SYNOVIAL SARCOMA OF LARYNX- A RARE SITE

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Abstract : Synovial sarcoma is a soft tissue sarcoma of unknown histiogenesis and occurs predominantly in the lower limbs of young adults and the head and neck is a relatively rare site, there are about 10 cases with laryngeal localization in the world literature. We present a 52 year old male with synovial sarcoma of larynx. Total laryngectomy was done and patient is free from disease till date.

Keywords: Synovial Sarcoma, Head and Neck, Larynx

INTRODUCTION

Sarcomas of head and neck are extremely rare accounting for less than 1% of all neoplasms of head and neck. These sarcomas arise in both soft tissue and bone and thus cannot be treated by a single approach. The clinical behavior of these tumors varies considerably and patients with low grade lesions are prone to local recurrences, whereas those with high grade lesions develop both local and disseminated disease. Diagnosis and treatment of sarcomas of head and neck present a considerable challenge. Another significant prognostic indicator of survival for sarcomas is the size of the neoplasm. Lymphnode metastasis is rare; when it occurs it is almost exclusively associated with high grade tumors. Synovial sarcoma is a malignant mesenchymal tumor accounting for approximately 10% of all soft tissue sarcomas. Young adults and adolescents are affected, with male to female ratio of 1.2:1. The neoplasm often originates in proximity to major joints and bursa, particularly in lower limbs, while other locations, such as the retroperitoneum, pleura, lung, thorax and mediastinum are unusual sites.

Synovial sarcoma of head and neck is very rare and accounts for only 3% of all sarcomas. The first case was reported by Jernstorm in 1954 and since then only about 85 cases have been reported in world literature. Two different types have been described, referred to as Biphasic type composed of both epithelial cells and spindle cells and Monophasic type where a single cellular component is dominant. The case reported is of a Biphasic synovial sarcoma of larynx and clinical and histopathological features of this rare neoplasm are discussed stressing the importance of immunohistochemistry in confirming this type of diagnosis.

CASE REPORT:

A 52 year old male patient came to our hospital in October 2004, with swelling in the neck, and hoarseness of voice. The patient also complained about foreign body sensation in throat associated with mild stridor. He had no persistent cough or pain in the throat and apparently no other complaints. The patient was non-hypertensive, non-smoker and non-alcoholic. He doesn't have any history of bleeding diathesis or trauma to the throat. But in treatment history, in 1992, he developed a swelling in the neck, in the thyroid region with hoarseness of voice; clinical diagnosis of carcinoma thyroid was made with FNAC revealing adenocarcinoma of Thyroid. Patient underwent total thyroidectomy. The details of the surgery and histopathology were not available with the patient.

Patient was asymptomatic till May, 2000, when he developed recurrence of the swelling. Excision biopsy of the mass was done, which revealed synovial sarcoma. Patient was advised surgery but he refused to undergo laryngectomy, therefore radiotherapy was given.

On 3rd October, 2004 he presented with stridor. As the patient refused tracheotomy, he was managed conservatively and Thallium-201 and SPECT scan was done with a probable diagnosis of Thyroid gland mass or mass in the larynx. On 11th October, 2004 CT scan was done, when he came with the presenting complaints which revealed iso or hypodense mass lesion with calcification in subglottic region of the larynx. There was no cervical lymph node involvement. The radiologist opined the condition as mass larynx with post RT status, probably synovial sarcoma in view of the previous history.

CT findings of the case

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Fig : The operated Mass

Patient was convinced and counseled for total laryngectomy and surgery was done and mass sent for review histopathological examination and immunohistochemistry. The HPE showed synovial sarcoma-biphasic with both spindle and epithelial cells. Immunohistochemistry

revealed spindle cells positive for vimentin and epithelial cells positive for cyto-keratin and there was also weak positivity with S-100. All the above features are consistent with biphasic synovial sarcoma. It was not considered necessary in this case to carry out electron microscopy and cytogenetic studies since these assessments are costly and not available in our set-up, moreover, they are used in uncertain cases.

The patient was disease free and went to his native place. He was on regular follow-up and voice rehabilitation with Blom-singer's indwelling prosthesis was done one year back. He is free from disease till date.

DISCUSSION AND CONCLUSION:

Synovial cell sarcoma is generally considered as a malignant neoplasm with biologically an aggressive behavior and with high probability of recurrence. The tumor manifests as an indolent growing mass with the sites of onset in decreasing order of frequency - hypopharynx, neck, face, larynx, rhino & retropharynx³, it usually occurs as a well circumscribed mass covered by a thin fibrotic capsule making it easy to enucleate from the surrounding structures. This however is a pseudo capsule, beyond which the neoplasm usually tend to infiltrate.

CT and MRI are non-specific in diagnosis, but are useful in planning the surgical strategy. In 30-60% of synovial sarcoma cases, calcifications have been found at CT examination. And these when present are to be considered a positive prognostic factor⁴ (as seen in this case). The factors with worst prognosis include: age more than 25 years, tumor size more than 5 cm. and low grade of differentiation.

The probability of local recurrence has been reported to range between 30-40%, with 50% probability of distant metastasis. Onset of recurrences and distant metastasis may manifest even after a prolonged period (as seen in this case). Therefore regular long term follow-up is essential. The most frequent sites of metastasis is the lung (49%) followed by skeleton (24%), liver (14%) and brain (11%)³. Survival at 5 years ranges from 23.5% & 45% and between 11.2% and 30% at 10 years.

Synovial sarcoma classically shows biphasic cellular pattern comprising two cellular components, spindle and epithelial cells. Monophasic variants are also described.⁵ Use of immunohistochemistry in these cases is very important diagnostic tool. Positivity for vimentin in mesenchyma like areas and positivity for cytokeratin in epithelial like areas, are crucial for diagnosis⁶ (as is with the case reported above).

Surgery remains the treatment of choice; post operative radiotherapy is advised to improve local control. Chemotherapy has no role in the management of synovial sarcoma⁷.

In conclusion, synovial sarcoma of larynx is a very rare tumor of head and neck region. Immunohistochemistry allows diagnosis to be made accurately. An aggressive surgical approach and post operative radiotherapy are the options of choice in the management of these cases. A thorough follow up offers possibility of identification of metastasis or recurrence as in the case reported above, and may often be successfully subjected to surgical treatment.

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