Synovial Cell Sarcoma Arising from Pleural Cavity: A Rare Case Report

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Abstract

Synovial cell sarcoma is the third most common case of soft tissue sarcoma in adolescent and young adult age group. Synovial sarcomas are rare malignant neoplasms of unknown histogenesis, most common affecting the lower extremities and frequently arises adjacent to joints or tendon sheaths. Synovial sarcoma is a misnomer because the tumor does not arise from the synovium; it only resembles synovial tissue at light microscopy. Since it appears to arise from as yet unknown multipotent, stem cells that are capable of differentiating into mesenchymal and/or epithelial structures and lack synovial differentiation. Synovial cell sarcoma arising from the pleural cavity is very rare in incidence and even rarely documented in literature. We here present a case of synovial cell sarcoma in a 25-year-old male arising in the pleural cavity.

Key words: Pleural cavity, Soft tissue sarcoma, Synovial cell sarcoma

INTRODUCTION

The name synovial cell sarcoma is a misnomer. The origin of synovial cell sarcoma is unclear. The name synovial cell sarcoma is given because of similarity between tumor cells and synoviocyte. Extremities is the most common site but a part from the extremities; synovial sarcoma may arise within head and neck, esophagus, retroperitoneum, and also in the thorax; mediastinum, heart, lung, pleura, or pericardium with lesser frequency.^{1,2} It preferentially affects young individuals.3 Recent data suggest the neurological origin of synovial cell sarcoma.4 Chromosomal translocation in synovial cell sarcoma is t(X;18)(p11;q11). Due to the fusion of gene SYT on chromosome 18 with gene SSX on chromosome X. Synovial cell sarcoma usually presents as a some nodule over extremities which remains benign for long duration and then increases rapidly in size. Prognosis of synovial

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cell sarcoma-like another soft tissue sarcoma is very poor. Surgical resection should be performed, whenever possible with neoadjuvant chemotherapy.

CASE REPORT

A 25-year-old male presented to outpatient department with complaint of left sided chest pain since 1 month and dyspnea on exertion since 1 month. There was no significant past history. On examination, breath sound was decreased on left infraxillary, inter- and infrascapular area. Chest X-ray posteroanterior view (Figure 1) and left lateral view (Figure 2) were suggestive of homogenous opacity occupying left lower zone with blunting of left costophrenic angle and cardiophrenic angle. Ultrasound chest revealed a pleural based mass. Computed tomography (CT) chest showed – A mass lesion arising from pleura and occupying the lower half of left hemithorax (Figure 3). CT guided biopsy was done which showed - spindle-shaped ells arranged in interlacing fascicles with intervening collagenous bundles (Figure 4). Cells have nucleus oval to elongated nuclie with moderate cytoplasm and indistinct cell borders. Immunohistochemical study was positive for smooth muscle actin (SMA), epithelial membrane antigen (EMA)

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Figure 1: Chest X-ray postero-anterior view showing homogenous opacity in left lower zone blunting left cardio- and costophrenic angle



Figure 2: Chest X-ray left lateral showing a mass lesion in left lower lobe

and Bcl2. A diagnosis of synovial cell sarcoma was made based on biopsy and immunohistochemistry report.

DISCUSSION

Synovial cell sarcoma is very rare tumor arising from immature mesenchymal cells. Pulmonary sarcoma comprises around 0.5% of lung malignancies. Chest pain, cough, hemoptysis are the common presenting complaints. The usual age of presentation is 25-30 years. Synovial cell sarcoma as often misdiagnosed as solitary fibroma, malignant fibrous histocytoma due to rarety of its incidence. Histology along with immunohistochemistry studies are diagnostic. Immunohistochemically, synovial sarcomas are nearly uniformly positive for cytokeratin, EMA, bcl-2, and vimentin, and negative for S-100, desmin, SMA, and vascular tumor markers. prognosis is very poor with 5 years survival rate of <50%.6 Synovial sarcomas are

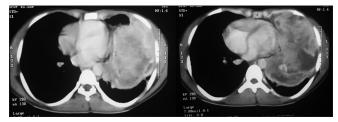


Figure 3: Contrast enhanced computed tomography chest showing mass lesion with contrast enhancement with areas of necrosis

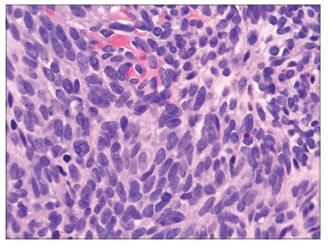


Figure 4: Pathology appearance of synovial cell of sarcoma

chemosensitive to ifosfamide and doxorubicin, however, the response rate is only 24%.

CONCLUSION

This case is one of the few case reports of primary pleural synovial sarcoma described in literature. It is often misdiagnosed as tuberculosis especially Indian subcontinent, which delays the diagnosis. Owing to its rarity and the paucity of data regarding its natural history, there are no guidelines for optimal treatment. Meanwhile, it consists of surgical resection associated with chemotherapy and/or radiotherapy. In our case, arriving at the correct diagnosis, with contemporary interventional pulmonology methods, and chemotherapy has improved the outcome of this otherwise aggressive tumor.

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