IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 12 Ver. II (December. 2016), PP 08-09
www.iosrjournals.org

Metastatic Clear Cell Sarcoma- Role of Temazolamide

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Abstract
Objective: *To describe outcome of patients with relapsed Clear cell sarcoma when treated with Temazolamide.*
Methods: *36 year gentle man a diagnosed case of clear cell sarcoma of foot post wide local excision was being treated with adjuvant RT. On regular follow up patient had multiple time regional recurrence along with distant metastasis for which he received Temazolamide and had partial remission.*
Result: *With regional therapy (surgical) and systemic chemotherapy in form of temazolamide such aggressive tumour can be managed and patient can achieve long term survival.*
Conclusion: *clear cell sarcoma are a rare, heterogeneous group of solid tumor in need of improved therapeutic optionsTemozolomide has demonstrated an encouraging activity in the treatment of CCS metastasis in heavily pre-treated patients and was safe and well tolerated.*
Keywords*: temazolamide , clear cell sarcoma ,*

I. Introduction

Clear-cell sarcomas is a rare tumor account for less than 1% of all soft tissue tumours[7]. It has morphological similarities to malignant melanoma but a distinct genetic background including a chromosomal translocation, t (12; 22) (q13; q12), or a resultant EWSR1-ATF1 fusion gene[5]. The tumor has high propensity for metastasis and recurrence with 5- year survival rate of 50%. Chemotherapy has been shown to have limited benefit, with platinum-containing regimens offering the most potential benefit. We report a case of clear cell sarcoma of foot and his survival.

II. Case report

A 36 year man reported in onco-surgery department of NIMS, Hyderabad with a history of left foot ulcerative growth(of 6 month duration) post excision biopsy wide local excision (done outside) in august 2007. There was h/o warty growth at similar site in 2006 which was excised but biopsy was not done. The histopathology reports revealed amelanotic melanoma, review of the slides was done. On IHC the tumor was found to be HMB+ve and S100+ve[1]. It was diagnosed to be a case of clear cell sarcoma. The previous HPE report did not mentioned about the margin status. Clinically scar was present over the dorsum of left foot, no evidence of lymphadenopathy, chest xray-NAD. Patient was advised RT (radio therapy). He received RT from 06.10.2008 to 18.11.2008 60Gy /30#. Patient was on regular follow up thereafter. Patient developed left inguinal swelling in june 2009. FNAC revealed metastatic clear cell sarcoma, chest CECT revealed sub pleural nodule in right upper lobe? Metastatic. Patient underwent left ilio-inguinal block dissection + eversion of sac at our hospital on 26.06.2009. HPE report clear cell sarcoma.

Patient developed recurrence at left inguinal region which was confirmed by FNAC report in September 2009. Palliative chemotherapy was started and patient was started on Tab. Temozolamide300mg ,5 days/month for 1 year. The swelling subsided on treatment but it again recurred in Nov, 2010. CECT chest and abdomen revealed right upper lobe of lung and metastatic left inguinal lymphadenopathy. Patient underwent left ILBD in jan.2011. there after chemotherapy with temazolamide was resumed.

After 3 months in march 2011 patient developed metastatic lymphadenopathy in right inguinal region with metastatic nodules at nape of neck. On treatment with Tab. Temazolamide for 6 months the swelling subsided.

During treatment the patient developed low back ache. PET scan was done which showed lumber vertebrae L5 metastatic deposits and left inguinal & right thigh soft tissue deposits[9]. Patient received external RT to lumbar spine from 11.12.12 to 22.12.12 30 Gy/10#. Patient was restarted on chemotherapy. In the last visit on 28.05.13 patient noticed swelling over the right thigh.

III. Discussion

This case report highlights the challenges faced in management of clear cell sarcoma. Such propensity of clear cell sarcoma to metastasise is rarely reported. In spite of having recurrent metastasis patient is still doing well. The average age at the time of the first manifestation of clear-cell sarcoma is about twenty-five

years, and the most frequent site of the primary tumour is the foot[8]. Clinically, the mass is of moderate size and grows slowly. Only 50% of patients report pain[12]. Therefore, years may elapse between the first occurrence of the tumour and treatment.

To differentiate clear-cell sarcoma from malignant melanoma, cytogenetic analysis can be helpful; while the translocation t(12;22)(q13;q13) seems to be a primary feature of clear-cell sarcoma, this translocation has not yet been observed in malignant melanoma[4].

Most authors have emphasized the highly malignant potential of this entity. Wide local excision is the appropriate surgical treatment. If wide margins cannot be achieved, radical excision or even amputation is required. Regional lymphadenectomy is not obligatory. The most common sites of metastatic spread include the regional lymph nodes, lungs, skeletal system, liver, and heart. The frequency of skeletal involvement ranges between 15% and 26%. This patient has multiple metastases to different site like bone, pulmonary, lymph node and soft tissue. With regional therapy (surgical) and systemic chemotherapy in form of temazolamide such aggressive tumour can be managed and patient can achieve long term survival. Further studies are needed to define the role of temazolamide in metastatic clear cell sarcoma.

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