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Case Report

A rare case of 22 year old male having spindle cell sarcoma and distant metastasis

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ABSTRACT

Sarcomas are a group of cancers that have varied presentations in different locations in our bodies, can metastasize and show a range of histopathological features and are very uncommon. We report a case of a 22-year-old male presented with a case of high-grade spindle cell sarcoma in the posterior mediastinum of size 13×13×8 cm3 along with left moderate pleural effusion of the left lung. There are also multiple enhancing hypodense lesions in other parts of the body indicating neurofibromatosis. The tumor was detected using CECT Chest, Abdomen, an MRI was also done and results were confirmed by IHC. The patient was given chemotherapy however that only resulted in subsequent increase in size of the tumor since the patient did not report for the treatments regularly. This shows how important it is to be consistent with the treatment, failure of which results in grave consequences.

Keywords: Spindle cell sarcoma, Neurofibrosarcoma, Immunohistochemistry, Corpus callosum, Distant metastasis

INTRODUCTION

Sarcoma is a term for a wide group of cancers that starting in the bone and soft connective tissues. They are uncommon and hence their actual incidence is difficult to determine as its occurrence is less than 1% of all cancers.[1]Sarcomas have different representations at different locations in the body and have an inclination to metastasize.[2] Concurring to the morphology of cells found, sarcomas can be of small cell type, spindle cell type, epithelioid type, pleomorphic type etc.[3] Spindle cell sarcoma is a rare type of soft tissue sarcoma, undifferentiated in nature.[4] Males are more likely to be affected by it than females.[5]

Treatment includes surgery, fundamentally to remove the tumor and adjuvant radiations that can be used as primary therapy.[6] Here we discuss a case of 22-year-old male presented with spindle cell sarcoma along with neurofibromatosis and the results following neglect to regular follow-ups.

CASE REPORT

This is a case of 22-year-old male patient who presented to the hospital with complaints of chest pain and breathlessness along with swellings on the chest and abdomen. The patient is a defaulter and came to the hospital 3 months after the diagnosis of spindle cell carcinoma was made, with complaints of chest pain, back pain, abdominal pain, loss of appetite, pain during breathing, and loss of 10kg of weight. He has a significant family history. His mother has similar soft swellings on chest and abdomen while brother had a brain tumour 4 years ago. The patient had a single episode of GTCS and he hasn't been on any medication since childhood. A CECT of chest revealed well defined multiple rounded lesions in posterior mediastinum soft tissue with largest one measuring 13×13×8 cm. It was

inferred that left moderate pleural effusion seen along with the collapse of underlying lung parenchyma may represent metastasis. CECT of abdomen and pelvis revealed multiple enhancing hypodense lesions in subcutaneous plane with the largest measuring 3.8×2.7 cm in the left infraumbilical region. An MRI brain showed moderate dilatation of bilateral lateral ventricles with para ventricular edema, thinning of corpus callosum and an indeterminate lesion in R ambient cistern. Diagnosis of neurofibrosarcoma and left pleural effusion was made which was followed by a biopsy and IHC for confirmation. The IHC reports concluded high grade spindle cell sarcoma and a probability of malignant peripheral nerve sheath tumour and case of neurofibromatosis.

After 6 months the patient came in with complaints of severe chest pain and weight loss. CT of neck, thorax and abdomen showed that the tumour mass had expanded to the size of 27.6×1.8×17.7 cm. this tumour mass caused the displacement of left kidney, pancreas, liver, spleen, aorta and the esophagus .it was causing the collapse of left lung and expansion of the left hemithorax. It also showed a few heterogenous enhancing lesions in the left lobe of liver which indicated a possibility of metastasis. Management includes surgery with radiotherapy or chemotherapy. He was also given a blood transfusion of 390 ml. The patient has received 40 chemotherapy sessions till date. He is now put on palliative care.

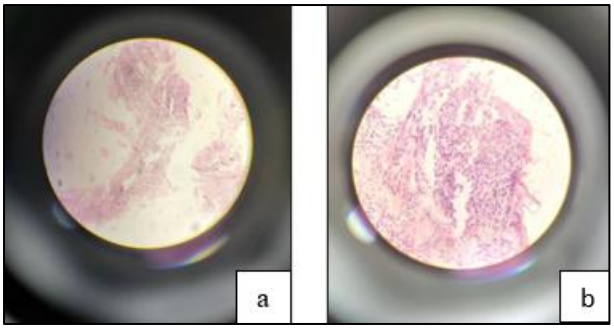

Figure 1 (a and b): Histopathological examination of tumour.

DISCUSSION

Sarcomas represent abnormal cell clusters that develop in bone and connective tissue. Spindle cell sarcoma is an uncommon type of cancer which falls under its umbrella. In this condition, the cells resemble elongated spindles under microscopic examination, characterized by their long and narrow shape with tapered ends akin to candlesticks. They commonly form in the long bones such as femur, humerus and tibia. There are various factors which enhance the development of this condition such as osteomyelitis, bone infarction, fibrous dysplasia, Paget's disease of bone and previous radiation therapy. Their clinical manifestations include bone pain, fracture, fatigue, malaise and tenderness or swelling at the site of tumour.[7] People of almost all ages are affected by spindle cell sarcoma, though it is more common in middle and older age groups with the median age at diagnosis being 61 years. A sex ratio of 1.11:1 (male:female) has been observed in the ones affected with the disease. The race distribution indicates that it is more common in the white population.[8] The differential diagnosis of spindle cell sarcoma includes liposarcoma and other spindle cell neoplasms like malignant melanoma and spindle cell lipoma due to the presence of similar histopathological findings of spindle shaped cells. Another differential is intradermal nodular fasciitis. Sarcoma can be mistaken for nodular fasciitis when rapid mitotic activity and increased cellularity are observed on biopsy. But, while nodular fasciitis grows quickly, it's relatively small in size and doesn't show pleomorphism, atypical mitoses and nuclear hyperchromatic as seen in the sarcoma. Hence, in order to prevent misdiagnosis, it is essential to perform repeated biopsies and imaging studies.[9,10] Imaging plays a crucial role in diagnosing sarcoma, often revealing the tumor's size rather than its specific type. In our case, a CECT scan revealed chest area enlargement and a lesion, while biopsy and IHC tests confirmed neurosarcoma.[11]

The prognosis of sarcoma relies on the tumor's site, its grade and its stage, which encompasses metastasis.[12] Accurate grading of tumour is essential for prognosis and to determine the treatment plan. It can be done either by cytologic grading or the histological grading of which the former is more important. Cytologic grading takes into consideration four factors, nuclear atypia, nuclear overlap, mitosis and necrosis. On this basis, there are four grades. Its sensitivity and specificity are 95.23% and 80.76 % respectively. While histologic grading takes into account three factors: tumour differentiation, mitosis count and tumour necrosis. On this basis, there are three grades. This criterion can be used on fine needle aspiration cytology samples of spindle cell sarcomas with 88 % accuracy.[13]

Treatment is done in stages and includes chemotherapy, surgery and radiation therapy in most cases. In this, both preoperative and postoperative radiation therapy offer distinct advantages in improving outcomes.[11] As part of our patient's management plan, he has completed a total of 40 chemotherapy sessions to date, aimed at excising the tumor. Precise removal is crucial for treating patients with Spindle cell sarcoma, otherwise, an imprecise removal could affect the risk of local relapse.[14] This also applies to chemotherapy i.e., which drug should we go with. CNS metastases in SpSCC can be addressed using pembrolizumab, an anti-PD-1 therapy. Additionally, for treating spindle cell sarcoma in the nasal cavity, 5-fluorouracil has shown efficacy.[15,16] Currently, pembrolizumab plus platinum and fluorouracil are approved as the first line treatment.[16]

CONCLUSION

After lighting on clinical findings of chest pain, back pain, stomach pain, loss of appetite, after a thorough investigation by CECT of chest and abdomen, MRI and biopsy followed by confirmation with IHC, the patient was diagnosed with spindle cell sarcoma. It is a very rare form of cancer with less than 1 percent occurrence amongst all types of cancers.[1] Finding a center with the right expertise for treatment along with patient's cooperation is extremely important in treating such type of cancers pain during breathing patient has diagnosed of Spindle cell sarcoma in the posterior mediastinal region. So, when it comes to soft-tissue sarcomas, it's important to go to a center with the right expertise. They can provide the best treatments and keep an eye on your progress with check-ups and imaging. It's all about giving you the best care possible. Also, patient should be punctual, aware and conscious about his/her treatment, this also matters at many levels.

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