Clear Cell Sarcoma of Cervical Cord Region: An Unusual Anatomical location

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ABSTRACT

Introduction: Clear cell sarcoma of soft tissue (CCSST) is a rare tumor accounting for less than 1% of all sarcomas. The most common location is extremities. We report an extremely rare case of CCSST of cervical cord region. **Case Presentation:** A 44 year old male patient presented with complaints of right neck swelling and pain cervical spine region radiating to right arm. Lymph node biopsy showed metastatic clear cell sarcoma of soft part (melanoma of soft part). Whole body PET CT scan reported metabolically active enlarged right posterior cervical lymph node and hypermetabolic focal lesion in right side of spinal canal The intraspinal tumor was unresectable so patient received External Beam Radiotherapy. Follow up MRI after six months reported stable disease. **Conclusion:** CCSST is a rare tumor and our patient had tumor location in cervical cord region which is an unusual location. In surgically inoperable case like this, radiotherapy may be considered as treatment option and it has resulted in symptom-free survival in this patient till date.

Key Words: Clear cell sarcoma, Intraspinal, cervical cord

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INTRODUCTION

Clear cell sarcoma of soft tissue (CCSST), first described by Enzinger in 1965, is a sarcoma with melanocytic differentiation[1]. It is a slow growing tumor derived from melanoblast like cells located within subcutaneous tissue, tendon and aponeuroses of young adults

aged 20-40 years. It is a rare tumor accounting for less than 1% of all sarcomas. It occurs most commonly in extremities. Tumor rarely arises in head and neck, bone, gastrointestinal tract and kidney. CCS derives its name from abundant amount of intracytoplasmic vesicles. We report an extremely rare case of CCSST of cervical cord region.

CASE REPORT

A 44 year old male patient presented with complaints of right neck swelling and pain cervical spine region radiating to right arm for 4 months. FNAC from right posterior triangle lymph node reported metastatic poorly differentiated carcinoma. Lymph node biopsy showed metastatic clear cell sarcoma of soft part (melanoma of soft part) (Figure:1,2).

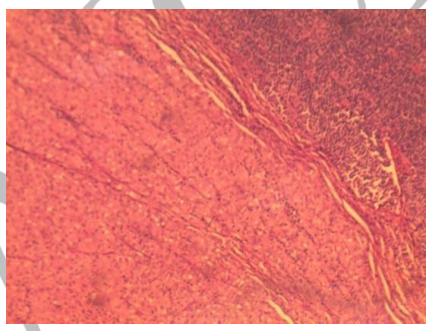

Figure:1 100x view showing tumor cells arranged in small clusters metastatic to lymph nodes

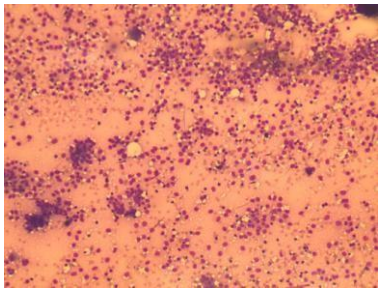

Figure: 2 400 x view showing nests of tumor cells having abundant cytoplasm,round to polygonal nuclei, metastatic to lymph nodes

Immunohistochemistry reported malignant cells expressing Vimentin, S100 and HMB-45. Pre and post contrast MRI of oral cavity and neck showed enhancing altered signal intensity lesion in right C3-C4 neural canal with contiguous epidural intraspinal component extending from C2 to C4 vertebral levels, suggestive of neurogenic mass (Figure:3).

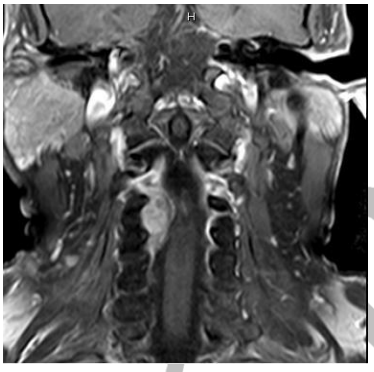

Figure: 3 CEMRI Cervical spine showed enhancing altered signal intensity lesion in right C3-C4 neural canal with contiguous epidural intraspinal component extending from C2 to C4 vertebral levels

Size of the lesion was 3.6(CC)x2.2(Tr)x1.4(AP) cm and there was focal effacement of thecal sac with mild focal indentation of the cord. A centimeter short axis left upper deep cervical node was seen. Whole body PET CT scan reported metabolically active enlarged right posterior cervical lymph node (1.3x1.6cm, SUV-55.3) and hypermetabolic focal lesion in right side of spinal canal (displacing cord towards left) at C3 vertebral level and extending into neural foramina of C3-C4 vertebra (SUV max-57.1). The intraspinal tumor was unresectable so patient was referred for External Beam Radiotherapy in view of pain. He received dose of 50.4 Gy/28fr to the spinal cord lesion and 58.8 Gy/28fr to nodal region. Chemotherapy was not administered, as requested by the patient. Follow up MRI after six months reported stable disease.

DISCUSSION

CCS is a rare tumor representing around 1% of all soft tissue sarcomas.[1] It is also known as melanoma of soft parts, acknowledging the similarity in the neural crest origin as well as in the

cytological, immunohistochemical and ultrastructural features of both tumors. The unique t (12; 22) (q13; q12) translocation, younger age of presentation, location in deeper soft tissues accompanying with tendons/aponeuroses, absence of epidermal involvement, is generally located in non-pigmented areas and predilection to the extremities help in distinguishing it from malignant melanoma[2,3].

The usual sites of the neoplasm are in the extremities, especially the region of the foot and ankle, followed by the knee, thigh and hand. a few reports of CCS have occurred at unusual sites which include lung, chest wall,[4] cervical spinal cord[5], scapula[6], retroperitoneum[7].

Patient with CCSST have variable unpredictable course of the disease. It has high propensity of lymph node metastasis (10-18%) with poor prognosis. Other sites of metastases often include lung, skin, bone, liver, heart and brain. Immunoreactive markers used to delineate CCS from epithelial tumors are S100 and HMB-45. Molecular genetics study includes t(12;22) (q13;q12) chromosomal translocation, typically not present in cutaneous malignant melanoma. Surgical resection, radiotherapy or chemotherapy have no reported significant advantage of one therapy over another.

Necrosis and a larger tumor size (>5 cms) is associated with a high rate of distant failure[8]. Prognosis is also significantly better for patients whose tumors arise in the extremities rather than at other sites where complete tumor excision is more difficult.

CONCLUSION

CCSST is a rare tumor and our patient had tumor location in cervical cord region which is an unusual location. In surgically inoperable case like this, radiotherapy is a treatment option and it has resulted in symptom-free survival in this patient till date.

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