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Case Report

Superior vena cava obstruction as manifestation of synovial cell sarcoma: a case report and review of literature

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ABSTRACT

Synovial cell sarcomas are one of the most common soft tissue tumors affecting adolescents and young adults. The common location of these tumors is often extremities. Primary involvement of mediastinum is a very rare presentation of this tumor, only few cases have been reported so far. We are reporting the case of a 27-year-old male patient who presented to us with a history of dyspnoea and chest discomfort of one week duration along with features of SVC obstruction. Chest is a frequently involved site of metastasis and lymphomas but rarely involved in synovial sarcoma. The learning point from this case is that rare causes like synovial sarcoma of mediastinum should always be included in differential diagnosis of mediastinal mass and SVC obstruction. We are reporting this case in view of its rarity. Though uncommon this case report emphasizes that synovial cell sarcoma should also be considered in differential diagnosis of mediastinal mass and early recognition with HPE, IHC and prompt institution of treatment can be lifesaving.

Keywords: Chemotherapy, Mediastinum, Superior vena cava, Synovial cell sarcoma

INTRODUCTION

Synovial sarcoma is one of the most common soft tissue tumors of adolescents and young adults. Approximately 1/3rd cases occur during first two decades of life.[1,2] It is of mesenchymal origin. Common sites of involvement include appendicular structures mainly involving the knees. The other unusual sites of involvement include head and neck region, abdomen. The involvement of mediastinum is very rare. Due to its rarity in the published literature we are reporting this case.

CASE REPORT

A 27-year-old male patient presented to us with a history of dyspnea and chest discomfort of one week duration, with aggravation of symptoms on exertion. On examination patient had features suggestive of SVC obstruction. Testicular examination was normal. His CBC was normal. Serum AFP, B-HCG and LDH levels were normal.

Chest X ray PA view showed mediastinal widening and this was followed by PET CT scan which showed 6.3x10.7x12.0cm mass occupying superior mediastinum with an SUV of 18.6 from clavicle to T8 vertebra encasing the arch of aorta displacing the carina completely. No lesion was detected elsewhere in the body. Patient underwent CT guided biopsy which revealed malignant spindle cell tumor and IHC was positive for Vimentin and BCL 2 and negative for LCA, Ki- 67 was 25-30% confirming synovial cell sarcoma. Patient was planned for Ifosfamide and adriamycin chemotherapy with sperm cryopreservation followed by surgical assessment. Patient was reevaluated after three cycles of chemotherapy with PETCT which revealed

stable disease. Patient was started on palliative chemotherapy with gemcitabine and docetaxel in view of non-operable disease.

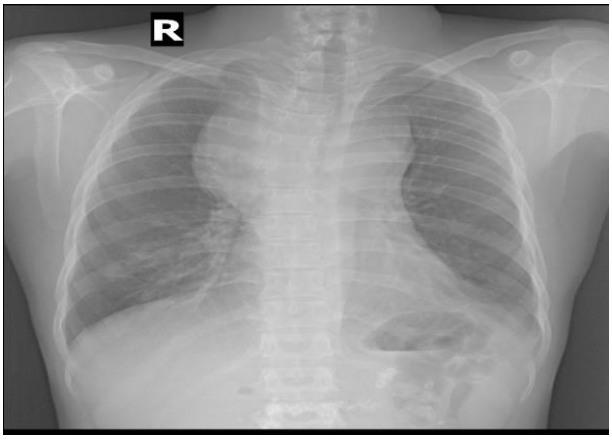

Figure 1: Chest X-ray.

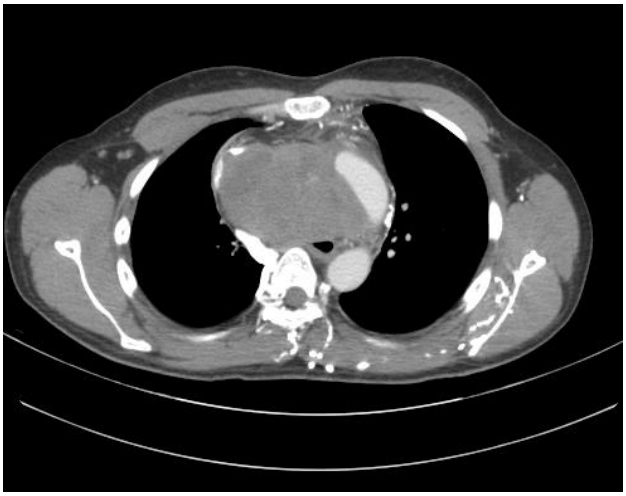

Figure 2: CT Thorax.

DISCUSSION

Synovial cell sarcoma are one of the most common soft tissue tumors affecting adolescents and young adults. The common location of these tumors is often extremities (85%).[1,2] Common sites of involvement include appendicular structures mainly involving the knees. The other unusual sites of involvement include head and neck region, abdomen, thoracic cavity pleura. Primary involvement of mediastinum is a very rare presentation of this tumor, only few cases have been reported so far.[3]

It is called synovial sarcoma due to its resemblance to synovial tissue on light microscopy. It is commonly seen in 2nd to 4th decade of life and it primarily involves extremities particularly the knees. The tumor is thought to arise from pluripotent mesenchymal cells. The differential diagnosis includes other mediastina l tumors such as sarcomatous mesothelioma, smooth muscle tumors, spindle cell sarcoma, thymoma, lymphoma and pleuropulmonary blastoma.

Pathological examination

On gross examination tumors are well circumscribed displaying both solid and cystic areas.

On Histological examination classical tumor demonstrates a biphasic pattern (i.e. presence of both epithelial and spindle cell components) but it may be monophasic as well. Another type worth mentioning is poorly differentiated type.[3]

The cytogenetic study usually reveals translocation (X;18) (p11, q11) which is detected in up to 90% of all the subtypes.[3]

On IHC the tumor displays strong positivity for Vimentin, EMA and BCL2 and usually necessary and in dispensable tool in diagnosing and guide further management of the disease.[3]

Though ideal management of such cases would be complete excision with good margins, but it is seldom feasible due to adjacent vital structures and morbidity associated with the procedure. Ifosfamide and adriamycin chemotherapy is administered in both adjuvant and neoadjuvant setting.[4-6] Role of radiotherapy is for achieving local control after surgical resection.

CONCLUSION

Though uncommon this case report emphasizes that synovial cell sarcoma should also be considered in differential diagnosis of mediastinal mass and early recognition with HPE, IHC and prompt institution of treatment can be lifesaving

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