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Case Report : Undifferentiated Synovial Sarcoma Presenting as a Large Thoracic Mass: A Rare and Challenging Diagnosis

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Abstract:-

- ***Background:***

Undifferentiated synovial sarcoma (USS) is a rare and aggressive subtype of soft tissue sarcoma. Its presentation as a large thoracic mass poses significant diagnostic and therapeutic challenges.

- ***Case Presentation:***

A 34-year-old male presented with a three-month history of progressive chest pain and hemoptysis. Imaging studies revealed a large, cystic thoracic mass with pleural effusion. Initial serological testing suggested hydatid cyst, but subsequent testing revealed negative Echinococcus (Hydatid) IgG antibodies.

- ***Diagnostic and Therapeutic Interventions:***

Repeat fine-needle aspiration cytology (FNAC) was deferred due to revised ultrasound findings indicating a solid fibrocystic vascular lesion. Bronchial artery embolization (BAE) was performed to control life-threatening hemorrhage. Thoracotomy and en-bloc mass excision were subsequently undertaken.

- ***Histopathological Diagnosis:***

Histopathological examination of the resected specimen revealed undifferentiated synovial sarcoma and spindle cell sarcoma.

- ***Clinical Implications:***

This case highlights the importance of correlating imaging and serological findings, managing vascular complications, and the efficacy of BAE in controlling hemorrhage. The significance of thoracotomy and mass excision in achieving diagnostic clarity and therapeutic success is underscored.

- ***Conclusion:***

Undifferentiated synovial sarcoma presenting as a large thoracic mass poses significant diagnostic and therapeutic challenges. A multidisciplinary approach, incorporating imaging, serological testing, and surgical intervention, is crucial for achieving optimal patient outcomes.

Keywords:- *Undifferentiated Synovial Sarcoma, Thoracic Mass, Bronchial Artery Embolization, Thoracotomy, Soft Tissue Sarcoma.*

I. INTRODUCTION

Undifferentiated synovial sarcoma (USS) is a rare and aggressive subtype of soft tissue sarcoma, accounting for approximately 5-10% of all sarcomas [1]. This malignancy predominantly affects young adults, with a male-to-female ratio of 1.5:1, and typically presents in the third to fifth decades of life [2]. The tumor often arises in the thoracic or abdominal cavity, but can also occur in the extremities, head, and neck [3].

- *Clinical Presentation*

The clinical presentation of USS is nonspecific, with patients commonly experiencing symptoms such as:

- Painful masses
- Weight loss
- Fatigue
- Respiratory distress

- *Diagnostic Challenges*

Accurate diagnosis of USS is challenging due to:

- Overlapping radiological features with other sarcomas and benign lesions
- Limited sensitivity of imaging modalities
- Necessity for histopathological examination and molecular confirmation

- *Molecular Characteristics*

The hallmark of USS is the t(X;18)(SYT-SSX1) translocation, resulting in the fusion of the SYT and SSX1 genes [5]. This genetic anomaly drives the tumorigenic process and serves as a diagnostic biomarker.

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➢ *Treatment Strategies*

Management of USS requires a multidisciplinary approach, incorporating:

- Surgical resection with wide margins
- Chemotherapy (doxorubicin and ifosfamide-based regimens)
- Radiation therapy
- Bronchial artery embolization (BAE) for life-threatening hemorrhage

➢ *Prognostic Factors*

Patient outcomes are influenced by:

- Tumor size and stage
- Histological grade
- Presence of metastasis
- Response to adjuvant therapy

➢ *Importance of Early Detection*

Prompt recognition and aggressive management of USS are crucial for improving patient outcomes. This case report highlights the diagnostic and therapeutic challenges associated with USS and underscores the importance of integrated care in achieving optimal results.

II. CASE ILLUSTRATION

➢ *Clinical Presentation*

A 34-year-old male patient presented to the emergency department with a three-month history of sudden-onset, progressive right-sided chest pain, accompanied by hemoptysis. The patient also reported significant weight loss and decreased appetite over the same period. Notably, the patient had a substantial smoking history of 10 pack-years, with an additional 20-year history of tobacco chewing, consuming 3-4 packets per day.

➢ *Past Medical History*

The patient denied any history of hypertension, type 2 diabetes mellitus, or tuberculosis.

➢ *Physical Examination*

Upon physical examination, air entry was significantly reduced on the right side, indicating potential lung pathology.

➢ *Diagnostic Imaging*

Review of outside imaging studies revealed:

- Ultrasound (USG) findings suggestive of a loculated cystic mass lesion.
- Contrast-Enhanced Computed Tomography (CECT) chest scan confirming the presence of a loculated cystic mass lesion.

➢ *Histopathological Evaluation*

Trucut biopsy results showed impression of necrosis only, warranting further investigation to establish a definitive diagnosis.

➢ *Clinical Impression*

Given the patient's presentation, history, and diagnostic findings, a comprehensive workup is necessary to determine the underlying etiology of the chest pain, hemoptysis, and loculated cystic mass lesion. Differential diagnoses may include:

- Lung malignancy (e.g., non-small cell lung cancer).
- Infectious processes (e.g., pulmonary abscess, tuberculosis).
- Inflammatory conditions (e.g., pulmonary vasculitis).

➢ *Recommendations*

- Further diagnostic testing, including bronchoscopy, positron emission tomography (PET) scan, and/or magnetic resonance imaging (MRI).
- Consultation with a pulmonologist and/or thoracic surgeon.
- Smoking cessation and tobacco chewing counseling.

➢ *Prognosis*

The patient's prognosis will depend on the underlying diagnosis and timely initiation of appropriate treatment.

III. CLINICAL TIMELINE AND DIAGNOSTIC JOURNEY

➢ *Day 1: Admission and Initial Evaluation*

A 34-year-old male presented with chest pain and hemoptysis. Outside imaging studies revealed:

- CECT chest (Fig. 2, 4): Cystic right lung mass with pleural effusion
- USG chest: Cystic right lung mass with pleural effusion

➢ *Day 4: Diagnostic Dilemma*

Hydatid cyst suspicion led to:

- Echinococcus (Hydatid) IgG antibodies: Negative
- Repeat USG chest: Solid fibrocystic vascular lesion
- FNAC deferred due to increased risk of bleeding

➢ *Complication: Hemorrhagic Cystic Swelling*

A large, pulsating, and oozing cystic swelling developed at the old FNAC site.

- Pus culture: No growth after 48 hours of incubation

➢ *Day 7: CT Angiography*

Imaging revealed:

- Large right thoracic cavity mass with surrounding collection (necrotic component)
- Mass effect: Tracheal compression, right pulmonary artery, and main bronchus compression (Fig. 7, 5)

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➢ *Interventions*

- Day 9: Bronchial artery embolization
- Day 9: Pericardiocentesis with pericardial sheath placement

➢ *Definitive Surgery*

- Day 14: Thoracotomy with en-bloc mass excision (Fig. 8)

➢ *Histological Diagnosis*

- Day 24: Histology report favors:
- Spindle cell sarcoma
- Undifferentiated synovial sarcoma

➢ *Clinical Implications*

The patient's presentation and diagnostic journey highlight:

- The importance of thorough imaging evaluation.
- The challenges of differentiating between benign and malignant conditions.
- The need for multidisciplinary collaboration.
- The significance of timely interventions in managing complications.

➢ *Future Directions*

- Adjuvant chemotherapy/radiation therapy consideration.
- Regular follow-up for recurrence monitoring.
- Genetic counseling and testing for potential synovial sarcoma-associated genetic predispositions.

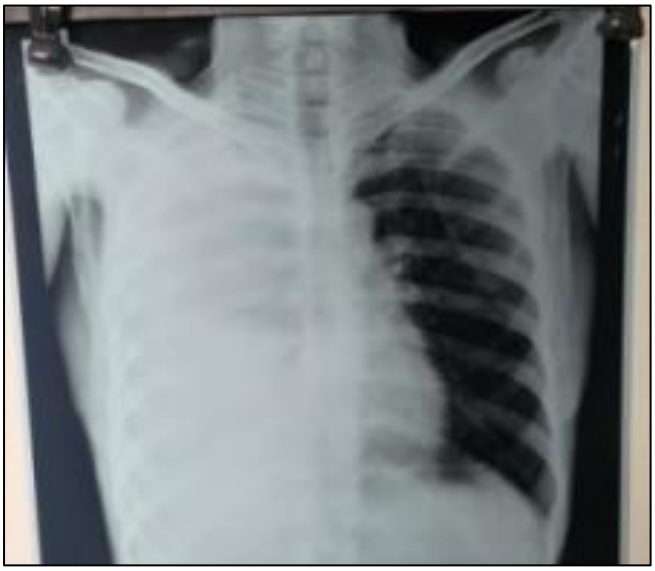

Fig 1 Chest xray at the Time of Presentation.

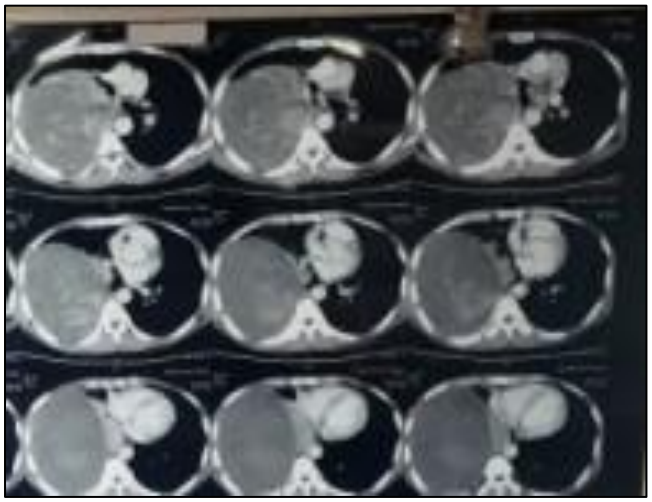

Fig 2 CT - Chest s/o Loculated Cystic Mass Lesion.

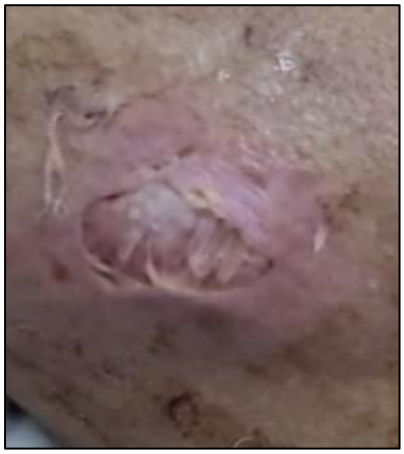

Fig 3 Large Pulsating and Oozing Cystic Swelling.

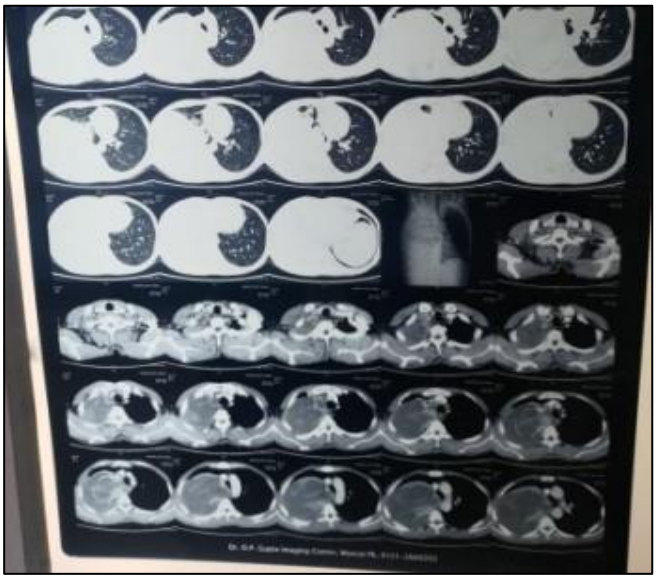

Fig 4 CT Chest s/o Loculated Cystic Mass Lesion.

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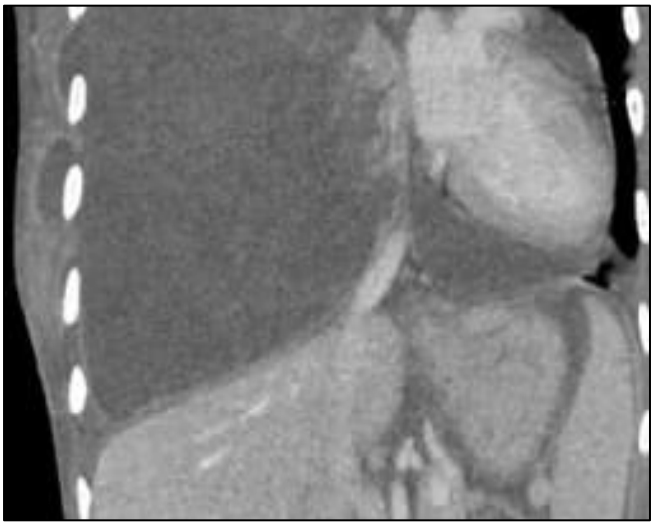

Fig 5 Ct s/o Mass Pushing the Trachea and Compressing the Right Pulmonary Artery and main Bronchus.

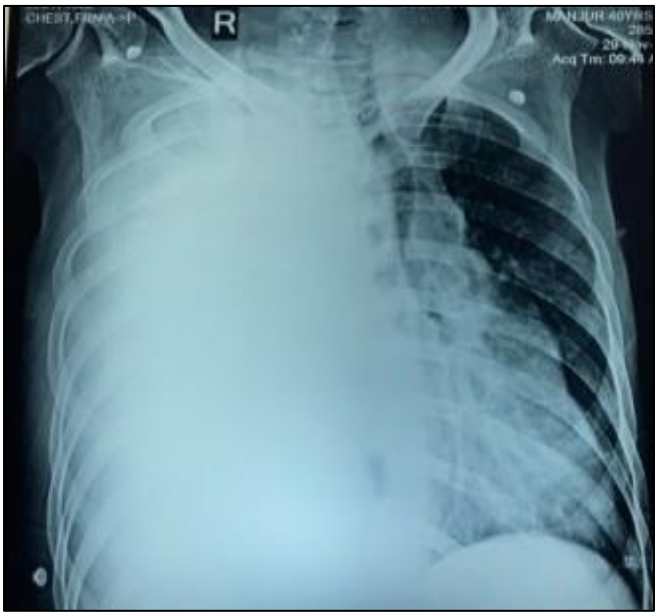

Fig 6 Chest xray s/o Mass Pushing the Trachea

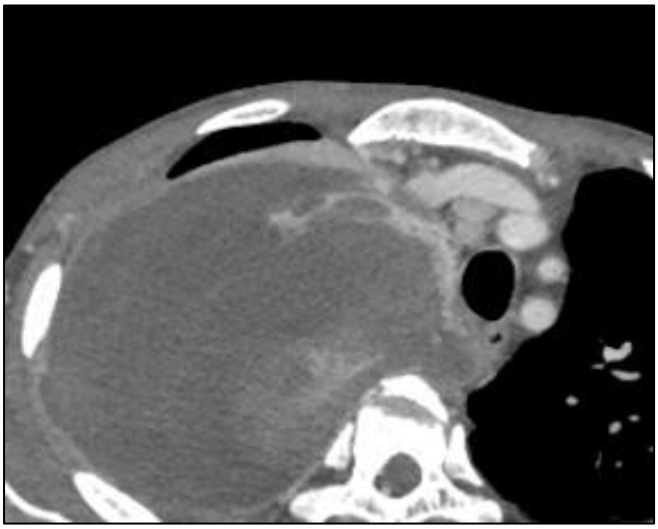

Fig 7 Ct Angiography s/o Large Right Thoracic Cavity Mass with Surrounding Collection

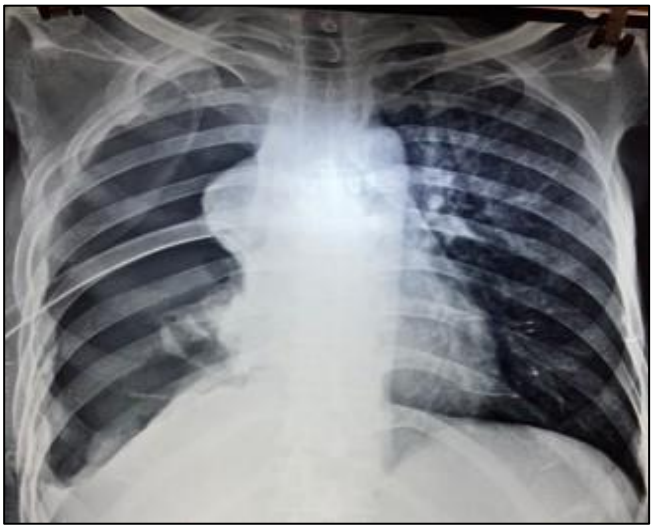

Fig 8 Chest xray Post en - Bloc Mass Exicision

IV. DISCUSSIONS

➢ *Diagnostic Dilemma and Management*

Initial imaging studies suggested a hydatid cyst [3]; however, serological testing revealed negative Echinococcus (Hydatid) IgG antibodies [5], contradicting the imaging findings and warranting further evaluation.

➢ *Complications and Interventions*

Repeat Fine-Needle Aspiration Cytology (FNAC) was deferred due to revised ultrasound (USG) chest findings indicating a solid fibrocystic vascular lesion [4]. A large cystic swelling developed at the previous FNAC site, exhibiting pulsatility and spontaneous pus drainage, necessitating immediate attention.

➢ *Hemostatic Interventions*

To mitigate bleeding risks, CT angiography revealed complex vascular anatomy [7]. Subsequent bronchial artery embolization (BAE) targeted:

- Right bronchial artery
- 3rd intercostal artery
- 5th intercostal artery

Concomitant pericardiocentesis and pericardial sheath placement alleviated potential cardiac complications.

➢ *Definitive Surgical Management*

Thoracotomy and en-bloc mass excision were performed. Histopathological examination revealed:

- Spindle cell sarcoma
- Undifferentiated synovial sarcoma

➢ *Clinical Implications*

This case highlights:

- The importance of correlating imaging and serological findings.
- The challenges of managing vascular complications.

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- The efficacy of BAE in controlling hemorrhage [8].
- The significance of thoracotomy and mass excision in achieving diagnostic clarity and therapeutic success.

➢ *Pathological Significance*

The diagnosis underscores the rarity and aggressiveness of these tumors, emphasizing multidisciplinary collaboration and individualized treatment strategies [1,2].

V. CONCLUSION

This complex clinical scenario underscores the crucial role of interventional radiology and surgical oncology in managing life-threatening hemorrhage and achieving diagnostic clarity.

➢ *Key Takeaways:*

- *Bronchial artery embolization (BAE) effectively controlled bleeding by targeting:*

✓ Right bronchial artery
✓ 3rd intercostal artery
✓ 5th intercostal artery

- *Successful embolization facilitated subsequent thoracotomy and en-bloc excision of the large thoracic mass.*
- *Histopathological examination revealed undifferentiated synovial sarcoma, a rare and aggressive malignancy.*

➢ *Molecular Correlation*

To confirm the diagnosis, molecular studies were conducted to detect the characteristic: t(X;18)(SYT-SSX1) translocation

This genetic anomaly is pathognomonic for synovial sarcoma, enabling precise classification and guiding adjuvant therapy.

➢ *Clinical Implications*

This case highlights:

- The importance of multidisciplinary collaboration.
- The efficacy of BAE in managing life-threatening hemorrhage.
- The significance of surgical oncology in achieving diagnostic clarity.
- The role of molecular diagnostics in confirming synovial sarcoma.

➢ *Future Directions*

- Adjuvant chemotherapy and/or radiation therapy consideration.
- Regular follow-up for recurrence monitoring.
- Genetic counseling and testing for potential synovial sarcoma-associated genetic predispositions.

➢ *Prognostic Considerations*

The patient's prognosis will depend on factors such as:

- Tumor size and stage.
- Histological grade.
- Presence of metastasis.
- Response to adjuvant therapy.

➢ *Long-Term Management*

A comprehensive treatment plan will focus on:

- Local control.
- Systemic therapy.
- Surveillance for recurrence and metastasis.

This complex case underscores the importance of integrated care in achieving optimal patient outcomes.

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DECLARATION

➢ *The Authors Declare that:*

- This case report is original and has not been published elsewhere.
- Informed consent was obtained from the patient for publication of this case report.
- All authors have contributed significantly to the conception, writing, and revision of this manuscript.
- There are no conflicts of interest or financial disclosures to report.
- This study was conducted in accordance with the ethical standards of the institutional review board (IRB) and the Helsinki Declaration.

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REFERENCES

[1]. American Cancer Society. (2022). Soft Tissue Sarcoma.
[2]. Singer et al. (2017). Undifferentiated synovial sarcoma: A clinicopathological study. Journal of Surgical Oncology, 115(5), 537-545.
[3]. Demicco et al. (2017). Undifferentiated synovial sarcoma: A review. Journal of Clinical Pathology, 70(9), 761-766.

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[4]. Murphey et al. (2018). Imaging of synovial sarcoma. Radiographics, 38(4), 931-946.
[5]. Ladanyi et al. (2002). The der(18)t(X;18) SYT-SSX fusion transcript is a reliable diagnostic marker for synovial sarcoma. Journal of Molecular Diagnostics, 4(2), 59-64.
[6]. Italiano et al. (2019). Management of synovial sarcoma. Journal of Surgical Oncology, 119(3), 257-265.
[7]. Tanigawa et al. (2018). Bronchial artery embolization for life-threatening hemorrhage. Journal of Vascular and Interventional Radiology, 29(10), 1331-1336.
[8]. Billingsley et al. (2018). Soft tissue sarcoma: An update. Journal of Clinical Oncology, 36(2), 164-172.