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Case Report

Synovial cell sarcoma of the pleura: an uncommon cause of a haemorrhagic pleural effusion

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ABSTRACT

Soft tissue tumors account for a small percentage of malignancies and synovial sarcomas account for 10% of soft tissue tumors in our body, among them are the synovial sarcomas (SS).These arise from various sites and are of mesenchymal origin, Primary pleural synovial sarcomas are very rare tumours and account for not more than 0.5% of lung malignancies, they are being diagnosed mainly by the immunohistochemistry and classical cytogentic translocation t(x;18). This tumor has no sex preference and is commonly seen in the age group of 30-45.Our case is of a elderly male with massive pleural effusion who was diagnosed as having biphasic variant of synovial sarcoma of pleura by the help of immunohistochemistry. Treatment is mainly surgical resection and chemotherapy with ifosfamide/ adriamycin or radiotherapy. It must be considered as differential for biopsy proven malignant mesothelioma and hence immunohistochemistry should be must for all biopsy proven mesotheliomas.

Keywords: Haemorrhagic effusion, Immunohistochemistry, Primary synovial sarcomas, Pleural based tumours, Pleural effusions, SS18/SSX1 fusion gene

INTRODUCTION

Synovial sarcomas are a type of soft tissue tumours which most commonly arise in the deep soft tissues most commonly around the knee joint which includes 80% of such cases, it has a malignant potential, synovial cell sarcomas are also reported to occur in the head and neck, mediastinum, vulva, heart, oesophagus, lungs, abdominal wall, mesentery and retroperitoneum.[1]

Synovial sarcoma of the synovial sarcoma of the pleura has a very rare is its diagnosis. Only around 100 cases have been reported in the literature and are particularly very rare in elderly population. With the inclusion of immunohistochemistry in the investigatory armamentarium these days synovial sarcoma has been diagnosed much more frequently.

CASE REPORT

A 71 year old male patient presented to our OPD with a history of dry cough, right-sided chest pain and breathlessness since 1 month, He was a chronic cigarette smoker with pack years of 25. General physical examination was normal. On percussion there was stony dull note, on auscultation he had reduced breath sounds on right side. Chest X-ray showed evidence of massive pleural effusion on right side. Routine haematological investigations were normal. Pleural fluid was haemorrhagic and its ADA was 31.63 IU/L, Fluid Cytology was negative for malignant cells. CT scan of

thorax was suggestive of massive right sided pleural effusion with diffuse nodular and irregular mildly enhancing pleural thickening involving mediastinal, posterior and anterior costal pleura with underlying right middle and lower lobe collapse (Figure 1) and a radiological differential was given of malignant mesothelioma, further bronchoscopy was performed and it was suggestive of distorted and narrowed opening of right main bronchus, thoracoscopy was suggestive of unhealthy costal pleura and biopsy from pleura was taken (Figure 2), histopathology report mentioned tumor comprised of spindle cells arranged in sheets and vague fascicles with mitotic figures impression was given of low grade spindle cell mesothelioma (Figure 3), further immunohistochemistry report was calretinin, panCK, CD34 Negative which ruled out mesothelioma but instead IHC was positive for vimentin, BCL-2, EMA, CD99 positive (Figure 4) giving an impression of Synovial Cell Sarcoma of the pleura. Patient has been now referred to radiotherapy department where Adriamycin and ifosfomide based chemotherapy has been started and patient is doing well.

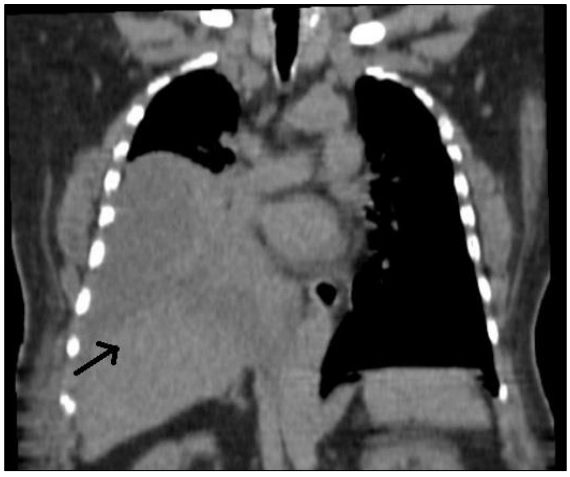

Figure 1: Massive right sided pleural effusion with arrow depicting diffuse nodular and irregular mildly enhancing pleural thickening.

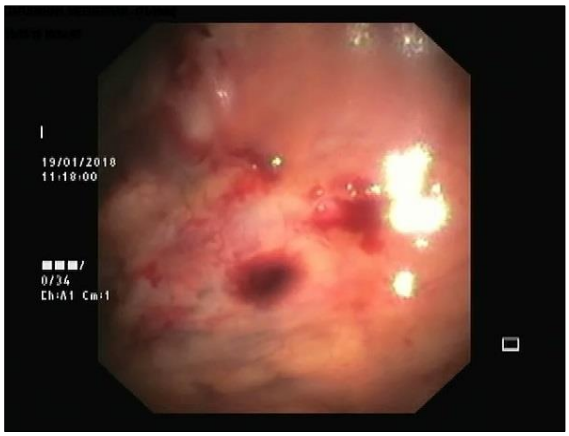

Figure 2: Thoracoscopy showing reddish elevated lesions from which biopsy was taken.

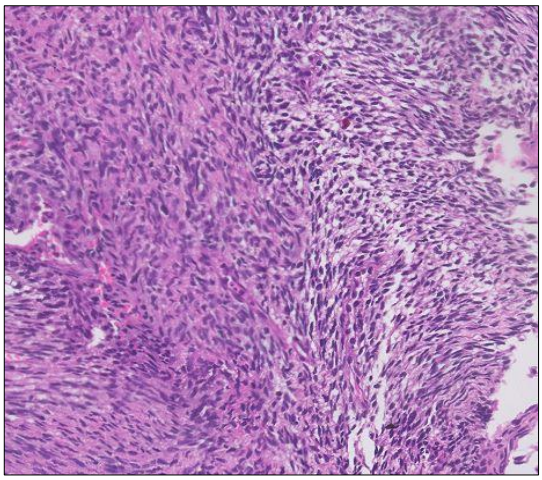

Figure 3: Tumor comprised of spindle cells arranged in sheets and vague fascicles with mitotic figures impression was given of low grade spindle cell mesothelioma.

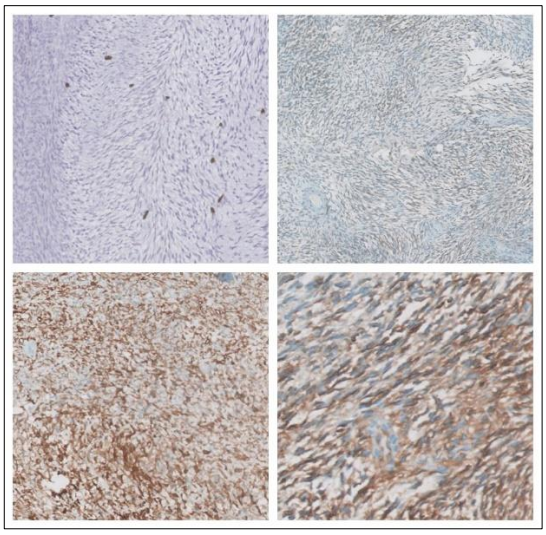

Figure 4: Immunohistochemistry positivity for vimentin, BCL-2, EMA, CD99 positive.

DISCUSSION

Primary pleural synovial sarcomas (PPSS) are rare neoplasms that have been in light since last two decades owing to advances in pathological investigations mainly immunochemistry. The first case was published by Gaertner et al in 1996.[2] They are exceedingly rare sarcomas which are mesenchymal in origin account for 10% of all soft tissue sarcomas. Synovial sarcomas should not be confused with the synovial fluid of joint. The presence of primary synovial sarcomas of pleura can be explained as the soft tissues and pleura both develop from the mesothelium. There is no sex predilection for this tumor and most of the cases in the literature are seen to be associated with 30-50 age group. There has been no causal association with smoking. Synovial Sarcomas have a highly aggressive nature when the primary are

arising from pleura, mediastinum or the chest wall.[3] Primary synovial sarcomas can be classified as monophasic, biphasic and poorly differentiated depending upon the cells present. Monophasic variety is composed entirely of spindle cells and the biphasic variety consists of both spindle cells and the epithelial cells. Very few cases are seen in elderly. Patient can present with plethora of symptoms but chest pain, dyspnea, dry cough being the most common. Radiological homogenous opacity with differential of pleural effusion is most commonly associated and CT scan may suggest well circumscribed mass with or without effusion, thickening of pleura is yet another common finding on CT scan. Because of the rarity of pleural synovial sarcoma and its similarity (clinical and histologic) to other pleural neoplasms, particularly sarcomatous mesothelioma. Presence of Bcl-2 protein represents a useful marker for differentiating synovial sarcoma. In our case the age group was not coinciding with the usual age group of presentation of PPSS and the biopsy was suggestive of malignant mesothelioma but Immunohistochemistry report was Negative for calretinin, panCK, CD34 which ruled out mesothelioma but instead IHC was positive for vimentin, BCL2, EMA, and CD99 positive giving an impression of Synovial Cell Sarcoma of the pleura (BIPHASIC VARIANT). The t(X;18)(p11.2;q11.2) translocation is present in more than 90% of these neoplasms, regardless of the histologic subtype and site.[4] The best treatment for synovial sarcomas of the pleura remains unclear also due to rarity of this tumour very few guidelines are available for the treatment of PPSS, mainstay of treatment is surgical resection with chemotherapy or radiotherapy. Ifosfamide and doxorubicin have shown good results in the 5 year survival period.[5]

CONCLUSION

PPSS are extremely rare neoplasms but should always be considered as a differential for haemorrhagic pleural effusions where HPE is suggestive of Mesothelioma mainly because of the different line of management. Also Immunohistochemistry has to be used as a investigation modality whenever possible and cytogenetic studies as GOLD standard for diagnosis whenever possible. There remains no gold standard therapy due to paucity of data and due its rarity currently Ifosfamide based regimens are currently standard of chemotherapy treatment along with radiotherapy. Future therapy mainly is directed to be more effective and less toxic SS18-SSX derived peptide vaccine is in the pipeline for the treatment for PPSS. An oral vascular endothelial growth factor receptor inhibitor PAZOPANIB has also been tried for the treatment of leiomyosarcoma and synovial sarcomas.

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