

## Recurrent Larynx and Hypopharyngeal Liposarcoma with Systemic Progression

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### Abstract

**Introduction:** Liposarcomas are infrequent tumors. Only 5.6% of these tumors occur in the head and neck. Primary liposarcomas of the larynx comprise ~10% to 15% of head and neck liposarcomas. Multiple surgeries and recurrences history are common.

**Case summary:** We present the case of a 33-year-old male whose illness started at 16 years of age, and who underwent multiple surgeries due to multiple recurrence (six) by a liposarcoma in the neck, all treated with organ preservation. In 2014 patient arrived with a big recurrence a total pharyngolaryngectomy with a jejunal are flap was performed. after 18 month of the follow up the patient comes to the visits and refered developed headache, ataxia and shortness of breath, that is why a MRI of the brain and CT of the torax shown intraxial lesions in the brain and multiple bilateral pulmonary nodules compatible with metastatic disease, We propose palliative treatment with radiotherapy but the patient refuse treatment so he was sent to palliative care.

**Discussion:** The mainstay of treatment is surgical resection. Multiple recurrence is the rule but metastasis to the brain was not reported before. The role of radiotherapy is not well know. Chemotherapy until now showed no sustancial benefit.

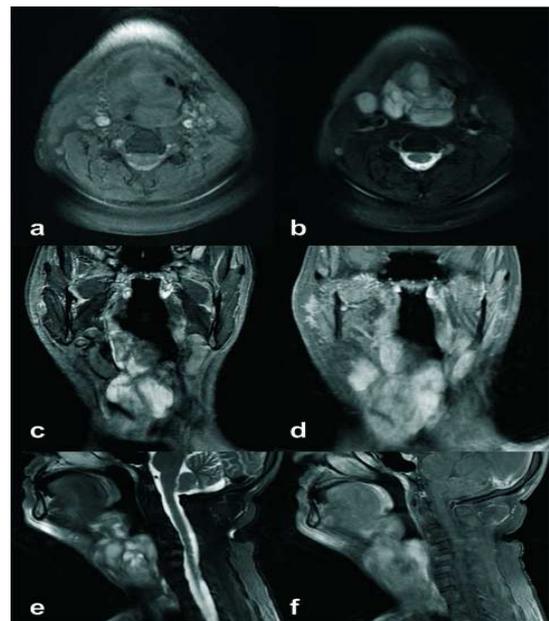
**Keywords:** Head and neck sarcomas; Liposarcoma; Metastasis; Brain; Recurrence

### Introduction

Liposarcomas are malignant mesenchymal tumors that most frequently occur in the extremities and retroperitoneum. Only 5.6% of these tumors occur in the head and neck area. Most of these originate in the soft tissues of the neck [1,2]. The first reports of head and neck liposarcomas were described during the 1960s with liposarcomas of the hypopharynx being extremely rare. Approximately 38 cases have been reported in the English, French and Spanish literature [3,4]. Primary liposarcomas of the larynx comprise ~10% to 15% of head and neck liposarcomas [5]. Approximately 60% of the cases occur in males. Mean age of presentation is between the 4<sup>th</sup> and 6<sup>th</sup> decades of life. Symptom presentation is mainly dysphagia and dyspnea [3,6-8]. We present the case of a young male patient who underwent multiples surgeries and recurrences until being subjected to extensive surgery with jejunal free flap reconstruction and final metastasis disease to the brain.

### Case Report

A 33-year-old male whose illness initiated at 16 years of age with dysphonia and dysphagia due to a tumor originating from the right piriform sinus and treated with two transcervical resections and diagnosis of myxoid liposarcoma. In 2005, the patient was referred to us for recurrence, a transoral surgical resection with CO<sub>2</sub> laser was performed, and surgical margins were negative. A year later, a recurrence and surgery was proposed and refused by the patient. Patient was lost of follow-up for 8-year, during that two additional transcervical resections were performed. In november 2014 he returned to our institution due to a 6<sup>th</sup> new recurrence. He presented with dysphagia to fluids and significant dysphonia with laryngeal stridor and dyspnea on minimal effort. Magnetic resonance imaging (MRI) demonstrated a tumor extending from the piriform sinus and right lateral hypopharyngeal wall to the paraglottic space, with massive infiltration of the larynx with no invasion to tongue base (Figures 1a-1f). A total pharyngolaryngectomy and reconstruction with a jejunal free graft. A tumor of 10 cm × 12 cm with epicenter in the right hypopharynx (Figures 2a-2d). Patient was discharged on the tenth postoperative day with a gastrostomy tube



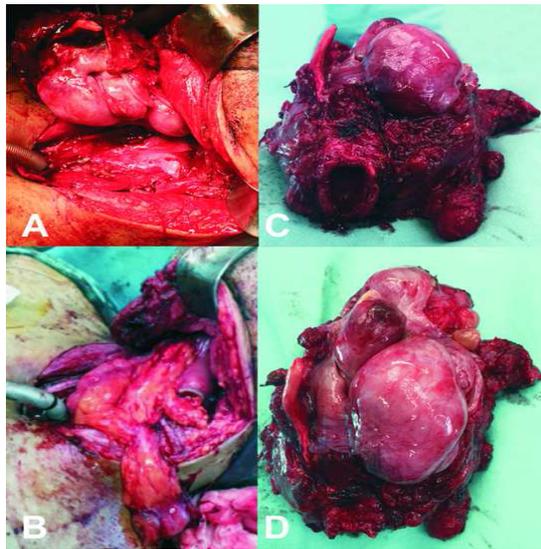
**Figure 1:** Gadolinium-enhanced MRI images which show a hyperintense lesion whose epicenter is located in the right hemilarynx (a,b), with major invasion to the right lateral wall of the hypopharynx (c,d), right paraglottic space and preepiglottic space (d,e and f).

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**Figure 2:** (a,c,d) Surgical specimen demonstrates a right hemilarynx based neoplastic lesion, with evident invasion to paraglottic space and right wall of the hypopharynx and minor invasion to preepiglottic space. (b). Inset of microvascular jejunal free flap to be anastomosed to remaining portion hypopharynx and cervical esophagus.

feeding and no complications. A myxoid liposarcoma with round cells component (20%) and lymphovascular invasion and margins were negative. Adjuvant radiotherapy was proposed but patient refuse. At 18 month of the follow up the patient refered headache, ataxia and shortness of breath, a MRI of the brain and CT of the torax shown intraxial lesions to the brain and multiple metastasis to the lung. We propose palliative treatment with radiotherapy but the patient refuse any treatment so he was sent to palliative care.

## Discussion

Liposarcomas of the hypopharynx are rare tumors accounting for only 3% to 8% of liposarcomas presenting in the head and neck. These generally occur in the soft tissues of the neck [9]. There have been ~30 cases of this type of tumor in the hypopharynx with a male: female ratio of 9:1 [4]. Risk factors for developing this type of tumor are unknown. Gerry et al. [9] report that the combined incidence in the larynx and hypopharynx was 6%. This study also reveals that the biologic behavior of head and neck liposarcomas is more favorable than those of the trunk. There are up to seven different histological subtypes of this malignancy reported in the literature. Of these, myxoid liposarcoma continues to be the most common in the head and neck region with ~79% of the cases. Our patient had mostly a myxoid component; however, areas of rounded cells were present in 20% of the fields studied in the surgical specimen [9].

The most common clinical presentation is associated with obstructive symptoms of the aerodigestive tract such as dysphagia

and dysphonia. Lymph node involvement at the time of diagnosis is infrequent.

There is no standardized surgical management for these tumors due to their infrequent presentation, but surgery is currently the cornerstone of treatment without real evidence on the role of radiotherapy. This is demonstrated in the study by Gerry et al. [9] where 79.6% of the patients were treated exclusively with surgery for head and neck tumors. The anatomic location makes it very difficult to perform surgery with wide margins in comparison with other locations such as the trunk or extremities due to the closeness of neural and vascular structures which, if resected, are associated with significant morbidity. These tumors are associated with a high rate of recurrence especially when organ preservation is attempted. One of the main prognostic factors in the treatment of sarcomas is the status of the resection margins. In the literature, surgical management varies in different case reports from management using marginal resection, transoral laser surgery, and wide resections [1,3,5,7,8,10]. All series were in agreement and the case reports indicate a high rate of local recurrence with locally aggressive behavior and, rarely, distant disease, there is not report in the literature with metastases to the brain [5,8]. There is not any randomized control trial in the literature who shows sustancial benefit in the addition of adyuvant chemotherapy.

## Conclusion

The mainstay of treatment is surgical resection. Multiple recurrence is the rule but metastasis to the brain was not reported before. The role of radiotherapy is not well known.

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