A rare case report of solitary pulmonary metastasis from a synovial sarcoma

Arvind Kumar Bhatt[1,*], Sunil Vyas[2], Gopal Purohit[3], CR Choudhary[4]

[1]Senior Resident, [2]Assistant Professor, [3]Senior Professor, [4]Associate Professor, C. Kamja Nehru Chest Hospital, Dr. SN Medical College, Jodhpur, Rajasthan

***Corresponding Author:**
Email: arvindbhatt3@gmail.com

Abstract

We had reported a 47 years old male patient who presented to us with scanty hemoptysis since five days with chest X-ray showing left upper zone opacity.While investigating for the cause, a 124mm×100mm×147mm sized lesion in left upper lobe was found on contrast enhanced computed tomography and a diagnois of pulmonary metastasis from a synovial sarcoma by histology and immunohistochemistry was made.This case highlights the occurance of solitary metastasis into lung by synovial sarcoma.Such presentations are common in young adults with cough, chest pain, shortness of breath, or hemoptysis, with a mass on X-ray and computerized tomography scan.

Keywords: Metastasis; Malignant; Soft tissue neoplasms; Surgery; Synovial sarcoma

Introduction

Synovial sarcoma was initially described by Simon [1] in 1865 and was termed as synovial sarcoma in 1938 by Sabrazes *et al.* [2] due to its resemblance to the developing synovial tissue under the light microscope. However, it has no demonstrable connection or relationship with the synovium [3]. It is a distinctive soft tissue tumor that displays epithelial differentiation. It is more prevalent in adolescent and young adults, 15-40 years of age. Males are affected more than females, with an average male:female ratio of 1.2:1 [4].

We report this case for two reasons first a secondary synovial carcinoma was not an etiological diagnosis and was unexpectedly diagnosed while fine needle aspiration cytology report of underlying lung opacity revealed some other story which compelled us to do immunohistochemical cytology of the thigh swelling. Generally multiple metastases to lung occurs while solitary metastasis to lung is rare entity. And second due to the fact that sarcomatous involvement of the lung tends to comprise multiple metastases, cases of a single metastasis have been described.

Case Report

A 47 years old male chronic smoker presented to Kamla Nehru Chest Hospital, Jodhpur in Rajasthan, India. He gave the history of blood in sputum, scanty in amount since three days, dull aching chest pain since one month, dry cough with no postural variation since 15 days. He had a history of anorexia and documented weight loss since two months. There was no history of fever, dyspnoea, palpitations, vomiting, or pain abdomen. Also there was no history of diabetes mellitus, tuberculosis, rheumatic heart disease. Family history was inconclusive and there was no history of tuberculosis contact. Patient was a smoker since 10 years, non alcoholic, and driver by occupation.

At the time of admission pulse 78/minute, blood pressure 124/84mm of Hg in supine position. Respiratory rate 20/minute, SpO_2 on pulse oximetry 97% on room air, CVS-S1,S2 normal, with no murmur. General examination did not reveal any abnormality except-a painless swelling having hard consistency, non fluctuating revealed on medial and posterior aspect of left thigh on palpation.

There was no clubbing, cyanosis, icterus, pedal edema and no lymphadenopathy. No raised jugular venous pulse. The physical exam was remarkable only for slight tachypnea and absent breath sounds in the left lung field. Review of other system was unremarkable.

A chest X-ray (Figure.1) in the emergency department showed a large left apical lung mass, left upper zone lung mass with underlying lung collapse confirmed on contrast enhanced computed tomography as a 124mmX100mmX147mm sized lesion in left upper lobe (Figure. 2) with post contrast enhancement of mass in CECT thorax-axial section (Figure.3).

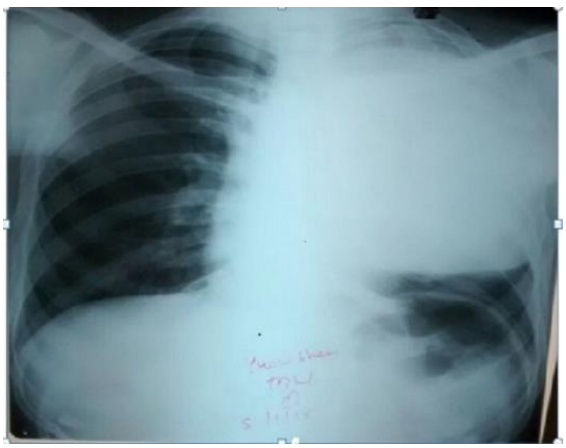

Figure.1: Chest xray showing left upper zone opacity

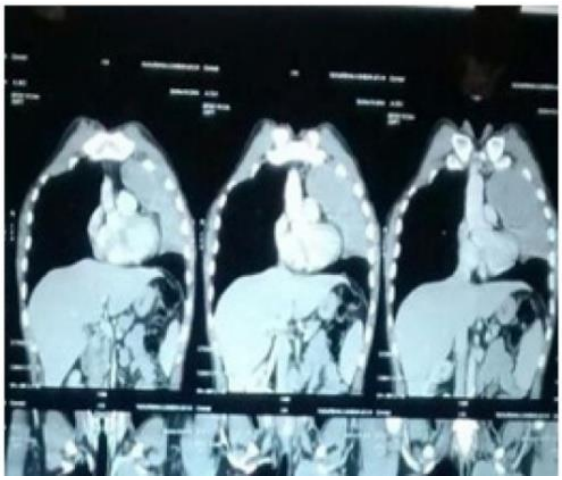

Figure.2: CECT thorax - mass in left upper zone [Sagital section]

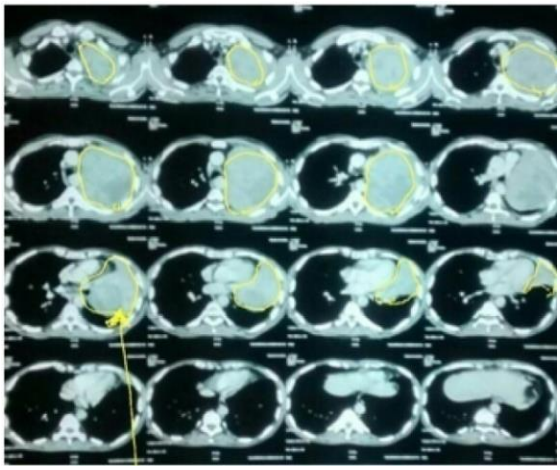

Figure.3: Post contrast enhancement of mass in CECT thorax [Axial section]

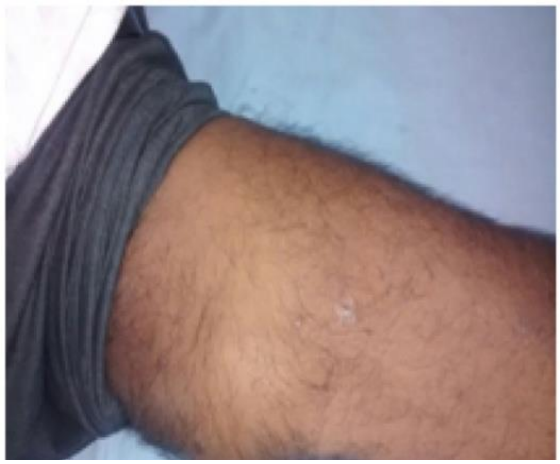

Figure.4: Clinical picture of patient reveals swelling on medial and posterior aspect of left thigh

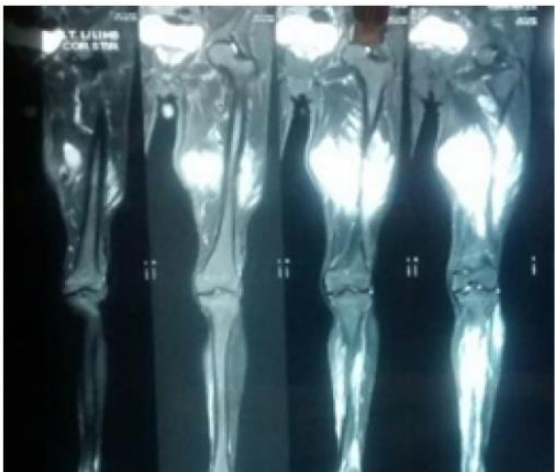

Figure.5: MRI Heterogeneously enhancing multi lobulated predominantly solid large mass lesion occupying the medial and posterior compartment of the left thigh extending from the neck of the femur till the mid thigh and with a track extending to the leg. The mass in the leg occupies the posterior compartment of the leg displacing the muscles anteriorly and posteriorly and extending into the lateral compartment posterior to the femur.Another track from the lower end of the mass extending till the ankle joint. Possibility of soft tissue sarcoma with local spread has to be considered above

USG abdomen was normal. Bronchoscopy was inconclusive. As a 47 years old male smoker with left upper zone opacity in lung, we were suspecting primary lung carcinoma, but USG guided fine needle aspiration cytology report was suggestive of a different story. Final impression of fine needle aspiration cytology was suggestive of non bronchogenic neoplasm.

On careful evaluation a swelling revealed on medial and posterior aspect of thigh. FNAC taken from the thigh swelling report suggestive of soft tissue sarcoma [Figure.4] which was consistent with FNAC from lung so the lesion appears to be a single entity. The lung mass may be considered to be metastatis from legs/thigh. Immunuhistochemistry from both sites was positive for CD[cluster of differentiation] 99, CD 56 Neuroendocrine marker), and CK(Cytokeratin which is epithelial cell marker) which are specific for synovial cell sarcoma, while EMA (Epithelial membrane antigen marker) was focally positive and S-100 (Schwannoma marker) was negative so synovial sarcoma, intermediate to high grade was diagnosed.

Discussion

Soft tissue sarcomas are often slow growing initially; the average duration of symptoms is 2–4 years [6)]. Pain and tenderness at the site of the mass are frequent, and some patients present with pain but no palpable mass [10]. This symptom is unusual compared with other soft-tissue sarcomas that typically manifest

as painless mass [10]. Weight loss and constitutional symptoms are unusual, but when present usually indicate a poorly differentiated tumor [6]. Local trauma may cause hemorrhage in a soft-tissue mass or may bring the mass to the attention of the patient or examining physician.Synovial sarcoma shows dual epithelial and mesenchymal differentiation composed of sheets of spindle cells and sharply segregated epithelial cells [14,15]. Synovial sarcomas are frequently multilobulated, and areas of necrosis, hemorrhage, and cyst formation are also common [16]. Monophasic synovial sarcoma represents 50%–60% (the most common subtype) of all lesions. The poorly differentiated synovial sarcoma represents a form of tumor that can occur in either monophasic or biphasic forms .The diagnosis of synovial sarcoma has been facilitated by the inclusion of molecular biological techniques in pathology and the recent incorporation of cytogenetics in the oncologic examination.Grading of synovial sarcoma is achieved by applying the FNCLCC (French Federation Nationale des Centres de LutteContre le Cancer) scheme, which uses a combined score from three separate parameters, including degree of differentiation, mitotic activity and necrosis [17,18]. Both biphasic and monophasic synovial sarcomas are usually intermediate grade (grade 2/3). Poorly differentiated synovial sarcomas are high-grade tumors.

The presence of keratin (epithelial marker) positivity (approximately 90% of cases), measured with immunohistochemical staining in correlation with the histologic appearance is diagnostic for synovial sarcoma [19,20]. Both the glandular component (diffusely) and the spindle cell component (focally) demonstrate single cells (monophasic and poorly differentiated subtypes) or clusters of cells (biphasic subtype) that are positive for epithelial markers,most notably pankeratins, EMA (epithelial membrane antigen), and CK7 in diagnosis of synovial sarcoma [21]. CD(cluster of differentiation) 99,which is found as a cytoplasmic membrane marker in Ewing and primitive neuroectodermal tumors, can also be positive in synovial sarcoma (62% of cases).

MRI (Magnetic resonance imaging) is the optimal radiologic modality for assessing the extent and intrinsic characteristics of synovial sarcomas [13,22–27]. Synovial sarcoma typically appears as a prominently heterogeneous multilobulated soft-tissue mass [22–27]. With prominent enhancement in synovial sarcomas. The enhancement is more commonly heterogeneous (83%–100% of lesions) than homogeneous (0%–17% of lesions) [12,16,25]. This heterogeneous enhancement reflects the intermixture of nonenhancing necrotic, cystic, or hemorrhagic regions and enhancing solid regions.

A large, single metastasis after long-term disease-free survival is very uncommon. Patients with synovial sarcoma, since it is an intermediate- to high-grade sarcoma, have a 5-year survival rate ranging from 36% to 76%. At 10 years, the survival rate has been reported to range from 20% to 63% [11]. The clinical course of synovial sarcoma is characterized by a high rate of local recurrence and metastatic disease.Local recurrence following resection occurs in 30%–50% of patients, and distant metastasis develops in 41% [11]. The majority of metastases occur within the first 2–5 years after treatment.Metastases are present in 16%–25% of patients at their initial presentation [28,29]. The most frequent metastatic site is the lung, which is affected in 94% of cases, followed by lymph nodes (4%–18%) and bone (8%–11%) [5-9,11,28,29].

Soft tissue sarcomas metastasize within the first two years of diagnosis. The most frequent metastatic site is the lung, which is affected in 94% of cases, followed by lymph nodes (4%–18%) and bone having 8%–11% [5–9,11,28,29]. Complete surgical excision of the tumor, nearby muscle and lymph nodes is the best way of treating this cancer. Depending on the location and size of the tumor, it may be necessary to remove all or part of a limb.Radiation is often used in conjunction with surgery to kill cancer cells. It can be given before surgery in order to shrink a tumor or afterwards to kill any remaining cancer cells. Chemotherapy can be given as a palliative treatment. Surgical management is the mainstay of treatment with chemotherapy and radiation typically used as adjuvant treatment.Although chemotherapy has a positive impact on survival, the prognosis is poor if metastatic disease occurs.All other forms of treatment are considered palliative. Management of isolated solitary or multiple metastases of metastatic soft tissue sarcoma is challenging. Surgery remaining the treatment of choice. Metastasectomy proved to be the choice treatment in the case of pulmonary metastasis.In some studies it is assessed that the impact on survival of three types of resection:minimal by laser or conventional device and lobectomy [30,31].

Conclusions

The solitary pulmonary nodule in this case was indistinguishable from primary pulmonary carcinoma. Evaluation with FNAC, CECT, MRI, IHC which led to the diagnosis of pulmonary metastasis instead of primary lung tumour.

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Conflicts of Interest: None declared

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