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Pulmonary Spindle Cell Sarcoma – A Rare Case Report

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***Abstract*: Spindle cell sarcomas (malignant fibroushistiocytoma, hemangiopericytoma, fibrosarcoma, leiomyosarcoma, synovial sarcoma) are the most common primary pulmonary sarcomas. Adult-type fibrosarcoma is a rare and highly aggressive subtype of soft tissue sarcomas. Fibrosarcoma is characterized by its low sensitivity towards radio- and chemotherapy as well as by its high rate of tumor recurrences**

***Keywords*: fibrosarcoma, primary pulmonary sarcoma, spindle cell sarcoma.**

1. Introduction

Spindle cell sarcomas (malignant fibroushistiocytoma, hemangiopericytoma, fibrosarcoma, leiomyosarcoma, synovial sarcoma) are the most common primary pulmonary sarcomas. Here we are presenting a case of a Spindle Cell Sarcoma which was in the end confirmed as a Fibrosarcoma after IHC.

Fibrosarcoma is a rare, highly malignant tumor of mesenchymal cell origin. It derives from pathologically transformed spindle shaped fibroblasts with an excessively high division rate. The diagnosis of fibrosarcoma is by one of exclusion. Using immunohistochemical and molecular techniques, it is possible to further subdivide the various subtypes of fibrosarcoma which can be very similar in their morphology, tumor genetics and clinical manifestation.

2. Case History

An 18 year old male who presented with cough with streaky hemoptysis, breathlessness, swelling over the left side of forehead and associated significant weight loss for a period of 3 months duration. He and his family never had any significant medical history. Physical examination revealed a swelling of size 5x4x3cm swelling seen in the left frontal region fixed to the underlying bone with swelling extending into left eye causing ptosis, another swelling of size 3x3 cm on the occipital region was also palpable which was also fixed to the underlying bone a third swelling was palpated on right side of neck 3x2 cm which was having the same consistency as previously mentioned swelling. There were reduced breath sounds over the right lung. On abdominal examination there was 6x6 cm swelling palpated on the left umbilical, left hypochondrium extending 6 cm below the left costal margin on midclavicular line the swelling was ballotable and not moving with respiration. Rest of the systemic examination was normal.

3. Diagnostic Work Up

Laboratory findings were normal. Chest Xray (a) showed non homogeneous opacity involving Right MidZone/Lower Zone with multiple rounded opacities involving bilateral lung feilds. We then followed up with a HRCT Thorax (b) which showed a fairly well defined smooth intraparenchymal lesion involving the right middle and lower lobe with multiple well defined non enhancing lesions scattered throughout bilateral lung parenchyma with few of them showing cavitation. USG (A+P) Well defined solid cystic lesion arising from upper pole of Left Kidney 11x10x9 cm showing significant internal vascularity and the lesion abutting the tail of pancreas the lesion is not separately visualized from the adrenal gland.

CT Brain(c)showed a large mass of size 7x 6 cm in the frontal bone it abuts the underlying brain parenchyma with mass effect, similar lesion was seen on the occipital region of size 3x3 cm. PET CT scan revealed multiple metastases to left kidney, mediastinal and cervical lymph nodes and skull deposits. We went ahead with a CT guided Tru Cut lung biopsy which showed Spindle cell morphology on HPE (d) and on further IHC examination showed Vimentin, SMA and CD99 Positive which confirmed the diagnosis of Fibrosarcoma.

4. Images

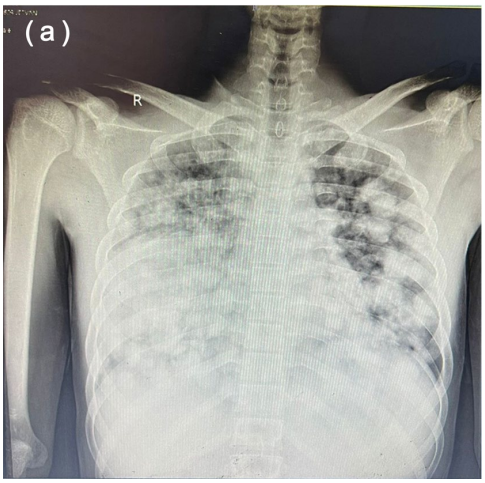

Fig. 1.

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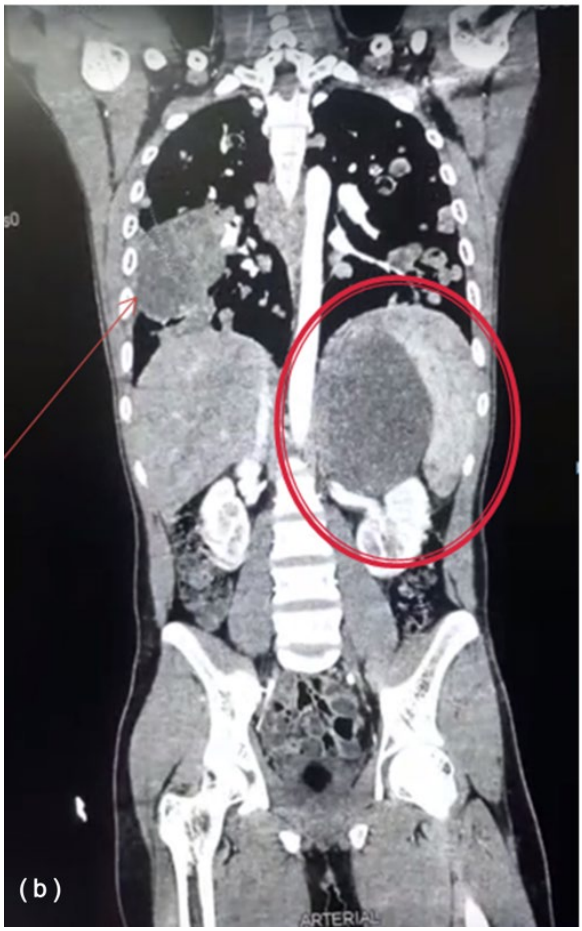

Fig. 2.

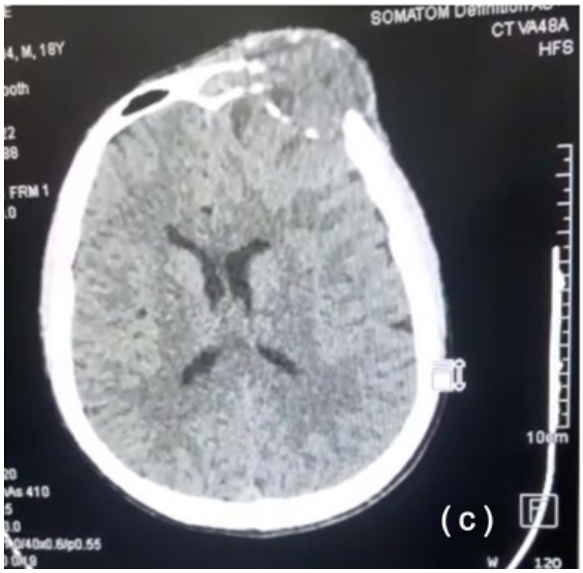

Fig. 3.

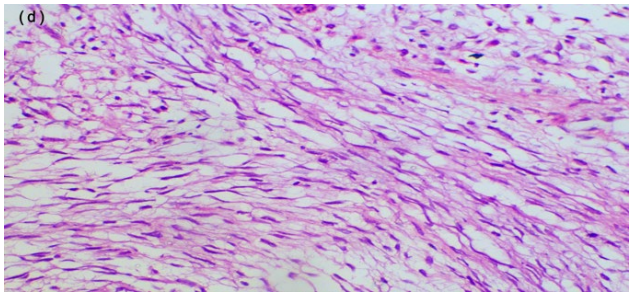

Fig. 4.

5. Treatment

Due to the extensive metastasis of the malignancy surgical resection couldn't be done and after Oncologist opinion patient was started on Inj Doxorubicin and Inj G-CSF, but due to extensive metastasis and aggressive nature of malignancy patient succumbed to the illness.

6. Discussion

The best current therapy of fibrosarcomas is complete surgical removal [1]. But surgical resection is not possible every time as sometimes the masse may be in the head, neck and there may be multiple metastases. Even though the response rate of fibrosarcoma towards radio and chemotherapy is very low, they are broadly used as a neoadjuvant and/or adjuvant tumour treatment [2]. Histopathology alone is not sufficient for a clear distinction between fibrosarcoma and other spindle-cell neoplasms.

Immunohistochemistry (IHC) is applied in the diagnostics of fibrosarcoma where specific antibody reagents allow the detection of differential diagnostically important tumor markers [3].

7. Conclusion

An accurate diagnosis is a prerequisite for the design of an adequate treatment plan.

The best prognosis is seen when;

i. complete surgical tumour resection with histological tumour-free margins,
ii. The use of agents that lead to a reduction in tumour proliferation and migration and/or inducing remission via neoadjuvant or adjuvant therapy,
iii. Preventing tumour invasion and metastasis [4].

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