aggregate of mature adipocytes. In fact, it is commonly challenging to distinguish healthy adipocytes from adenomatous cells. Liposarcoma is the one of the most common sarcomas of adulthood. The head and neck region constitutes approximately 3% of all locations. Retropharyngeal liposarcomas are rare, with only 8 prior reported cases.[2] This includes one case of suspected radiation induced sarcomatous transformation. There are 5 pathologic subtypes, each with their differing prognosis. Treatment is primarily surgical without prophylactic nodal dissection. Chemoradiation therapy is added for poor histologic characteristics or positive surgical margins. At our academic institution we have had the unique experience of treating both lipoma and liposarcoma of the retropharyngeal space and noted the similarities of the clinical presentations as well as the radiographic differences. A retrospective chart review was completed to identify illustrative cases. We present these cases and compare and contrast clinical, radiographic, and pathologic findings of a retropharyngeal lipoma with a retropharyngeal liposarcoma.

CASE REPORTS:

Case 1: A 66-year-old African American female initially presented to an outside hospital with a one-month history of progressive dysphagia and airway obstruction. She was admitted and her workup included computed tomography (CT) of the neck showing a retropharyngeal mass. She was discharged with outpatient follow up with an otolaryngologist. The patient re-presented to the emergency room a few weeks later with complaints of neck fullness, further difficulty with breathing, and dysphagia. A repeat CT again showed the mass and also a marked narrowing of the supraglottic airway. She underwent a tracheostomy and an incisional biopsy. Because of her increasing dysphagia, the patient also had a percutaneous gastrostomy tube placed. The biopsy was consistent with lipoma. The patient was subsequently transferred to our institution. The CT neck revealed a large retropharyngeal space mass (6.2 cm anteroposterior, 9.1 cm transverse, and 14.0 cm craniocaudad) centered at the level of the hyoid bone with anterolateral herniation of the mass into the left submandibular space (Fig.1).

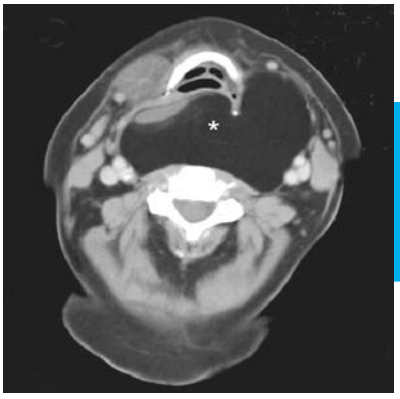

Fig: 1. CT scan of patient with large retropharyngeal mass (*) with slight herniation into the left neck. The fatty attenuation and homogeneity of the mass is consistent with a lipoma.

This homogeneous mass with fatty attenuation had occasional very fine hair-like septae noted within the center. The overall appearance was consistent with a lipoma. Her comorbid medical problems were optimized and she was taken to the operating room where a transcervical resection of the retropharyngeal mass was performed. There were no complications. Pathology confirmed a 10 x 9 x 4.8 cm lipoma without atypia (Fi. 2).

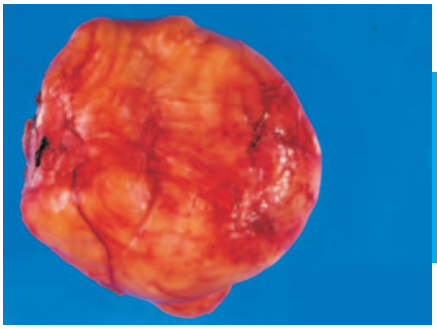

Fig: 2. Excised mass seen in Figure 1. Histopathology was consistent with a lipoma.

She did well following the surgery, and her tracheostomy tube was removed on the fourth postoperative day. She had no respiratory distress during her hospitalization and steadily regained normal vocal amplitude and quality over the course of her hospitalization. On postoperative day six she passed a bedside swallow study and was started on a regular diet which she tolerated without difficulty. Her PEG tube was subsequently removed and she has been asymptomatic after a 4-year follow-up.

Case 2: A 57-year-old Asian female presented to her primary doctor with a two month history of a rapidly enlarging left neck mass associated with dyspnea and dysphagia. On examination a left neck mass was appreciated and an urgent CT scan was ordered. The CT scan revealed a 2.5cm anteroposterior, by 6.5cm transverse, by 12.5cm craniocaudad mass from the level of the oropharynx extending to the mediastinum with pronounced tracheal shift (Fig:3).

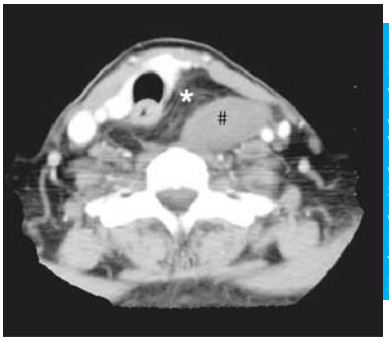

Fig: 3.CT scan of patient with large retropharyngeal mass (*) with fatty attenuation. There are prominent septations within the fatty attenuation areas compared to the mass seen in figure 1. In addition, a solid component is also seen posterolaterally (#). These features are suggestive of a liposarcoma.

The mass dissected between fascial planes to bilateral neck spaces with poorly defined borders. The mass showed fat attenuation with numerous thickened septae.There were no calcifications within the mass. Due to her worsening airway symptoms the patient was taken to the operating room and a tracheostomy and transcervical approach to this left neck mass was performed. The mass was noted to have a fatty consistency consistent with lipoma but more posteriorly and inferiorly had a solid component adherent to the esophageal muscular layer. This was resected without violation of the mucosa and the mass was delivered from the mediastinum without a sternotomy. Pathology revealed a 197-gram, 15 x 5.5 x 4 cm mass surrounded by fibrovascular capsule. The gross appearance was non-uniform, with one large gray-yellow 5cm nodule. Microscopic evaluation exposed a well-differentiated liposarcoma with de-differentiated component. The majority of the specimen consisted of relatively normal appearing adipose tissue with scattered enlarged, irregular, hyperchromatic nuclei. There was a smaller component that showed an abrupt transition with highly pleiomorphic cells in a fibrous background, including many cells having giant bizarre hyperchromatic and irregular nuclei with some being multinucleated. There was microscopic involvement of surgical margins at the left lateral esophagus as well as left lateral sternocleidomastoid muscle. The postoperative course was uneventful and the tracheostomy was decannulated and the patient started on peroral diet without incident. She was discharged home on postoperative day six. A postoperative CT showed no further evidence of tumor in the neck or mediastinum. Her case was reviewed at an interdepartmental sarcoma tumor board. Factors considered included the lack of residual gross disease and the morbidity of further resection, which would include laryngectomy and esophagectomy. The decision was therefore made to proceed with a combination of chemotherapy and radiation therapy for further management. After 4.5 years follow-up the patient is recurrence free.

DISCUSSION:

The retropharyngeal space is a potential space limited by the pharynx anteriorly and the prevertebral fascia posteriorly. The superior limit is the base of the skull and inferiorly, the fascial compartment ends within the superior mediastinum.The midline is divided by tough median raphe. Lymph nodes suspended within loose areolar tissue form the principal component. In their review, Yonus et al. compiled an exhaustive differential diagnosis of a retropharyngeal mass (Table 1).[3] Retropharyngeal lipomas are relatively rare. In 1940 Putney and Fry reviewed 15 cases presented in the literature from 1877 to 1934.[4]

Table: 1. Differential diagnosis of a retropharyngeal mass.

Developmental	Angiomatous Lymphoid Hamartoma
Traumatic	Chordoma Foreign Body Abscess Hematoma
Infectious	Acute/Chronic Abscess Lymphadenitis
Neoplastic	
Benign	Lipoma Fibroma Fibrolipoma Osteoma Osteochondroma
Malignant	Fibrosarcoma Lymphoma Liposarcoma Leiomyosarcoma Metastatic Carcinoma

Subsequently 12 additional cases were analyzed and further compiled by Yoshihara et al in 1998. Patient ages ranged from 3 months to 71 years, with a male to female ratio of 1.3:1.1 Various lipomatous subtypes such as infiltrative and ossifying have also been described with similar clinical presentations. [5-7] Retropharyngeal lipomas can grow to a considerable size before becoming symptomatic. The largest lipoma in Yoshihara's series was 17cm x 11cm on one side and 11cm x 8 cm on the other. These masses can also be found as an asymptomatic incidental finding. [7] Common presenting symptoms include dysphagia, globus sensation, sleep apnea, and dyspnea related to upper airway obstruction.[1,8-10] Lipomas typically arise from the subcutaneous tissue and rarely from deep soft tissue. Histologically they are composed of mature adipocytes without atypia and are often surrounded by a thin fibrous capsule. Lipomas can be diagnosed radiologically and findings on CT have been characterized as well-circumscribed masses with low attenuation related to fatty tissue (-50 to -150 Hounsfield units).[6] Therefore, given the benign nature, surgical excision is targeted at alleviating symptoms and/or obtaining the definitive diagnosis. However, for patients either not consenting to undergo surgery or unable to undergo surgery due to significant comorbidities, these cases can be successfully managed with expectant observation and serial imaging. [6,10] The natural course of retropharyngeal lipoma is therefore protracted without additional sequelae.

Liposarcoma is the malignant counterpart of a lipoma and most frequently arises in the deep soft tissue of the retroperitoneum and proximal extremities. The most recent World Health Organization classification of soft tissue tumors recognizes five categories of liposarcomas: well-differentiated, myxoid, round cell, pleomorphic, and dedifferentiated. Prognosis is correlated to the histologic subtype. Complete excision of well-differentiated and myxoid subtypes is considered curative with excellent 5-year survivals. Round cell and dedifferentiated subtypes have a high propensity for metastasis and 5-year survivals have been reported around 50%.[11] Liposarcomas of the head and neck are rare, thus retropharyngeal liposarcomas are rarer. Most recently, Ozawa et al. reviewed all 8 prior reports of retropharyngeal liposarcomas. [2] The youngest patient was 39 at the time of surgery. Well-differentiated subtype was the most common followed by myxoid type. All of the malignancies were excised with a cervical approach except for Ozawa's own patient, who underwent a transoral approach and resection. Follow-up for longer than 18 months has not been reported. [2]

A suspicion for liposarcoma is based on imaging studies. When evaluating a soft tissue mass with significant fat attenuation, it is crucial to hone in on suspicious radiographic findings. Kransdorf et al. compared both CT and MR images in order to distinguish between lipomas and well-differentiated liposarcomas. Thin septa were seen in both and were not significantly different. Through multiple logistic regression, only thick septa retained statistical significance for predicting liposarcoma. Calcifications were three times more common in liposarcomas than lipomas. However, 31% of lipoma images reviewed showed marked nonadipose areas, mainly correlating with fat necrosis, calcifications, and fibrosis with myxoid change. More specifically, many classically described liposarcoma findings can be also commonly found on lipoma imaging.[12] Galant et al. reviewed MR sequences of lipomas versus well-differentiated liposarcomas. The liposarcomas showed gross features resembling ordinary lipomas. 23% of lipomas contained non-characteristic findings such as thick septa, nodules, and heterogeneous hyperintensities. However, in well-differentiated liposarcomas, all but one case had findings of thick septa, nodules, or both. However, employing fat-saturated-T2 weighted STIR sequence imaging, hyperintensities are reported to have a 100% and 82% sensitivity and specificity respectively.[13] Suzuki et al showed the MRI presence of septa was significantly different between lipomas and liposarcomas. However, septa were still present in 10% of the lipomas. They also found no significant imaging diagnostic characteristics with the use of CT. Interestingly, FDG-PET could be used to quantify the metabolic activity of lipomatous lesions and can be useful in distinguishing between sarcomatous subtypes. However, the diagnostic accuracy with proposed Standardized Uptake Value (SUV) cutoffs are not dependable as of yet.[14] The clinical presentation our lipomatous masses of the retropharyngeal space did not reveal any significant differences. Notably however, the benign lipoma showed upper aerodigestive symptoms in a more rapid progression than its malignant counterpart. The severity of dyspnea and dysphagia has only been reported in a few retropharyngeal lipomas.[1] Both of our patients required tracheostomy prior to resection; however both patients were successfully decannulated in short order after the surgery. The comparison of the radiologic features between the benign and malignant masses is striking. Although the lipoma did show septae within the bulk of the mass, they were thin and fine than the septae of the liposarcoma. The septae of the liposarcoma were more numerous, thicker, and overwhelmingly abundant as displayed clearly within the coronal sections. Classically, sarcomatous calcifications were found in proximity to the lipoma and yet were absent from the liposarcoma imaging. The most telling radiographic finding was fascial plane dissection. Whereas the lipoma's bulk behaved as a singular bulk, displacing the normal fascial boundaries outwards; the liposarcoma's bulk seemed to dissect within tissue planes along the path of the retropharyngeal space. There was considerable mass effect in both CT scans, yet it was the liposarcoma which displayed a marked diffusely spreading growth pattern. It is this last characteristic that distinguishes the liposarcoma imaging from our benign case. It is also worth noting however, that histologically benign infiltrative lipomas can also display this investing appearance.[15] While three of the eight previously reported retropharyngeal liposarcomas required re-resection, the residual microscopic disease in our series responded well to postoperative chemoradiation.[2] The patient is four years post-treatment without evidence of residual disease, without the need for re-resection. Consistent with the literature of retropharyngeal lipomas, our report also notes resolution of presenting symptoms after complete excision. This patient is free of recurrence and doing well.

CONCLUSION:

Illustrative cases of lipogenic masses of the retropharyngeal space have been presented: a lipoma and a liposarcoma. Both patients presented with severe airway symptoms and both required surgical airway for respiratory distress. Both were imaged with CT scans, with the liposarcoma showing two potentially diagnostic features: thick septae and a growth pattern showing dissection through fascial planes. Transcervical approach was adequate for resection of both lesions. Postoperative chemoradiation was required for the liposarcoma due to positive microscopic margins. Both patients have been followed for over four years post-operatively without evidence of recurrent disease. In the evaluation of retropharyngeal lipogenic masses, it is valuable to identify radiologic subtleties that distinguish lipomas from liposarcomas for proper surgical planning and appropriate treatment.

REFERENCES:

1. Yoshihara T, Kawano K, Mita N. Retropharyngeal lipoma causing severe dysphagia and dyspnea. J Otolaryngol 1998; 27: 363-6.
2. Ozawa H, Soma K, Ito M, Ogawa K. Liposarcoma of the retropharyngeal space: report of a case and review of literature. Auris Nasus Larynx 2007; 34: 417-21.
3. Younus M. Retropharyngeal lipoma. J Laryngol Otol 1980; 94: 321-6.
4. Putney FJ, Fry KE. Retropharyngeal lipoma. Ann Otol Rhinol Laryngol 1940; 49: 967-72.
5. Eisele DW, Landis GH. Retropharyngeal infiltrating lipoma- a case report. Head Neck Surg 1988; 10: 416-21.
6. Rosell A, Garcia-Arranz G, Llavero MT, Martinez-San-Millan J. Imaging case study of the month. Lipoma of the retropharyngeal space. Ann Otol Rhinol Laryngol 1998; 107: 726-8.
7. Blanshard JD, Veitch D. Ossifying lipoma. J Laryngol Otol 1989; 103: 429-31.
8. Aland JW Jr. Retropharyngeal lipoma causing symptoms of obstructive sleep apnea. Otolaryngol Head Neck Surg 1996; 114: 628-30.
9. Di Girolamo S, Marinelli L, Galli A, Ottaviani F. Retropharyngeal lipoma causing sleep apnea syndrome. J Oral Maxillofac Surg 1998; 56: 1003-4.
10. Hockstein NG, Anderson TA, Moonis G, Gustafson KS, Mirza N. Retropharyngeal lipoma causing obstructive sleep apnea: case report including five-year follow-up. Laryngoscope 2002; 112: 1603-5.
11. Wong CK, Edwards AT, Rees BI. Liposarcoma: a review of current diagnosis and management. Br J Hosp Med 1997; 58: 589-91.
12. Kransdorf MJ, Bancroft LW, Peterson JJ, Murphey MD, Foster WC, Temple HT. Imaging of fatty tumors: distinction of lipoma and well-differentiated liposarcoma. Radiol 2002; 224: 99-104.
13. Galant J, Martí-Bonmatí L, Sáez F, Soler R, Alcalá-Santaella R, Navarro M. The value of fat-suppressed T2 or STIR sequences in distinguishing lipoma from well-differentiated liposarcoma. Eur Radiol 2003; 13: 337-43.
14. Suzuki R, Watanabe H, Yanagawa T, Sato J, Shinozaki T, Suzuki H, et al. PET evaluation of fatty tumors in the extremity: possibility of using the standardized uptake value (SUV) to differentiate benign tumors from liposarcoma. Ann Nucl Med 2005; 19: 661-70.
15. Senchenkov A, Werning JW, Staren ED. Radiographic assessment of the infiltrating retropharyngeal lipoma. Otolaryngol Head Neck Surg 2001; 125: 658-60.