Laryngeal Leiomyosarcoma: A Case Report And Review Of Literature

J Nair, R Atri, P Kaur, S Kumar, V Kaushal

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Abstract

Laryngeal leiomyosarcomas are extremely rare malignancies that arise from smooth muscle cells. The extend of its rarity is supported by the fact that only less than 50 cases of pure laryngeal leiomyosarcomas and less than 10 cases of hypopharyngeal leiomyosarcomas have been reported in modern medical literature.1, 2 Even though the clinical presentation mimic that of a laryngeal carcinoma; which form the major bulk of the laryngeal malignancies; the differences in management warrants an accurate diagnosis. An accurate histological diagnosis may be difficult; but when supplemented by electron microscopy and immunohistochemical tests, the diagnosis can be reached with certainty. We report a case of this very rare malignancy presenting in the supraglottic region with an even rare feature of hypopharyngeal extension. An extensive review of literature has been done highlighting the clinical features, histological and radiological diagnosis and management of this extremely rare malignant entity.

INTRODUCTION

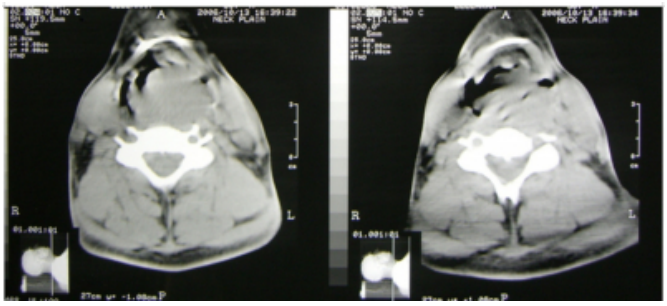
Malignant mesodermal neoplasms of the larynx are very rare; accounting for less than 1% of all malignant laryngeal tumors.[3] Leiomyosarcomas of head and neck constitute 3% of all leiomyosarcomas.[4] Of these head and neck leiomyosarcomas; only less than 50 cases have been reported originating from laryngeal region and less than 10 cases from the hypopharyngeal region. This extremely rare malignancy which mimics a laryngeal carcinoma in its clinical presentation and imaging, demands accurate diagnosis using immunohistochemical modalities because of the differences in the management. The rarity of this malignancy has lead to uncertainty regarding the accurate line of management and prognostic information. We present an extremely rare malignancy of supraglottic laryngeal leiomyosarcoma with hypopharyngeal extension with review of modern medical literature highlighting its clinical features, diagnosis and management.

CASE REPORT

A 40 year old male presented with alteration of voice for past 3 months, which was associated with pain on left side of neck of same duration, which later radiated to left ear. The patient developed respiratory stridor for which emergency tracheostomy was done. Direct laryngoscopy showed a proliferative growth in the supra glottic region more towards the left side, extending to the left piriform sinus and left arytenoids . The vocal cords could not be made out due to the overlying growth. There was slough covering the growth. A para-laryngeal lymphnode was palpable of size 1X 1 cm, firm in consistency and mobile. The biopsy of the tumor suggested a malignant mesenchymal tumor. An X-ray of the soft tissue neck showed an irregular soft tissue opacity arising from the prevertebral soft tissues anterior to C4 and C5 vertebral bodies in the region of the thyroid and cricoid cartilages. The opacity was projecting into the airway and causing significant obstruction. A CT scan [LEGEND 1] of the patient showed mass in the supra glottic larynx with involvement of glottis and pyriform fossa on the left side.

Figure 1

Legend 1: CT scan showing growth larynx with hypopharyngeal extension.



The patient subsequently was subjected to

laryngopharyngectomy with left radical neck dissection. The histopathology of the operated specimen (LEGEND 2 & 3) proved to be Leiomyosarcoma.

Figure 2

Legend 2: Spindle shaped cells having hyperchromatic nuclei and eosinophilic cytoplasm with numerous mitotic figures. H&E stain (200 X)

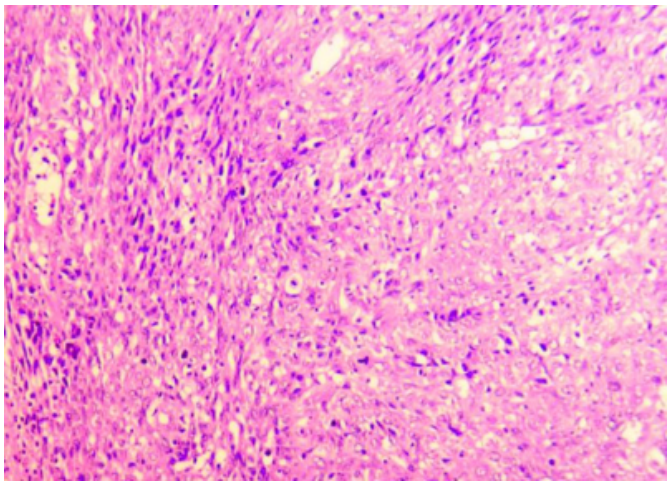
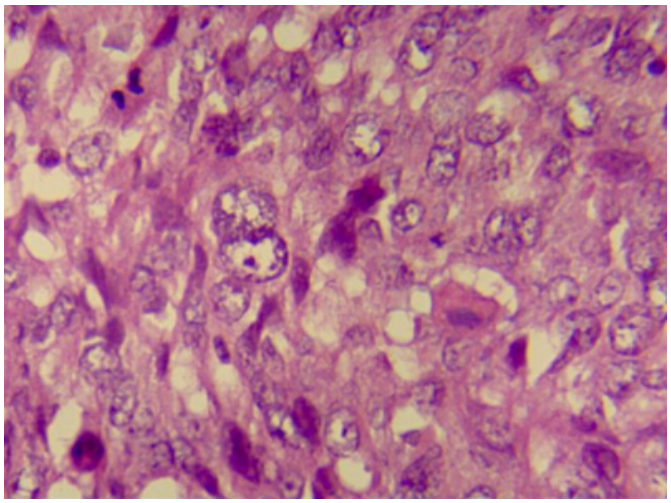


Figure 3

Legend 3: Highly pleomorphic cells with mitotic figures. H&E stain. (400 X)



The immunohistochemistry of the specimen was positive for alpha smooth muscle actin, desmin and vimentin and negative for S-100 and pancytokeratin; consistent with the diagnosis of leiomyosarcoma larynx. The patient was then investigated to rule out the possibility of dissemination; which all were negative. Post operatively, the patient was treated by adjuvant radiotherapy with 60 Gy delivered in 30 Fractions; and was put on follow-up. He was on close follow up for the past 4 months. Patient is clinically and radiologically disease free till now.

DISCUSSION

Malignancies of the larynx accounts for 1-5% of malignancies diagnosed annually. Majority of these tumors are squamous cell carcinomas. Mesenchymal tissue malignancies or sarcomas form a very insignificant proportion of laryngeal malignancies; accounting roughly 1%.[4] Leiomyosarcoma of the larynx was first reported by Jackson and Jackson in 1939.[5]

Leiomyosarcomas originate from the smooth muscle tissues. Their sites of common occurrence is in regions where smooth muscles are in plenty; namely uterus, the gastrointestinal tract and the retroperitoneum. Head and neck leiomyosarcomas account only for 3% of all leiomyosarcomas.[6]The low incidence in the head and neck region is explained as a result of smooth muscle scarcity in the head and neck, which is limited to erector pili muscles, vessel walls and the esophagus. The common sites of head and neck leiomyosarcomas are paranasal sinuses, scalp and cervical esophagus.[4]

Majority of the cases of leiomyosarcomas of the larynx originate in the supraglottic or glottic area.[7] Marioni etal in 2000; in his review of 31 cases, reported the incidences in laryngeal subsites. The authors noted that the incidence each sites was: glottis (48%), supraglottis (32%), supraglottis-glottis (6.5%), subglottis (6.5%), supraglottis-glottis-subglottis (3.5%) and glottis-subglottis (3.5%).[8] In our patient the tumor also showed extension to the hypopharynx (piriform sinus), which is an even rarer clinical feature. Extensive review of modern literature showed that there were only less than 10 cases of hypopharyngeal leiomyosarcoma reported; adding to the rarity of the present case.[2]

The etiological factors which play in the genesis of this rare malignancy are still unclear. Unlike the squamous cell carcinomas of the larynx which have smoking and alcohol as an important etiological component, their role in leiomyosarcoma larynx is still uncertain. An aberrant differentiation of mesenchyma is postulated as a factor, suggested by the factor that a leiomyosarcoma can occur in an area where smooth muscles are in rarity, namely the head and neck, which can be a possibility in our patient. [9] Another argument which is stated in medical literature as a etiological factor is a post operative healing process deviating from its normalcy; which might not be the cause here; as the patient doesn't have a history of any surgical interventions in the past.[10] There have been reports of

Ebstein-Barr virus genome detected associated with this rare malignancy; especially in immunosuppressed patients.[11]

The diagnosis of a leiomyosarcoma larynx is difficult to appreciate clinically as they are clinically indistinguishable from a laryngeal carcinoma. Hoarseness is a common complaint. But this non-specific complaint occurs both in benign as well as malignant laryngeal growths.[4] Laryngeal leiomyosarcoma presenting with obstructive features have been reported in medical literature. Tewary et al had reported a case which required an emergency laryngectomy for airway preservation.[12] Also reports of patients managed with emergency tracheostomy.[13] Our patient had obstructive complaints which were managed by an emergency tracheostomy.

A symptom which has not been reported in previous literature; but was a significant complaint in our patient was referred otalgia. These symptoms might be due to the extension into the hypopharynx, or due to the activation of internal branch of superior laryngeal branch of tenth cranial nerve with referral to auditory nerve of Arnold, causing referred otalgia. Even though this symptom is common with laryngeal and hypopharyngeal carcinomas; our search of medical literature did not reveal any mention about this relevant symptom associated with laryngeal leiomyosarcomas.

Head and neck leiomyosarcomas rarely present with lymphnode metastasis. In our case, the small lymphnode was palpable which again is rare.[14]

The role of conventional microscopy is limited in making a morphological diagnosis of laryngeal leiomyosarcoma. When the findings are supplemented by immunohistochemistry, and ultrastructural investigations, the diagnosis of this rare tumor is made with high degree of certainty. Histopathologically, laryngeal leiomyosarcoma is characterized by prominent interlacing bundles and fascicular arrangement of spindle-shaped cells with cigar-shaped, blunt-ended nuclei and eosinophilic cytoplasm. Our patient's histopathology also had a similar picture confirming the diagnosis of a spindle cell carcinoma of the laryngo-pharynx.

Other spindle cell tumors include rhabdomyosarcoma, melanoma, schwanomma, malignant fibrous histiocytoma and sarcomatoid carcinoma. The advent of newer monoclonal antibody techniques, aiding an immunohistochemical diagnosis has made the diagnosis easier. Leiomyosarcomas are usually positive for alpha smooth muscle actin and negative for cytokeratins and epithelial membrane antigens. In our case, the tumor was positive for alpha smooth muscle actin, desmin and vimentin and negative for pancytokeratin and S-100 antigens.

Marioni et al in 2005 stressed the diagnostic role played by immunohistochemistry in the diagnosis of this rare malignancy. The authors stressed that, even though this tests allow a reliable diagnosis, it can produce ambiguous or inconclusive results when the tumor cells lack specific immunohistochemical reactivity. In this rare scenario, the authors suggest the diagnosis to be substantiated by electron microscopy; because in addition to academic interest; any wrong assumption might lead to inappropriate clinical management.[15]

Radiological investigations like CT and magnetic resonance imaging play a critical role in determining extent of the primary in planning surgery as well as to deliver information regarding the lymph node status.

The rarity of this tumor has resulted in the lack of any conclusions regarding the standard modality of management. Abbas et al in 2005 in their review suggested that the first line of management be resection of the whole tumor with wide surgical margins with curative intent. In certain cases, endolaryngeal resection or partial laryngectomy has been tried, to preserve functionality in early disease status. Radical neck dissection is said to be reserved in case with obvious cervical lymphadenopathy. The rate of local recurrence following surgical excision range from 35-50%.[3] Our patient was subjected to laryngo-pharyngectomy and left radical neck dissection for complete removal of the disease with a curative intent.

The role of radiotherapy is in being an adjuvant therapeutic modality. Radiation can also have a role in recurrence or in residual disease. But as a primary treatment modality, radiotherapy is considered less effective.[16] Our patient was given adjuvant external beam radiotherapy the region by parallel opposed fields. Chemotherapy has also a limited role in leiomyosarcomas of the larynx.

The scarcity of cases reported has caused the indecisiveness in chalking out the prognosis of this rare tumor. Literature search has shown that head and neck leiomyosarcomas generally have a survival of 35-50%.[4] Medical literature review showed that the longest period of postoperative disease survival is 11 years. A long period of follow up is

mandatory as the risk of recurrence persist long time after treatment. [3]

With the advent of new immunohistochemical techniques; the diagnosis of leiomyosarcomas of the larynx can be picked out from the big list of differential diagnosis of spindle cell tumors of the larynx. Such modalities should be liberally used in the diagnosis when ever suspected due to the wide difference in the treatment methods, giving the patient a better cure and quality of life.

CORRESPONDENCE TO

Dr. Vimoj J. Nair, Junior Resident, Department Of Radiation Oncology, PGIMS, Rohtak-124001. EMAIL-vim4u4ever@hotmail.com Phone-09199920255 19

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Author Information

J. Vimoj Nair, MBBS
Junior Resident, Department Of Radiation Oncology, Regional Cancer Centre

Rajeev Atri, MD
Senior Resident, Department Of Radiation Oncology, Regional Cancer Centre

Paramjeet Kaur, MD
Associate Professor, Department Of Radiation Oncology, Regional Cancer Centre

Sanjay Kumar, MD
Assistant Professor, Department Of Pathology, Post Graduate Insitute Of Medical Sciences [PGIMS]

Vivek Kaushal, MD, DNB
Head & Senior Professor, Department Of Radiation Oncology, Regional Cancer Centre