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A Rare Presentation of Soft Tissue Sarcoma

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Abstract : *Soft tissue sarcoma are one of the most common malignancy with high incidence of lung metastasis which occur in about 80% of patient within 2 years duration. Though brain metastasis is a relatively a rare event in the natural history of soft tissue sarcomas it occurs in about 7% of the patient with soft tissue sarcoma. A direct metastasis to brain without lung metastasis s very rare and only few cases have been reported. The increasing use of chemotherapy may have caused a reduction in local relapses as well as distant failures leading to an improvement in survival, thereby allowing metachronous seeding of the brain, a sanctuary site. The purpose of this report is to increase awareness amongst clinicians regarding such a possibility.*

Keywords : *soft tissue sarcoma, brain metastasis*

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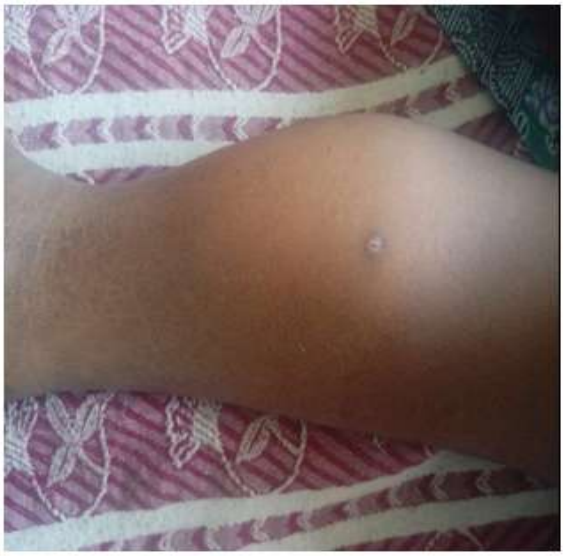
I. Introduction

Soft tissue sarcomas are rare and unusual neoplasms, accounting for approximately 1% of adult human cancers and 15% of pediatric malignancies[1]. Sarcomas continue to carry biologic and clinical interest and significance disproportionate to their clinical frequency because of their often clearly defined molecular genetics and the vast expansion of cytogenetic and molecular genetic information that has been discovered over the past 10 years[2]. Our patient had a rare presentation of soft tissue sarcoma in her left leg with metastasis to brain which is very rare. Adding to its peculiarity is that patient did not have a lung metastasis which is very common with soft tissue sarcoma.

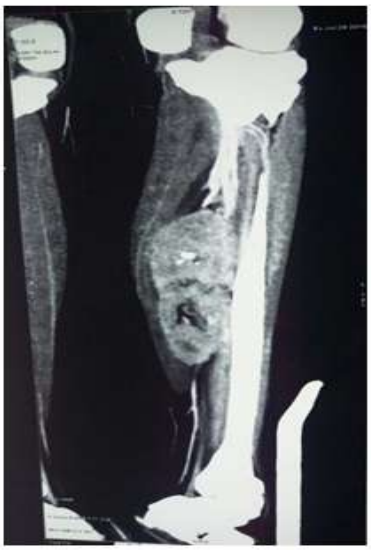
II. Case report

A 29 year old female patient presented to us with chief complaints of swelling in the left leg for the past one month duration. Swelling was insidious in onset, initially small and gradually increased in size to attain the present size. Patient had no pain or any signs of inflammation. No history of loss of weight and appetite She also complained of headache and vomiting for the past 2 weeks. No other neurological deficit. Patient had no history of chest discomfort or bone pain. On examination there was a swelling in left leg of size 8 X 5 cms extending from 2 cm below the left popliteal fossa up to 3 cms above the ankle joint. Swelling was hemispherical shaped, surface was smooth, and skin was normal and was firm in consistency. Swelling had restricted mobility on contracting the calf muscle. There was no lymphadenopathy and other system examination was normal.

There was no focal neurological deficit. A provisional diagnosis of soft tissue sarcoma was made. We did MRI of the local part which showed the soft tissue lesion which has also invaded the peroneal nerve. We proceeded with trucut biopsy which revealed extra skeletal Ewings sarcoma. We proceeded with metastatic work up. We did CT chest and there was no metastatic leision. Since patient had persistent headache and vomiting not related to food intake we took a CT brain for the patient. CT brain showed a metastatic leision in the left temporal region.

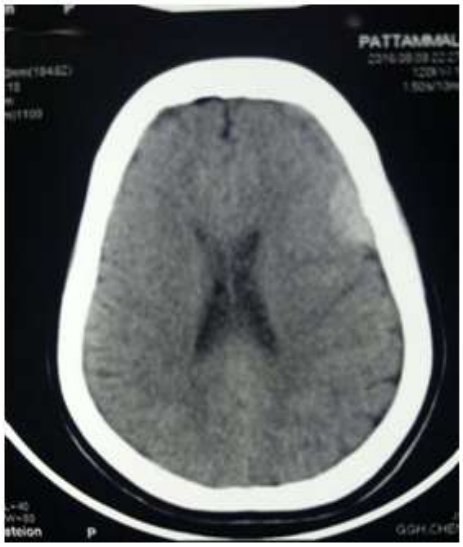


[Soft tissue sarcoma- left leg]



[MRI showing the leision]

[CT chest showing no lesion]



[CT brain showing Metastatic lesion in left temporal region]

After discussing in the tumor board, patient was sent to radiation oncology department for whole body irradiation.

III. Discussion

Soft tissue sarcomas are tumors arising from a diverse and complex group, as they usually display varying degrees of mesenchymal differentiation. Simple surgical excision is sufficed for benign soft tissue tumors. Less than 1% of the overall human burden of malignant tumors is soft tissue sarcomas but remain life threatening, and approximately 40% of patients with newly diagnosed soft tissue sarcoma die of the disease, corresponding to approximately 4,000 deaths each year in the United States[1]. Soft tissue sarcoma, diagnosed at an early stage, is eminently curable. When diagnosed at the time of extensive local or metastatic disease, it is rarely curable. The relatively small numbers of cases and the great diversity in histopathologic features, anatomic sites, and biological behaviors have made comprehensive understanding of these disease entities difficult. A better understanding is urgently needed to accelerate the development of new treatments.[2]

A total of 45% are located in the extremities, with 30% of all lesions occurring in the lower limb (most commonly in the thigh); 38% are intra-abdominal, divided between visceral (21%) and retroperitoneal (17%); 10% are truncal; and 5% are head and neck[3, 4]. Soft tissue sarcomas become more common with increased age, and the median age at diagnosis is 65 years. However, the median age varies significantly by histologic type and subtype[4]. In general, the median age of onset tends to be 20 to 50 years in the translocation-associated sarcomas

and 50 to 70 years in the complex sarcoma types. The most extensive resection, amputation, should be only rarely indicated for soft tissue sarcoma[5].At MSKCC, the amputation rate, which was 50% in the late 1960s, is now <5%. Outcomes from amputation versus limb-sparing surgery for extremity lesions were addressed by a prospective, randomized trial with well over 10 years' follow-up. Although local recurrence is greater in those undergoing limb-sparing operation plus irradiation than in those undergoing amputation, disease free survival is not different.[6] Moreover, the level of handicap can be significantly lower in patients treated with limb-sparing surgery. Amputation should be reserved for tumors that cannot be resected by any other means, in patients without evidence of metastatic disease and with potential for good long-term functional rehabilitation.[7]

Brain metastases from soft tissue sarcomas are usually rare. However, patients with neurological symptoms should be appropriately investigated promptly as presented in our case. Surgical resection of brain metastasis can be taken into consideration for solitary brain metastasis in non-eloquent areas. Palliative radiotherapy is appropriate for patients with multiple brain metastases or co-existing extra-cranial disease[8, 9]. Whole Brain Radiation Therapy (WBRT) has been the standard treatment for patients with brain metastases. As WBRT is widely available, is easy to deliver safely and accurately, is effective in providing palliation for the majority of patients, and does not require sophisticated technologies, it has long been considered the gold standard for treatment of brain metastases and represents the most common treatment modality worldwide. Symptomatic improvement has been poorly assessed and is variable[10].

IV. Conclusion

Although brain metastases from STS are rare, symptomatic patients should be appropriately investigated. Neurosurgical resection may be considered for solitary brain metastasis confined to no eloquent cortex associated with significant mass effect. WBRT should be appropriate palliative treatment for patients with multiple brain metastases or co-existing extra-cranial disease. CECT thorax is needed to rule out pulmonary metastases definitely. In case of synchronous lung metastases, palliative chemotherapy may be offered. The outlook of these patients continues to remain grim despite aggressive therapy.

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