

Case report

An atypical lipomatous tumor of the hypopharynx, a unique entity? A rare case report

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ABSTRACT

Introduction: Liposarcomas are thought to be the most prevalent kind of soft tissue sarcoma in adults. A well-differentiated liposarcoma that has a higher incidence of a local recurrence following surgical excision is referred to as an atypical lipomatous tumor. The incidence are extremely rare in which <1 % of head and neck sarcoma cases. This is unusual liposarcoma localization warrants great interest in reporting such a case.

Presentation of case: In this report we present a case of a 50-year-old male complained with difficulty swallowing solid food and continuous presence of lump in the throat. Fiber Optic Laryngoscopy (FOL) showed a tumor filled the hypopharynx and CT scan suggested a benign mass that was most likely a fibrolipoma.

Discussion: Tumor was found infiltrating the lateral pharyngeal wall and protruding into the hypopharyngeal lumen. Because the tumor had spread to the right thyroid lobe, surgical excision via a transcervical approach was combined with a right thyroidectomy. There was a positive margin at the end of resection, thus a chemoradiation was added. Two years post-operative evaluation shows no sign of recurrence.

Conclusion: The main treatment for hypopharyngeal liposarcoma is based on surgery, either through an endoscopic or transcervical approach which determined by the size of the tumor and surgical field. Adjuvant chemoradiation is given to help prevent the recurrence.

1. Introduction

Liposarcoma is defined as an adipose cell tissue-originated malignancy. The tumor is usually well-defined, tend to be attached to surrounding tissue, encapsulated, pedunculated, or polypoid, grows slowly and painlessly [1]. Approximately 20 % of all mesenchymal tumors originate from liposarcoma, which usually affect the trunk and extremities. The incidence of liposarcoma is <1 % among head and neck sarcoma cases [2]. Cases of hypopharyngeal liposarcoma are rare, dominant in males with an age range of 40–60 years [3,4]. Up until 2016, there were no >26 cases reported in the literature. According to the World Health Organization, liposarcoma are classified into four subtypes based on tumor histopathological findings atypical lipomatous tumor or well differentiated liposarcoma (ALT/WDLPS), dedifferentiated liposarcoma (DL), myxoid liposarcoma (ML) and pleomorphic liposarcoma (PL) [4].

The arising clinical symptoms depend on tumor's location and size. The symptoms may include dysphagia, a feeling of a lump in the throat,

dysphonia, snoring, and airway obstruction [3,5]. The main treatment for hypopharyngeal liposarcoma is wide surgical excision with an endoscopic or transcervical approach [6]. Here we are reporting a rare case of hypopharyngeal liposarcoma of a 50-year-old man who underwent transcervical excision followed by chemoradiation. This work has been reported in line with the SCARE criteria [7].

2. Presentation of case

A 50-year-old male presented with a chief complaint of difficulty swallowing solid food, the continuous presence of a lump in the throat, voice becoming nasally voice for four months. Patient also complained of shortness of breath which got worse one week before presenting along with a lump on his neck since two weeks ago (Fig. 1). He had a previous tracheotomy and hypopharyngeal biopsy. Family history of malignancy, history of radiation therapy and smoking habits were denied by the patient.

On physical examination, there was a neck lump located on the

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Fig. 1. Lump on the neck (red arrow).

anterior colli, level VI, solid, fixed with a size of 3 × 3 × 1 cm, moving when swallowing with the impression of pushing the trachea to the left. Tracheocanule number 7.0 was attached to the neck with good function. The patient had a previous direct laryngoscopy-derived biopsy representing fibrolipoma. The recent indirect laryngoscopy revealed a fibroepithelial polyp.

Pre-operative CT scan findings showed a solid 22 Hounsfield Unit (HU) lesion with a fat component (−53 to −23 HU), well-defined, irregular edges measuring ±5.3 × 3.0 × 3.1 cm in the right supra-glottic dominant. In contrast, there was a slight enhancement (58 HU) in the solid part. The lesion extends from the left carotid space to the paravertebral space until the right carotid space at the level of 4–7 cervical vertebrae. This profile suggests a benign mass that was most likely a fibro lipoma (Fig. 2).

Fiber Optic Laryngoscope (FOL) showed a tumor in the mesopharyngeal area that filled the hypopharynx with a smooth surface, did not bleed easily, hypersalivation, epiglottis, and vocal cords could not be evaluated (Fig. 3).

Based on the clinical, imaging, and histological data, the patient was diagnosed with hypopharyngeal fibro lipoma. The patient underwent transcervical excision of the tumor. During surgery, the tumor appeared to be attached to the right thyroid lobe and was difficult to separate, thus right thyroidectomy was performed. It also infiltrated the lateral pharyngeal wall and enter into hypopharyngeal lumen. The operation lasted for 7 h and the tumor was found to measure 12 × 9 × 3.2 cm, grayish white in color, well-defined like fatty tissue, not easy to bleed (Fig. 4). Upon histological examination, a well-differentiated

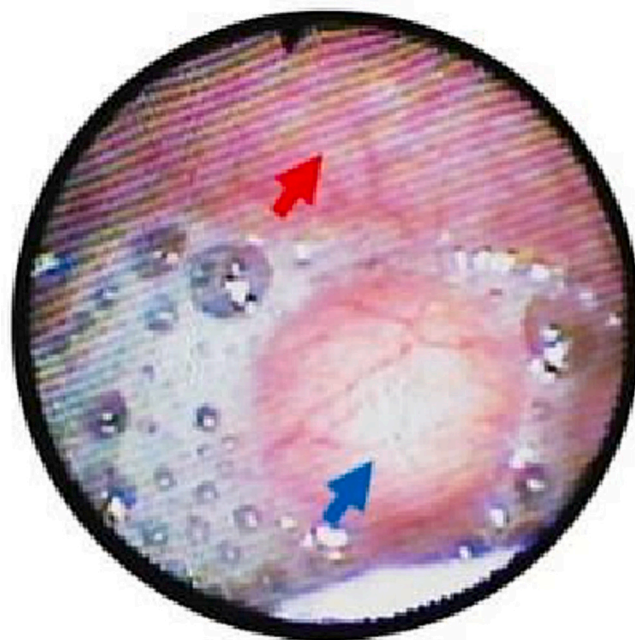


Fig. 3. Fiber optic laryngoscope before surgery. Tumor in the mesopharynx (blue arrow), posterior pharyngeal wall (red arrow).

liposarcoma was discovered.

The patient was discharged ten days after surgery. An adjuvant chemoradiation was added starting one month after the procedure consisting of 6-cycle of chemotherapy followed by radiotherapy 33 times. Post-operative evaluation on the first, seventh, and tenth months revealed no complaints and no tumor recurrence based on physical examination and FOL.

3. Discussion

Liposarcoma of the head and neck was found to be predominance in males with an age range of 20–90 years but at the peak of the fifth decade of life [4,8]. Family history of malignancy and a history of radiation exposure are important factors in the etiology of liposarcoma. The smoking habit is suspected as a predisposing factor to the occurrence of this malignancy [4]. Our patient had difficulty in swallowing, felt stuck in the throat, had a nasal voice, lost weight, and had a shortness of breath. The report demonstrated that the first symptoms were the

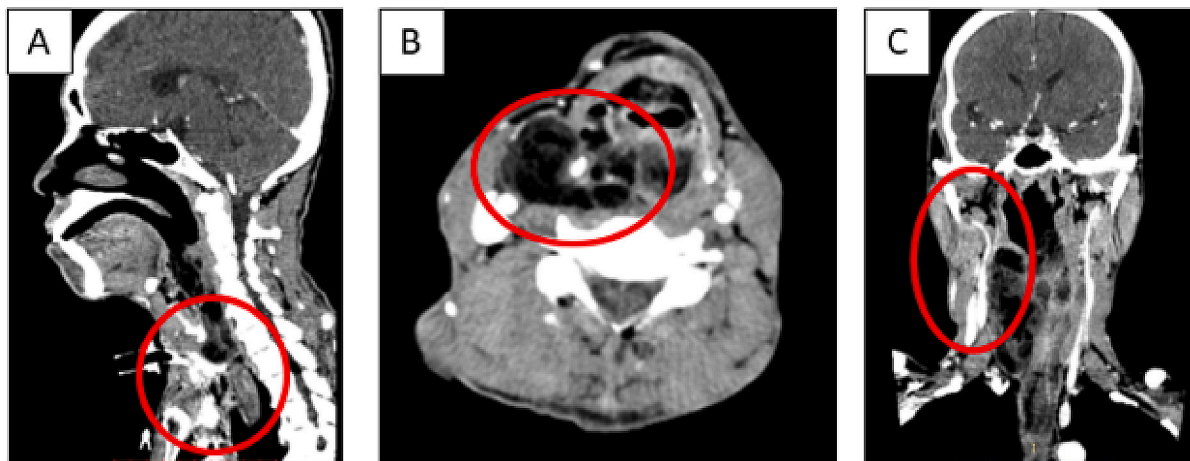


Fig. 2. CT scan of the head and neck. A. Sagittal section: supraglottic lesion (red circle); B. axial section: right-sided dominant lesion, on contrast, slight enhancement of 58 HU in solid part (red circle) is seen; C. coronal section: The lesion encases the right carotid artery (red circle).

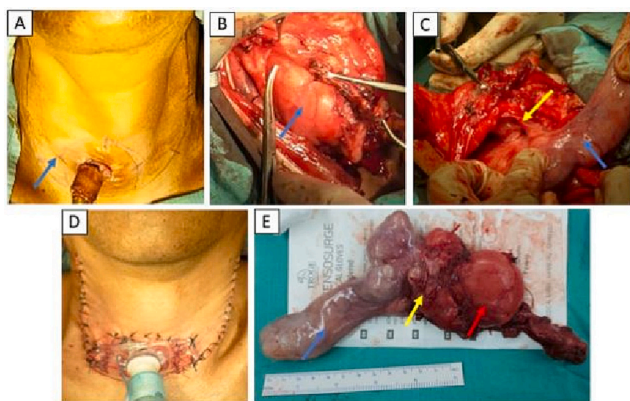


Fig. 4. Excision of the tumor with a trans-cervical approach. A. “U”-shaped incision (blue arrow). B. Tumor in the right thyroid lobe (blue arrow). C. Tumor of the right pharyngeal lateral wall (yellow arrow), tumor (blue arrow). D. Closure of the surgical wound. E. Excised tumor (blue arrow), lateral pharyngeal wall (yellow arrow), right thyroid lobe (red arrow).

progressive worsening of dysphagia, followed by a feeling of a lump in the throat, hoarseness, airway obstruction, and weight loss [9].

The tumor filled the hypopharynx, according to a FOL. The piriform sinus (16 cases), the posterior hypopharyngeal wall (6 cases), and the post-cricoid region are the sites of the genesis of hypopharyngeal liposarcoma (3 cases). Liposarcoma can be seen as polypoid and pedunculated tumors [9]. CT scan displayed a solid lesion with a fatty component, and well-defined irregular borders, suggesting a benign mass, most likely of fibro lipoma. Intratumoral hemorrhage and necrosis, as well as unclear boundaries of the tumor also indicate malignancy [10–12].

The management in this case was a transcervical approach of surgery alongside a right thyroidectomy because the tumor had spread to the right thyroid lobe. Wide surgical excision is the mainstay of the therapy and the approach used is determined by the size of the tumor to be able to visualize the tumor as a whole to facilitate the surgical process. The surgical technique used may be a transoral endoscopic approach or a transcervical excision of the tumor [9,10]. The transoral approach is chosen when the tumor size is <2 cm and the tumor appears pedunculated, however, the excision margin is difficult to evaluate. A transcervical approach is an option for >2 cm in diameter liposarcomas because it provides better and easier access surgical field. As opposed to the lateral pharyngotomy (cervical route), the transoral endoscopic technique is preferred since it is associated with fewer surgical morbidities and complications (such as the injury of the spinal accessory nerve), a shorter hospital stay, and the avoidance of tracheostomies [9,13].

Histopathological findings of tumors in patients showed fibro lipoma on pre-operative examination and liposarcoma on post-operative with a positive margin at the edge of the resection. Soft tissue tumors larger than 5 cm and deep soft tissue tumors have a high risk (about 10 %) of becoming sarcomas [14]. While the histopathology of lipomas and well-differentiated liposarcomas is often difficult to distinguish, the treatment is very different [15]. The histology of liposarcoma found pleomorphism, nuclear atypia, nuclear hyperchromasia, and infiltrative nature of lipoblasts [16].

Furthermore, adjuvant chemoradiotherapy was added following the surgery. It based on the recurrence rate will be lower if the surgical margin is free from the tumor [17]. Around 80 % of cases of incomplete tumor excision were associated with an increased risk of local recurrence. In the case of limb sarcoma, postoperative radiotherapy significantly reduced the local recurrence rate from 60 % to 40 % although does not change the survival rate or the rate of metastasis [18]. A significant increase of 5-year survival rate in patients with head and neck sarcoma who received radiation therapy [19]. The administration of

adjuvant chemotherapy was also associated with a significant reduction in relapse, in both locoregional and systemic [10].

There was a defect in the hypopharyngeal lateral wall along ± 3 cm at the time of tumor removal. Patient was hospitalized for 10 days under close supervision for any complications. Possible complications that can occur include fistulas due to the flow of which generally occurs in patients. Delaying oral feeding can be achieved by providing nutrition through a Nasogastric Tube, which is maintained for 7–10 days [20].

After surgery and chemotherapy/radiotherapy, follow-up visits are made every 3 to 6 months for up to 5 years. On physical examination and Imaging, the post-operative periodic control evaluation up to 2 years later revealed no symptoms or tumor recurrence. Hypopharyngeal liposarcoma had a mean recurrence time of ± 42.28 months. Changes in tumor differentiation should be suspected if there is a recurrent case [10]. Well-differentiated liposarcoma grows slowly and rarely metastasizes with a 1-year survival rate of 94.12 % [9]. Sarcomas usually have a poor prognosis ranging from 45 to 73 % with high recurrence risk for all stages [21].

4. Conclusions

Hypopharyngeal liposarcoma is a rare case. Because of its rarity, resemblance to other tumor types, and lack of aggressiveness, it is easily confused with other hypopharynx masses. Well-differentiated liposarcoma of the upper aerodigestive tract is rare and diagnostically challenging. It is important to differentiate it from a benign lesion before planning treatment because the simple removal of a visible tumor will result in local recurrence. The delicate anatomy of the head and neck limits the possibility of having wide surgical margins, preferably 2 cm or more around the macroscopic tumor, thus the excision surgery approach is determined by the size of tumor. The possible complication of fistula can be reduced by minimizing the stress at the esophageal site. Adjuvant chemoradiation therapy is also considered due to positive margins.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

A copy of written consent is available for the review by the Editor-in-Chief request.

Ethical approval

The study was conducted according to the Declaration of Helsinki and was approved by the ethical review board (approval number: 0954/117/4/IX/2021).

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ACR: Conceptualization, acquisition of data, analysis data, supervision, writing manuscript

CVS: Conceptualization, acquisition of data, analysis data, writing manuscript.

Declaration of competing interest

The authors report no conflicts of interest.

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