Primary Angiosarcoma of the Larynx

GOPAL KRISHNA MAHESHWARI1, HARSHAD ACHARATLAL BABOO1, MAHESH HIRJIBHAI PATEL2, USHA GOPAL3, KUMAR WADHWA4

1The Gujarat Cancer and Research Institute, Departments of 1Radiation Oncology, 2Surgical Oncology, 3Radio-Diagnosis and 4Pathology, Ahmedabad-India

ABSTRACT

Sarcomas constitute fewer than 1% of the head and neck cancers. They represent less than 1% of laryngeal cancers. Primary angiosarcoma of the larynx is an extremely rare malignancy. The available literature on this medical oddity is in the form of isolated case reports only. The purpose of this article is to add another case of primary angiosarcoma of a rare site, the larynx, of which only 21 cases have so far been reported in the literature. The present patient, a 28–year–old woman is the first case being reported from India among all reported cases of angiosarcoma of the larynx. [Turk J Cancer 2004;34(4):166-168]

KEY WORDS:
Angiosarcoma, larynx

CASE REPORT

A previously healthy 28-year-old woman presented with 1-month history of pain during swallowing, dyspnoea, cough, hoarseness of voice. She also complained of recurrent hemoptysis for last 10 days. Her general physical examination findings were unremarkable. There was no cervical lymphadenopathy. Examination of oral cavity and oropharynx was also normal. Indirect laryngoscopic examination revealed an exophytic mass in the left supraglottic hemilarynx. Further evaluation was not possible. Chest X-ray was normal. Except anemia (Hb=8.2 gm%), her hematological and biochemical investigations were within normal limits.

CT scan of the neck (Figure 1) demonstrated an extensive soft tissue mass involving left side of epiglottis and extending down to both false and true vocal cords on left side and also crossing the midline. Evidence of spread in to paraglottic or parapharyngeal space was also noted. The mass showed heterogeneous post-contrast enhancement. No neck node enlargement was noted on CT scan. Tracheostomy was performed. Direct laryngoscopic examination under anesthesia and biopsy of the lesion was done. Histopathological examination (Figure 2 A,B) of the endoscopic biopsy material revealed squamous epithelium with intact basement membrane. The tumor was mainly submucosal and sheets of spindle shaped tumor cells invaded submucosal connective tissue. Presence of vascular proliferation without lobules with central mitosis, vesicular and pleomorphic nuclei and cytoplasmic clear spaces consistent with diagnosis of angiosarcoma (grade I) was noted. Immunohistochemical
staining of the tumor cells resulted in positive staining with desmin, vimentin and factor VIII antigens.

The patient was started on hemostatic radiotherapy. A total dose of 16 Gy/1 week/4 fractions by two parallel opposed lateral portals on 6 Mv Linear Accelerator was delivered and total laryngectomy was performed. The patient had a smooth post-operative period with oral feeding started from 12th post-operative day and was discharged from the hospital on 18th post operative day. The histopathological examination showed presence of 3.2 cm size polypoid growth arising from laryngeal surface of the epiglottis, aryepiglottic fold, false vocal cord and true vocal cord on left side. A margin of healthy tissue was seen all around the tumor.

The patient currently remains asymptomatic without clinical or radiological evidence of any loco-regional recurrence or distant metastases 40 months after her disease was diagnosed.

DISCUSSION

Angiosarcoma is a malignant tumor of vascular endothelial cell origin. It is malignant counterpart of the hemangioma and one of the rarest among the various soft tissue sarcomas, comprising less than 1% of all sarcomas (1). It is also known as malignant hemangioendothelioma, angioblastoma, hemangiosarcoma, and intravascular endothelioma. The exact etiology of angiosarcoma is unknown. Trauma, radiation and anectesia have been linked with its etiology (2). It may be multifocal in origin. Although it can be found throughout the human body and affects various organs and structures, the extremities are the most commonly involved sites. Angiosarcoma occurs most commonly in the skin and superficial soft tissues. This phenomenon contrasts sharply with deep location of most soft tissue sarcomas. Angiosarcoma of the head and neck is very rare and when it does occur in this region, the scalp and facial skin are the most common sites of origin.

Sarcomas are rarely found in the larynx and represent less than 1% of malignant tumors in this location. More than 50% of laryngeal sarcomas are fibrosarcoma (3). Among sarcomas arising in the larynx, angiosarcoma is one of the rarest accounting for vanishing small proportion of all laryngeal tumors. The first case of an angiosarcoma of the larynx was reported in 1924 by Yankauer (4). A review by McRae et al. (5) showed that till 1990, only 20 cases including a case of their own have been reported in this region. A search of the literature enabled us to trace an additional case reported by Sciot et al. (6) in 1995, making a total of 21 cases (7-10). The present case is the first patient of laryngeal angiosarcoma being reported from
Angiosarcoma of Larynx

India. Because of rarity of this tumor, it practically never enters into the clinical differential diagnosis of malignant lesions in this location.

Angiosarcoma of the larynx has been reported equally in male and female. More often this tumor has been seen in the middle aged patients. It can affect any site in the larynx. Dysphagia, bleeding, change of voice, dyspnoea, stridor are common manifestations of lesion in this location. In the present case, the lesion was extensive and involved supraglottic and glottic regions. Two special techniques useful in differentiating angiosarcoma from other lesions are immunohistochemical staining for factor VIII and the reticulin stain. The former is of value in confirming the endothelial origin of the tumor whereas latter demonstrates that the malignant cells are located on the luminal side of the vessels.

Angiosarcoma usually spreads by hematogenous route and pulmonary metastases are the most common site of distant spread. Lymphatic spread is reported in less than 20% cases of angiosarcoma of head and neck region (2). In the present case, no lymph node/ distant metastases were observed at initial presentation or during follow-up period of 40 months.

Surgery remains the mainstay of the treatment. A combined modality of treatment has been used, where radiotherapy is used as adjuvant to surgery (pre or post-operatively). Laryngeal angiosarcoma is a wildly aggressive neoplasm that results in frequent local recurrences, early metastases and a low 5 year survival rate. However, few cases with long term disease free survival have been reported (10,11). Our patient was treated with hemostatic radiotherapy and surgery. She remains in good health 40-months after the initial presentation. We recommend that all such cases of angiosarcoma of the larynx should be reported in order to know their biological behavior and ultimate prognosis.

References