Pleural Effusion and Pregnancy: A Masquerading Presentation of a Leiomyosarcoma

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ABSTRACT

Introduction: Sarcomas arise from either soft tissues or bone. Leiomyosarcoma is a type of soft tissue sarcoma. It is an aggressive tumor arising most commonly from uterus, gastrointestinal system and vascular sites like pulmonary artery and inferior vena cava.

Case report: This is a case report of a patient with 20 weeks of gestation presented with fever and rapidly progressive dyspnoea which was not responding to symptomatic management. Examination was suggestive of right pleural effusion which progressed to superior vena cava obstruction during hospital stay. MRI thorax revealed a large soft tissue mass with areas of necrosis with a thin rim of pleural effusion and mediastinal shift to the left side. Histopathology revealed high grade leiomyosarcoma with brisk mitotic activity.

Conclusion: Leimyosarcoma in our patient had an initial presentation similar to pleural effusion which was eventually diagnosed after invasive procedure though its site of origin could not be determined. Thus we are reporting this case due to the rarity of incidence, atypical initial presentation, age of onset, site, possible relationship with estrogenic stimulation (pregnancy) and the rapidity with which it progressed.

Keywords- Leiomyosarcoma, Mediastinal tumor, Pregnancy, Superior Vena Cava Obstruction

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Conflict of interest: None

INTRODUCTION

Sarcomas account for less than 1% of all solid malignancies in adults [1]. Sarcomas are normally found in soft tissues and bones. Of the soft tissue sarcomas (STS), approximately 2-9% are leiomyosarcoma[2]. It is an aggressive STS, derived mainly from the smooth muscle cells of the uterine, gastrointestinal or vascular system. The occurrence in mediastinum is rare. Mean age of diagnosis of STS is 58 years [3] with young adults experiencing the lowest

incidence of STS[4]. It is more common in women than in men[5]. Incidence of malignancy in pregnancy is infrequent with incidence rate of 0.07% to 0.1% of all pregnancies [6]. The role and influence of pregnancy in the development and progression of STS including leiomyosarcoma is controversial and not known.

We are reporting a case of leiomyosarcoma of unknown origin within right mediastinum in a pregnant female who presented with rapidly progressing dyspnoea.

CASE REPORT

A 26 year old married female with 20 weeks of gestation, presented to our ER with progressive shortness of breath associated with intermittent fever and dry cough over last 15 days. She also complained of right sided pleuritic chest pain which resolved over 2 days. Patient did not complain of hemoptysis, orthopnea and wheezing. There was no significant past medical history. Prior to admission she had visited a primary care physician and was prescribed a course of antibiotics. However there was no response to therapy. The patient was referred to our hospital for increasing dyspnoea.

On examination the patient was febrile, with a pulse rate of 136/minute, regular with all other peripheral pulses normal. Patient had respiratory distress with a respiratory rate of 30/minute and SpO2 of 97% while breathing ambient air. She had pallor without lymphadenopathy. Other general examination findings were unremarkable. On examination of the respiratory system, there was an upper and lower mediastinal shift to left side with a stony dull note on percussion and absence of breath sounds in the right side of the chest. Other system examination revealed no abnormality. Provisional diagnosis of pleural effusion was considered and patient was given symptomatic management.

On day 1 investigation revealed hemoglobin 8.5 gm/dl, normocytic normochromic anemia, with low corrected reticulocyte count. Total leucocyte count was 13600/cumm with 78% polymorphs. The Erythrocyte sedimentation rate was 100mm in 1sthr with normal CRP levels. The liver function tests, renal function tests and metabolic parameters were normal. Viral serology was negative. Electrocardiogram showed sinus tachycardia. Ultrasound for fetoplacental profile showed a single intrauterine live

fetus of 16 weeks gestation with no liquor. Therapeutic thoracocentesis was done because of increasing dysponea. Pleural fluid study revealed an exudative pleural effusion, mononuclear lymphocytosis with low adenosine deaminase level. Aerobic culture for bacteria, DNA PCR assay for tuberculosis and Pap smear was negative. Parenteral antibiotics were started.

However, the patient continued to have fever with rapid progression of dysponea. On day 4 of hospital stay, she complained of facial flushing and puffiness. Examination revealed pulse rate of 140/min, respiratory rate 40/min, and engorged non pulsatile jugular vein without any signs of cardiac involvement. At this stage the provisional diagnosis was reviewed and the possibility of superior vena cava (SVC) obstruction was considered.

A two dimension echocardiography revealed external right atrial wall compression without any other endocardial, myocardial or pericardial disease. As the patient was in gestation, chest radiograph or CECT thorax could not be done; patient underwent a screening USG of thorax. It revealed a heterogeneous mass in right hemithorax with SVC compression and multiple satellite lesions involving the collapsed right lung.

To further delineate the mass, MRI thorax (T2 and STIR) was done [figure 1a and 1b]. It revealed a large soft tissue mass (169 mm x 181mm x 228 mm in anteroposterior, lateral and craniocaudal dimension respectively) occupying almost complete right hemithorax with extension in mediastinal region and on the left side. There were small areas of necrosis seen. A thin rim of right sided septated pleural effusion was also noted.

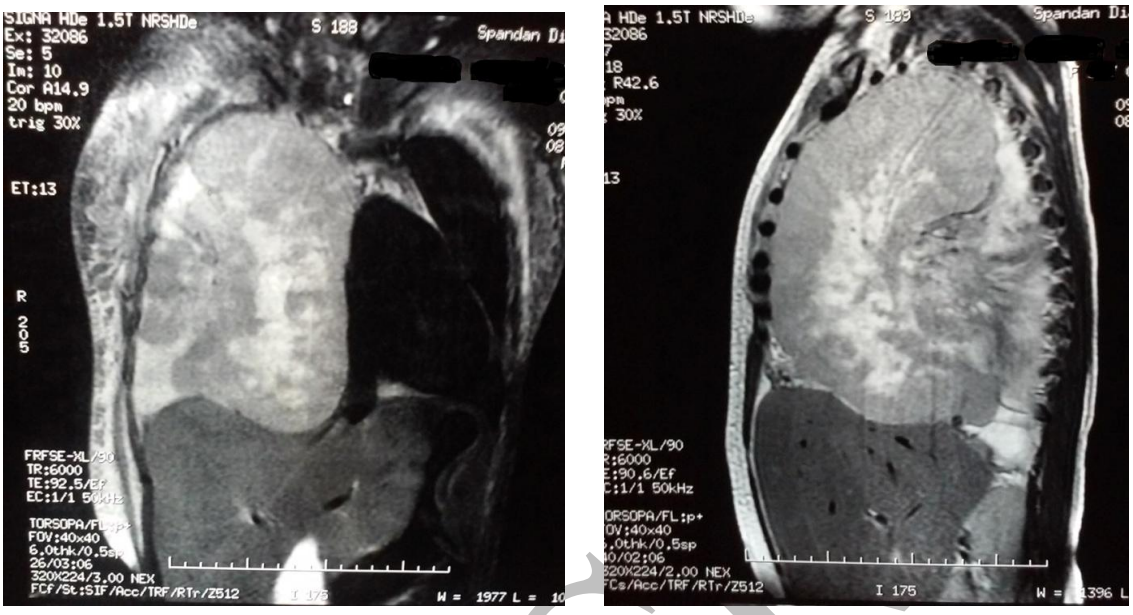

Figure1a and 1b: MRI showing a large soft tissue mass (169 mm x 181mm x 228 mm in anteroposterior, lateral and craniocaudal dimension respectively), occupying almost complete right hemithorax, with extension in mediastinal region and on the left side. There were small areas of necrosis seen.

Serology for common tumor markers revealed serum alpha-feto protein-176.03 ng/ml, serum βHCG -51751mIU/ml, both of which were raised but within the normal limits in pregnancy. Serum LDH was 592 U/ml.

A real time ultrasound guided core biopsy from the mass was undertaken [Figure 2]. Histopathology of the specimen [Figure 3] showed a tumor composed of spindle shaped cells with plump nuclei with pleomorphism and brisk mitotic activity. The tumor expressed EMA, desmin, SMA, h-Caldesmon and was negative for cytokeratin, c-kit, CD 34, S-100 protein, PgR, calretinin, WT-1, HMB 45 and Melan A; suggestive of leiomyosarcoma.

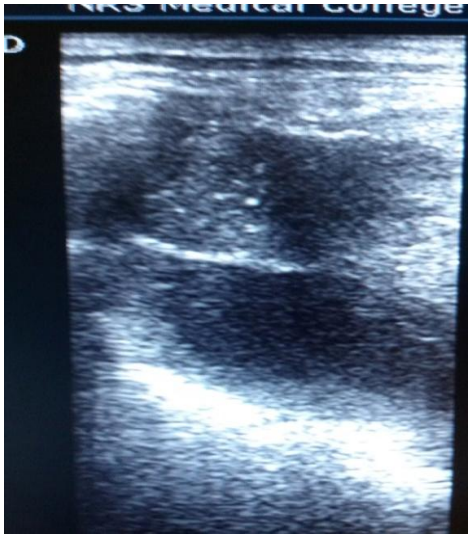

Figure 2: USG guided biopsy being taken from the mediastinal mass

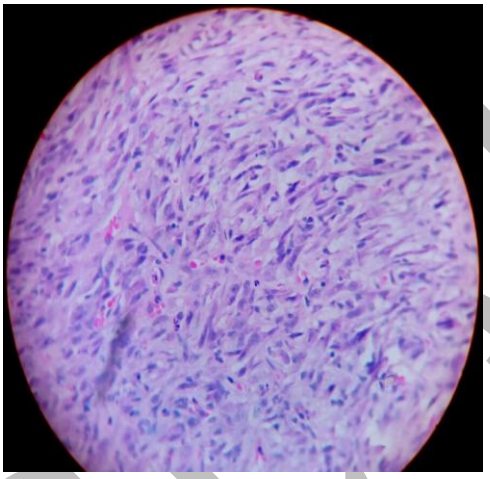

Figure 3: Spindle shaped cells with plump nuclei with pleomorphism and brisk mitotic activity.

A consult from an oncologist, cardio-thoracic surgeon and obstetrician was taken. Neoadjuvant chemotherapy with surgical resection, if feasible, was planned [6]. After written consent from the patient and her husband, a course of chemotherapy with gemcetabine (900mg/m^2) and docetaxel (100mg/m^2) was administered. After the first cycle of chemotherapy the symptoms of the SVC syndrome subsided. She delivered a fresh stillborn male fetus two days after the chemotherapy. The shortness of breath improved with respiratory rate of 20/minute after 4 days of the first cycle. Patient was discharged in a stable state for follow up after a week for the next cycle of chemotherapy. However the patient succumbed to the disease, just after the next admission, within 45 days of onset of symptoms.

DISCUSSION

Several types of soft tissue sarcomas, occurring anywhere in the body, have been reported in association with pregnancy. These include liposarcoma, rhabdomyosarcoma, fibrosarcoma, synovial sarcoma, neurosarcoma, hemangiopericytoma, leiomyosarcoma, and unclassified types. Several tumors may show growth acceleration during pregnancy, among these are leiomyomatosis peritonealis disseminate, dermatofibrosarcoma protuberans, malignant fibrous histiocytoma, liposarcoma, malignant melanoma and meningiomas. Osteosarcoma may also be influenced by hormones during gestation, as

suggested by Huvos et al. [6]. However the relationship of leiomyosarcoma with pregnancy is not known. A case control study done in Northern Italy investigated the potential role of various female hormones and related factors. The only suggestive association was for woman who had become pregnant with her first child beyond the age of 29 years [7].

Leiomyosarcoma (LMS) is a tumor composed of cells showing smooth muscle histology. Although all LMS have similar histology, it is classified into three types, most common being LMS of deep soft tissue, cutaneous LMS which has the best prognosis, and rarest of all is LMS of vascular origin. Grading of this tumor is difficult, and mitotic activity appears to be the best indicator of prognosis when combined with location and size of the tumor [1]. It is an aggressive tumor, with rapid progression and minimal symptoms. The 5- year survival rate in stages I and II is 80% and 67 %, respectively, and 12% to 50% in more advanced stages [8].

Truncal LMS comprises of less than 10% of all cases [5]. It arises mainly from esophagus, pericardium and smooth muscle cells of large and small blood vessels [6]. In this patient, the exact origin of the leiomyosarcoma could not be discerned. However, pericardium was compressed and was free of encroachment with esophagus being pushed by the mass. The probability of these structures as the source is highly unlikely, raising the possibility of vascular structures as the origin.

In management of pregnancy related soft tissue sarcoma, there are no strict guidelines. In each case, the therapy is to be tailored in accordance with the curability of mother's disease. This patient had a high grade tumor, massive in size which was locally advanced. It was treated with chemotherapy with initial response followed by a downhill course.

CONCLUSION

Leimyosarcoma in our patient had an initial presentation similar to pleural effusion which was eventually diagnosed after invasive procedure though its site of origin could not be determined. Thus we are reporting this case due to the rarity of incidence, atypical initial presentation, age of onset, site, possible relationship with estrogenic stimulation (pregnancy) and the rapidity with which it progressed.

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