(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume4, Issue 2, Page No: 611-616
March-April 2021

Synovial Sarcoma Presenting As Right Sided Hemorrhagic Effusion: A Case Report and Review of Literature

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Type of Publication: Case Report
Conflicts of Interest: Nil

ABSTRACT

Bronchogenic malignancy and pleural secondaries are the major causes of massive hemorrhagic effusion. Synovial sarcoma of extremity presenting as pleural effusion is rarely reported in literature. Here we report the case of a 61 year old male who presented with right sided massive effusion. The diagnosis was made by biopsy taken via thoracotomy and confirmed by IHC. Subsequent evaluation by PET scan showed metabolically active lesion on left calcaneum. Here, we review the clinical details of this case and related literature.

Keywords: Synovial sarcoma, pleural effusion, thoracoscopy.

INTRODUCTION

Synovial sarcoma is a type of soft-tissue sarcoma usually seen in younger age group[1]. The disease starts most commonly in the legs or arms, but it can appear in any part of the body[2]. Synovial sarcoma presenting as pleural effusion is a rare entity.

CASE REPORT

61 year old male presented with progressive exertional dyspnea of 4 weeks duration. There was no h/o chest pain, cough or weight loss. He had past history of coronary artery disease and was on antiplatelets and also on medications for diabetes and hypertension. On evaluation his vitals were stable and there was stony dull note on right hemothorax with reduced breath sounds. His routine blood examination showed Hemoglobin- 12 g/dl, TC- 8800, ESR- 55mm/Hr, S. Cr- 0.9 mg/dl, and total protein /albumin - 7.8/ 3.9.

CXR (Fig: 1) showed right sided effusion and pleural aspiration from elsewhere revealed hemorrhagic fluid which was exudate, lymphocytic, low ADA with no malignant cells. CT thorax (Fig: 2) was s/o right massive effusion with collapse of underlying lung.

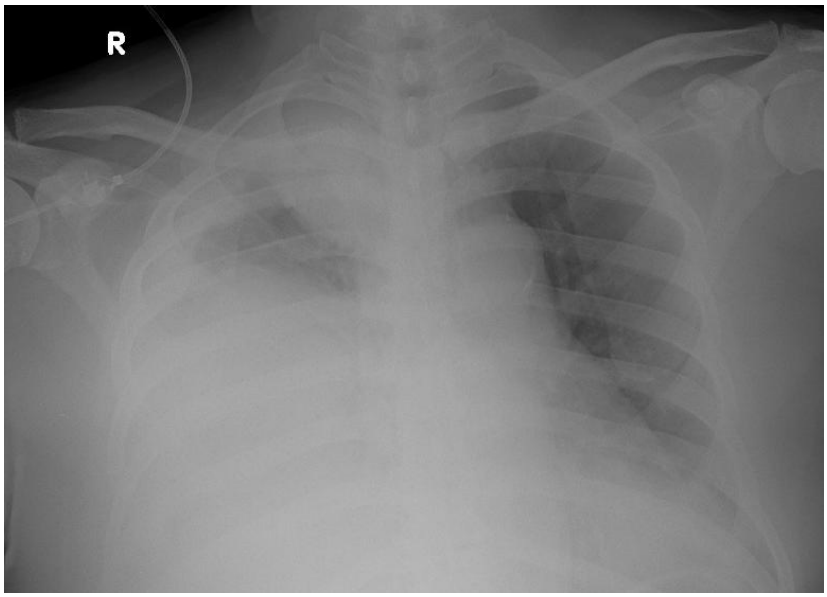

Figure 1: Chest X- ray showing right sided effusion

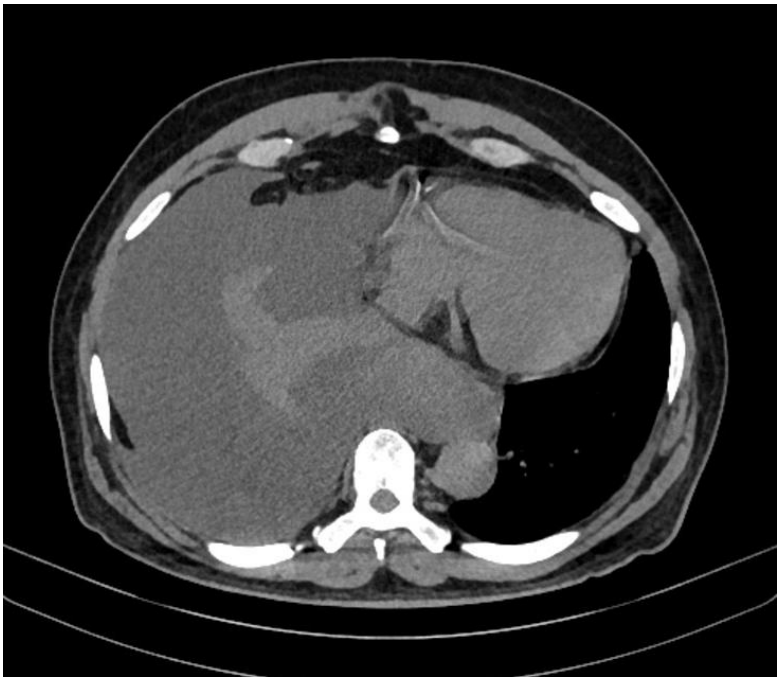

Figure 2: CT Thorax showing right massive effusion

We proceeded with thoracoscopy using Olympus LTF 160 and removed around 3 liters of hemorrhagic fluid (Fig: 3). There were no nodules over parital or diaphragmatic pleura and biopsy was taken from multiple sites from parital pleura. Lung showed multiple bullae of varying size. Bronchoscopy was within normal limits except for the extra luminal compression of right lower lobe. Thoracoscopic pleural biopsy showed chronic inflammation only. Patient improved clinically and intercoastal tube was removed once the drain was less than 100 ml /day and he was sent home with supportive measures.

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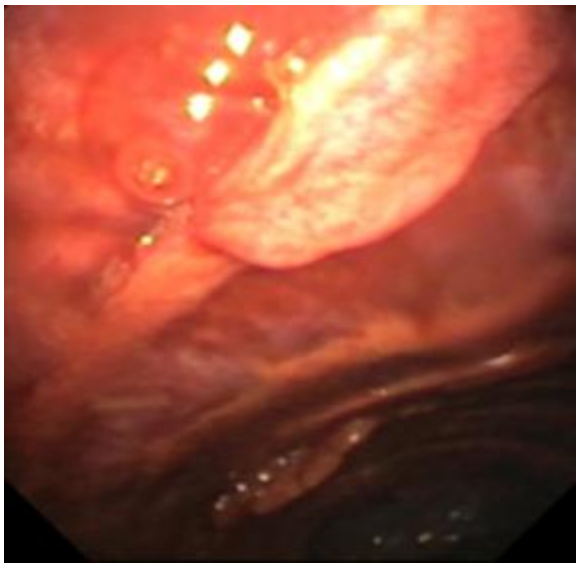

Figure 3: Thoracoscopic appearance of pleura

After 4 weeks, he presented to emergency department with right sided chest pain and increase in dyspnea. Clinical examination and CXR was suggestive of right effusion. CT thorax was repeated and showed right massive effusion with lung collapse with multiple hyper dense clumped areas possibly blood clots/ mass. We proceeded with intercoastal tube drainage and altered blood was drained. In view of reaccumulation of fluid and retained clots in pleural cavity cardiothoracic opinion was taken and decided to plan thoracotomy. Hematoma was evacuated and decortication of right lower lobe was carried out. Soft tissue mass noted in the right costo diaphragmatic region was excised and sent for histology. Post procedure lung expanded well and patient was discharged after removal of ICD.

Histopathological examination showed high grade spindle cell lesion with cellular and necrotic areas. Individual cells showed nuclear atypia and brisk mitosis. Cellular area showed fascicles of spindle cells arranged haphazardly, in storiform and focal herring bone pattern (Fig: 4).

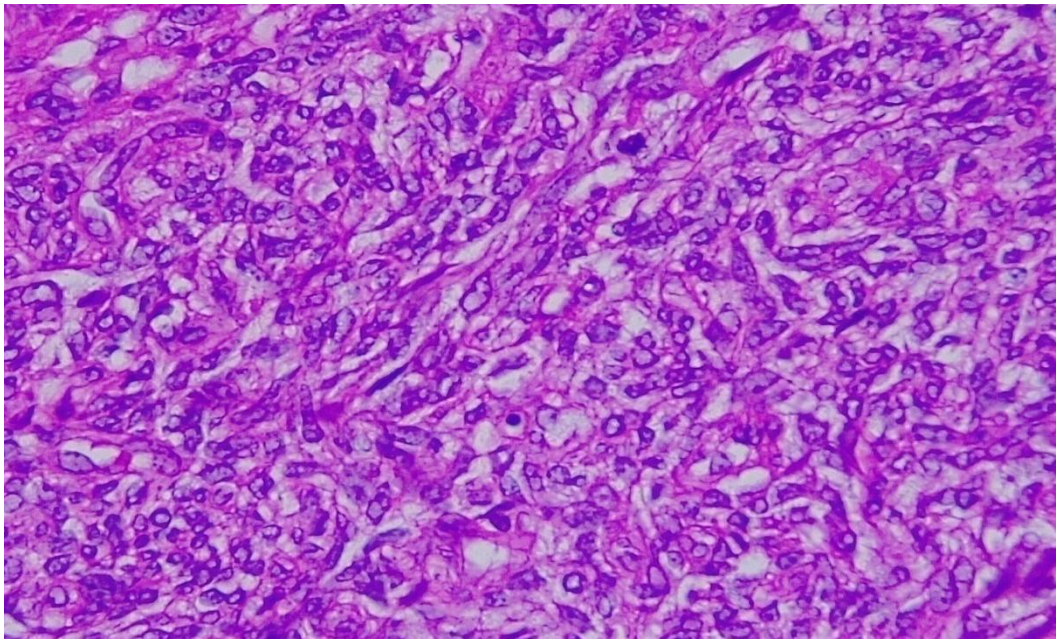

Figure 4: Spindle cells showing nuclear atypia

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These fascicles were separated by dense collagenous stroma. Ki 67 proliferation index was 15%, Immunohistochemistry showed diffuse strong positivity for CD99 (Fig: 5).

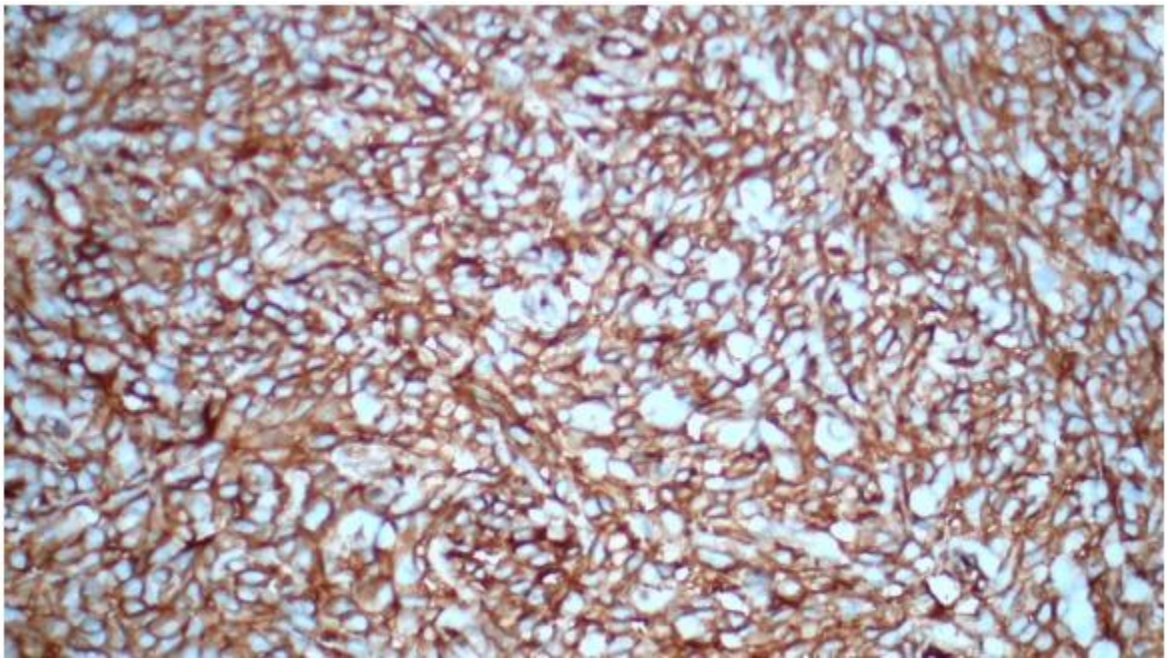

Figure 5: Immunohistochemistry for CD99 showing diffuse positivity in tumor cells

Tumour cells had patchy cytoplasmic positivity for Bcl 2, negative for CD 34 and STAT6. A final diagnosis of Synovial sarcoma of pleura, spindle cell type was made. After multidisciplinary discussion, since primary pleural synovial sarcoma is extremely rare, we decided to proceed with PET scan in search of any primary, which revealed a lytic lesion on his left calcaneum (Fig: 6). The patient was referred to medical oncology for further management.

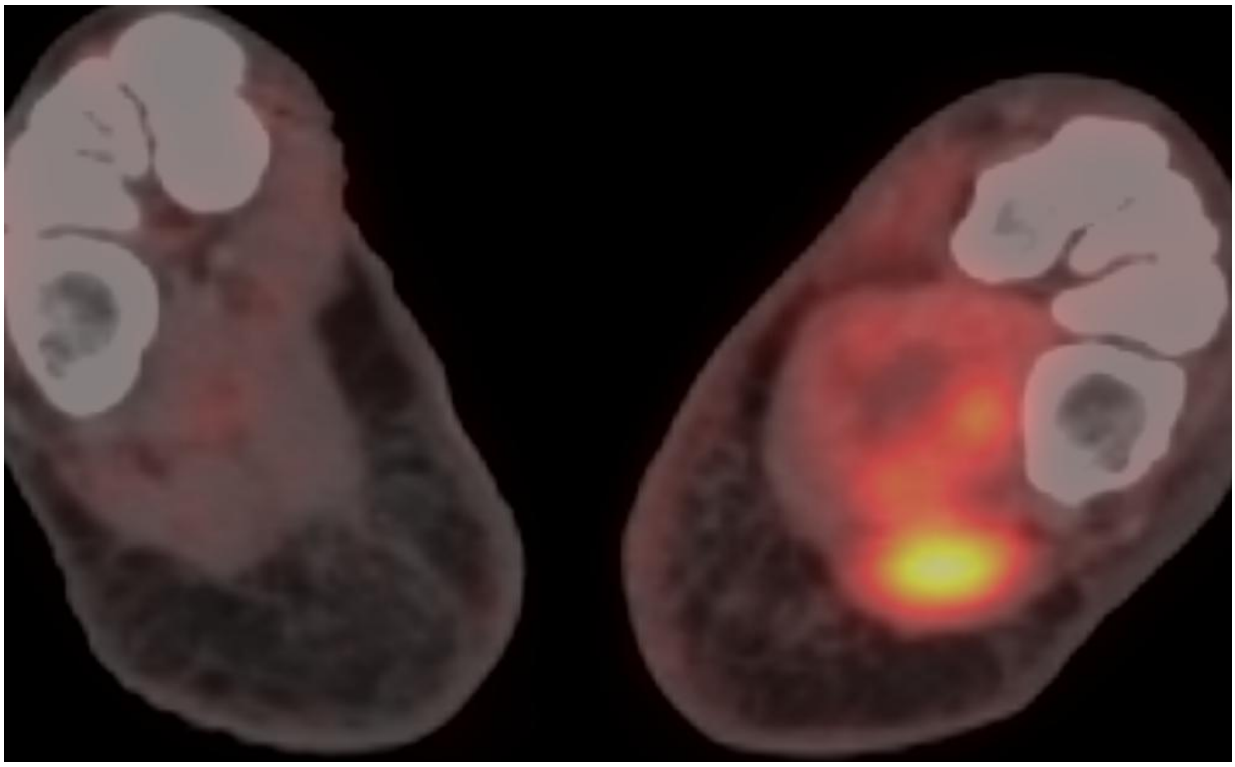

Figure 6: PET scan image showing lytic lesion in left calcaneum

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DISCUSSION

Synovial sarcoma is a malignant neoplasm of soft tissue arising from the pluripotent mesenchymal tissue. It must be differentiated from other spindle cell tumors as the clinical course is highly aggressive[3]. It arises from the extremities in 90% cases and 10% from various other sites[4]. The diagnosis of synovial sarcoma requires combination of clinical, radiological, pathological, and immunohistochemical evaluations to exclude alternative primary tumours[5]. Synovial sarcoma of the extremitiy remaining occult and presenting as exertional dyspnea with massive pleural effusion and pleural based lesions as in our case is uncommon. Primary pleuropulmonary synovial sarcoma is even rarer. The term describes the anatomic subset of primary synovial sarcomas originating from either the lung or from the pleura due to inherent difficulties in assigning a precise anatomic origin in most cases[6]. Primary pleuro pulmonary sarcoma comprises only 0.5% of all primary lung malignancies with only a few case reports in literature[7].

Synovial sarcoma is driven by the chromosomal translocation t(X;18) (p11;q11) involving genes SS18 and either SSX1, SSX2 or SSX4. The usual clinical presentation is in third or fourth decade. In case of pleural involvement, presentation is with exertional dyspnea. Radiologically there can be heterogeneously enhancing pleural based mass or nodules with ipsilateral pleural effusion, while mediastinal lymphadenopathy is rare[8].

Histologically, synovial sarcomas are classified as biphasic or monophasic. Monophasic synovial sarcomas are more common and composed of uniform spindle cells. Biphasic synovial sarcomas contain both epithelial and spindle cell components. Immunohistochemically, synovial sarcomas show focal expression of Cytokeratin and EMA. Furthermore, 30% are protein S-100 positive, 60% - 70% CD99 positive, and 75%-100% are Bcl-2positive[9]. The 5-year survival rate varies from 36% to 76% depending on the patient's age, tumor size, and tumor resectability[10]. Combination chemotherapy consisting of doxorubicin and ifosfamide has been associated with moderate sensitivity in patients with synovial sarcoma. External Beam Radiotherapy is another therapeutic modality used in unrsectable, incompletely debulked synovial sarcoma.

Declaration of patient consent:

The authors certify that they have obtained all patient consent forms. In the form, patient has given his consent for reporting the clinical data and images in the journal. The patient and relatives understand that their names and initials will not be published and due care will be maintained to conceal their identity, but anonymity cannot be guaranteed.

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