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Undifferentiated Pleomorphic Sarcoma of Larynx: A Rare Entity with Review of Literature

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Abstract
Sarcomas of the larynx are rare neoplasms that constitute less than 1% of laryngeal malignancies. We describe a rare case of undifferentiated pleomorphic sarcoma of larynx of a 62-year- old male who presented with hoarseness of voice. Laryngoscope showed growth over glottic region of larynx and biopsy revealed poorly differentiated carcinoma. Patient was treated with radical radiotherapy (RT). After 3 months of completion of RT, laryngoscopy and CECT revealed residual lesion. Cordectomy was done. Post- cordectomy histopathology report (HPR) and review of pre RT biopsy specimen showed tumour composed of highly pleomorphic spindle cells with moderate eosinophilic cytoplasm, vesicular nuclei with prominent eosinophilic nucleoli, frequent mitoses and occasional multinucleate cells. IHC showed tumour cells were positive for vimentin and focal positive for smooth muscle actin but were negative for Pan-CK, CK- 5/6, S-100, CD-34 and desmin. The overall features were consistent with undifferentiated pleomorphic sarcoma. Post-cordectomy PET-CT showed metastatic disease. Patient was further managed with 6 cycles of Ifosfamide and Epirubicin based palliative chemotherapy (CT) and after 6 months of CT completion, patient is symptomatically better. The characteristic clinical, histopathological features and management of this rare case are described with a literature review.

Keywords : *Larynx, undifferentiated pleomorphic sarcoma, metastases*

Introduction

Sarcomas are derived from the mesodermal tissue with a diversity of clinical behaviours due to various types of pathologic classifications and represent 1% of all head and neck malignancies [(1)]. Sarcomas of the larynx are rare neoplasms that constitute less than 1% of laryngeal malignancies[(2)]. Undifferentiated pleomorphic sarcoma has an uncertain histogenesis and it is mainly found in the limbs, abdomen and retroperitoneum[(3,4)]. Since its occurrence in larynx is rare, so limited evidence is available regarding its literature.

Case Report

A 62- year- old male, presented with 6 months history of hoarseness of voice. Patient was investigated and diagnosed outside to our institution as carcinoma Larynx of glottic region based on Laryngeal biopsy findings which showed poorly differentiated carcinoma. Patient was treated there with EBRT dose of70Gy in 35#. After 3 months of completion of EBRT, laryngoscopy showed residual growth (Figure1) and CECT revealed irregular soft tissue thickening in the region of anterior commissure. Type Va extended cordectomy was done. Post – operative HPR showed sarcoma. Patient was then referred to our institution. Patient's pre- RT histopathology specimen, post-cordectomy histopathology specimen were reviewed and IHC was done in our institution which showed tumour composed of highly pleomorphic spindle cells disposed in sheets and intersecting fascicles. The tumour cells were oval to spindle with moderate eosinophilic cytoplasm and elongated to oval vesicular nuclei with prominent eosinophilic nucleoli (Figure 2). Frequent mitoses (8-10/HPF) and occasional multinucleate cells were also noted. The tumour cells showed positivity for vimentin (Figure 3) and focal positivity for smooth muscle actin but were negative for Pan-CK, CK- 5/6, S-100, CD-34 and desmin (Figure 4).So overall features were consistent with undifferentiated pleomorphic sarcoma. Post cordectomy direct laryngoscopy showed growth over anterior commissure and PET-CT showed disease in anterior commisure with FDG- avid right upper paratracheal and meditational lymph nodes along-with multiple left lung metastases. Patient was further managed with 6 cycles of Ifosfamide and Epirubicin based CT which patient tolerated well and after 6 months of follow-up patient is symptomatically better.

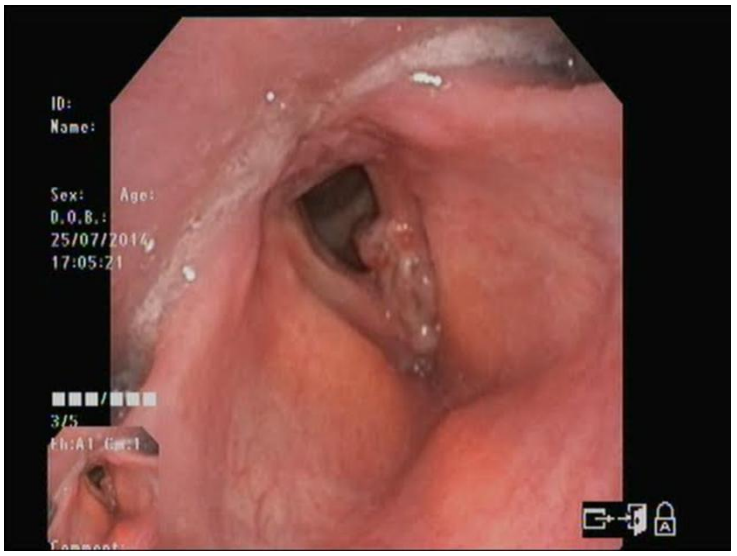

Figure 1. Post- RT laryngoscopy showing residual growth over anterior commissu

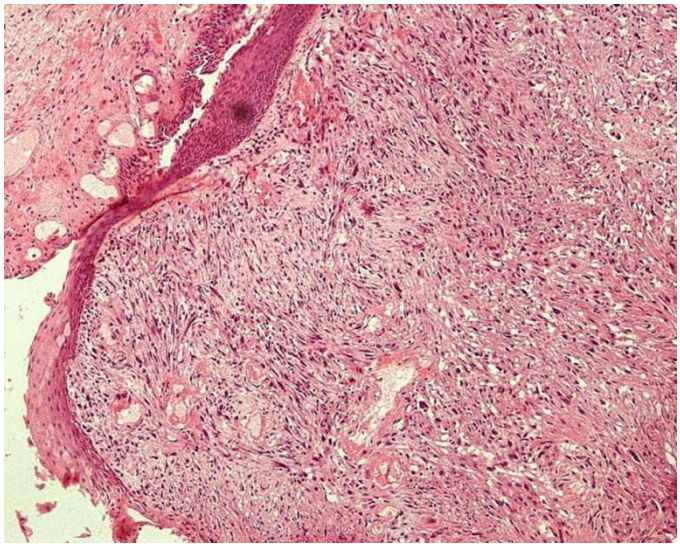

Figure 2. Section showing focal stratified squamous lining with underlying pleomorphic spindle cell proliferation and atypical mitoses 100X H&E.jpg

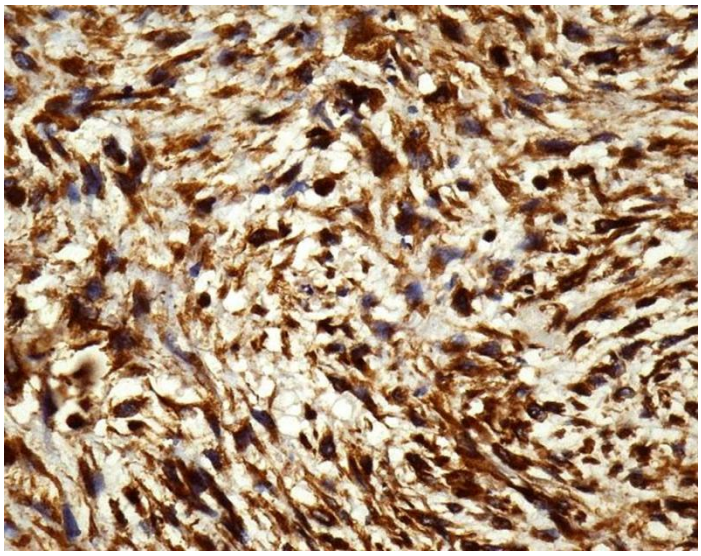

Figure 3. Pleomorphic spindle cells showing vimentin positivity 400X IHC.jpg

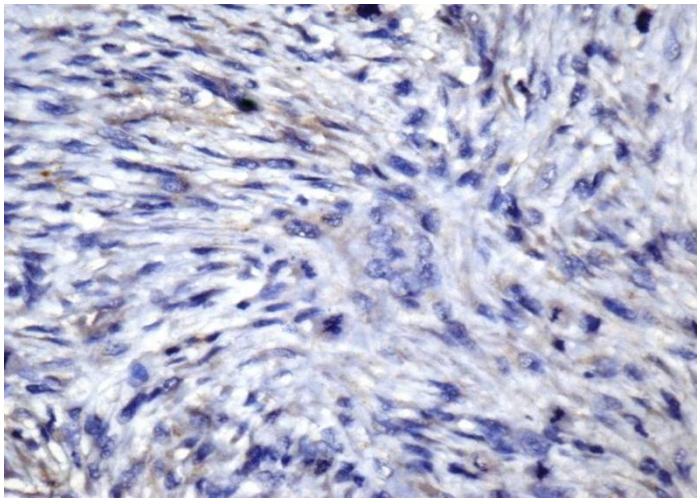

Figure 4. Pleomorphic spindle cells showing pan- CK negativity 400X IHC.jpg

Discussion

Approximately 3 to 10% of all undifferentiated pleomorphic sarcomas (also known as Malignant Fibrous Histiocytoma, MFH) are localized in the head and neck and the most common sites are naso-sinusal cavities (30%), cranio-facial bones (15–25%) and the larynx (10–15%), [5,6] .Less than 50 cases of laryngeal pleomorphic sarcoma have been published in literature till date [7]. Laryngeal pleomorphic sarcoma is an aggressive tumour associated with high rates of recurrence and distant metastases. It is more commonly found in elderly male. Frequently, the primary localization of these tumours in male isglottis as in our case, whereas in women it is the subglottis [5].

Dysphonia is the most common presenting symptom as reported by our patient [7]. Differential diagnosis includes:

Pleomorphic rhabdomyosarcoma, fibrosarcoma, spinocellulacarcinoma, angiosarcoma, hemangio-pericytoma, pleomorphic liposarcoma and lymphoma. It is derived from mesenchymal tissue composed by 5 cell types: fibroblast, histiocytic, indistinguished, giant multinucleated and xantomatous cells. Its origin is believed to be from tot potent cells for presenting two different cell types. Structural analysis through electronic microscopy, IHC and tissue culture are useful in making diagnosis [8]. In our case when patient came to us, there was diagnostic dialemma because patient was treated by EBRT for carcinoma but post- cordectomy HPR showed sarcoma. We were able to make the correct diagnosis only after reviewing the two specimens along with IHC. So here we suggest that in any case of poorly differentiated carcinoma of larynx, HPR review along with IHC should be done to exclude sarcoma.

Surgery is primary modality of treatment [9] and radical resection is usually recommended. Postoperative adjuvant RT is considered for high-grade tumours, positive surgical margins, larger tumour (> 5 cm) and recurrent lesions[10]. In our case persistence of disease after EBRT might be because initially surgery was not done since patient was treated in line of carcinoma. Chemotherapy has important role in metastatic disease, primarily Ifosfamide and Doxorubicin based combination regimens. Lung is most common site of distant metastasis like in our case. Because of its rarity, no convincing dataabout survival is known so far.

Conclusion

Undifferentiated pleomorphic sarcoma of larynx is a rare clinical entity with an aggressive clinical course. For poorly differentiated carcinomas we suggest histopathology review along with IHC. Surgery is the treatment of choice and post-operative radiotherapy is required for high grade,

positive margins, tumour size more than 5 cm and recurrent lesions. Chemotherapy has important role in metastatic disease. More case reports and series are needed to predict the outcome.

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