

# PLEOMORPHIC LIPOSARCOMA PRESENTING AS EXTRA-CARDIAC MEDIASTINAL MASS LEADING TO CARDIAC COMPRESSION

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## ABSTRACT

A 45-year-old male presented with a 3-months history of fever associated with dyspnea, orthopnea, weight loss, chest pain, and anorexia. Past history was notable for cigarette smoking of 5 packs per year. Examination findings suggested clubbing, raised jugular venous pulse and pedal edema. Characteristic findings on systemic examination were displaced apex beat, sternal heave, bibasilar crepitations, distended abdomen, and the presence of shifting dullness. Echocardiography revealed an extracardiac mass collapsing mostly the right side of the heart. Later, multidetector computed tomography scan (MDCT) was done which detected a mediastinal mass crossing midline, significantly compressing right sided cardiac chambers, adjacent segments of major arteries and veins, and displaced esophagus to the left. An ultrasound guided biopsy confirmed the diagnosis of pleomorphic liposarcoma. The objective of reporting this case of pleomorphic liposarcoma is to keep our differentials broad when we get cases with typical cardiac symptoms. Sometimes the etiology could be extracardiac.

**Key words:** Pleomorphic liposarcoma; Mediastinal mass; Extracardiac mass.

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## INTRODUCTION

Liposarcomas are the malignant tumors of mesenchymal origin, more common in extremities and retroperitoneum, and rarely arise in mediastinum. People aged 20-70 years are most commonly affected with a peak incidence in the fifth decade of life. They are classified based on histological appearance into well differentiated, dedifferentiated, myxoid (common in children), and pleomorphic. Pleomorphic sarcoma is characterized by a varying number of pleomorphic lipoblasts merged in the background of undifferentiated sarcomatous areas<sup>1,2</sup>. It has a prevalence of 5-10% of all of the liposarcomas<sup>3</sup>, and 1-2% of all mediastinal tumors. They are the most malignant and are initially ignored but signs and symptoms of compression or invasion usually follow later as their size increases. Symptoms vary depending upon the location of tumor<sup>4</sup>. Herein, we describe a mediastinal tumor that mimics the signs of right sided heart failure e.g., edema, raised jugular venous pressure (JVP), breathlessness, distended veins, ascites, and pleural effusion. The tumor is difficult to diagnose on imaging and histopathology due to low adipose

tissue content. The key to accurate diagnosis is taking adequate tissue samples from adipose and non-adipose tissues. Our patient presented with symptoms attributable to cardiovascular involvement and nonspecific constitutional signs and symptoms, and an extensive workup was done to reach the final diagnosis.

## CASE REPORT

A 45-year-old normoglycemic and normotensive male presented to the medical outpatients with a 3-months history of fever associated with rigors and chills. He also complained of shortness of breath on daily activities. He had lost about 8 kg weight and his weight was down from 74 kg to 66 kg during this period. There was also left sided chest pain and anorexia. Personal history was positive for cigarette smoking of 5 packs per year in the past and disturbed sleep due to orthopnea. He denied any changes in bowel and bladder habits. The past medical history, past surgical history and family history were not significant. On physical examination, he looked quite debilitated. He had a blood pressure of 90/60 mm Hg, pulse 90 beats/minute, temperature 98 F, and oxygen saturation of 96 %. Positive findings on examination included anemia, clubbing involving both fingers and toes, raised JVP, distended neck veins (see *Figure 1*), positive hepatojugular reflux, and pedal edema. No cyanosis and lymphadenopathy were appreciated. On chest inspection, there were intercostal

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recessions, and the apex beat was visibly displaced and located along the anterior axillary line in the 6<sup>th</sup> intercostals space. The sternal heave was positive on palpation. A pleuropericardial rub was audible on auscultation. Respiratory examination revealed a central trachea and bibasilar fine crepitations although heart sounds could be heard in lower chest zones when auscultated from the back. The abdominal examination demonstrated a distended abdomen, and positive shifting dullness. There was no hepatosplenomegaly. Central Nervous System examination was generally normal. Baseline investigations were sent to laboratory and we had the following results (see Table 1)

Abdominal and Pelvic ultrasound showed mild to moderate ascites and mild left sided pleural effusion. Kidneys on both sides were mildly echogenic. There was a predominantly echogenic lesion seen in epigastrium measuring 11 x 12 cm with no vascularity. On echocardiography, left atrium, left ventricle and right atrium were normal in size but right side was collapsed by an extra-cardiac mass especially at the right atrium and tricuspid valve level. Tricuspid valve was compressed giving a 12mmHg pressure gradient while other valves were normal. Extra-cardiac mass was collapsing mostly the right side of the heart. Inferior vena cava (IVC) was enlarged without respiratory variation, and there was sluggish flow across it. Moderate mitral regurgitation (MR), mild tricuspid regurgitation (TR) and pulmonary hypertension were noted. Left ventricular systolic functions were good and there was a trace aortic regurgitation (AR).

On multidetector computed tomography (MDCT), there was a large, mainly anterior and mid mediastinal right sided heterogenous attenuation mass crossing mid-line, extending to the left involving pericardium and significantly compressing right cardiac chambers, adjacent segments of superior vena cava, IVC, descending thoracic aorta, left atrium, right sided pulmonary veins, and also compressed and displaced esophagus to the left (see Fig 2). There was minimal pleural effusion, and left lower lung ground had glass haze. A small sub centimeter hypodense focus in the left hepatic lobe was found, which was indeterminate on a single phase study. Prominent mildly enlarged mediastinal nodes at sub-carinal aortopulmonary window and perivascular compartment were also noticed. Moreover, limited abdominal sections showed ascites and edematous liver.

Finally, an ultrasound guided biopsy of the mass was carried out that showed cores of fibro adipose muscular tissue showing a high grade sarcoma interspersed with

multinucleated lipoblasts, and 10-20 mitoses were seen in a single focus. A diagnosis of pleomorphic sarcoma was made based on the report. Table 2 enlists the common pathological lesions found in different locations in the mediastinum. A cardiovascular surgeon was involved keeping in mind the clinical picture and after interpreting the imaging scan and biopsy findings. The tumor could not be excised due to the extensive nature of the procedure involved; therefore, the patient was referred to oncology unit for chemotherapy.

## DISCUSSION

Liposarcomas are the malignant adipocytic tumours of mesenchymal origin and mediastinum is an uncommon site of such tumours. In the mediastinum, it is thought to arise from adult thymus or from mesenchymal cells capable of lipogenesis. WHO in 2002 classified liposarcomas based on histological types such as well differentiated, dedifferentiated, myxoid (common in children), pleomorphic, and mixed type. five to ten percent of all of the liposarcomas and 1-2 % of all mediastinal tumors are pleomorphic liposarcomas. They are the most malignant and contain variable number of pleomorphic lipoblasts. Areas of lipogenesis differ in different tumours, and sometimes pleomorphic lipoblasts are very scanty in number making it difficult to reach the diagnosis<sup>5</sup>. Prognosis is worse if centrally located, big in size (>10cm), and with high mitotic index<sup>6</sup>.

**Table 1: Baseline investigations of patient.**

Hb	12.23g/dl (11-16g/dl)
WBC count	11,200/cmm
Platelets	389,000/cmm
MCV	76fl
Total Bilirubin	0.57mg/dl
ALT	35.8U/l
ALP	257U/l
Creatinine	0.53mg/dl
Serum albumin	2.43mg/dl
ESR	80mm/1st hour
INR	1.3
PT	4 seconds prolonged
APTT	6 seconds prolonged
Serum Electrolytes	WNL
HBsAg (ELISA)	Non-reactive
Anti-HCV (ELISA)	Non-reactive

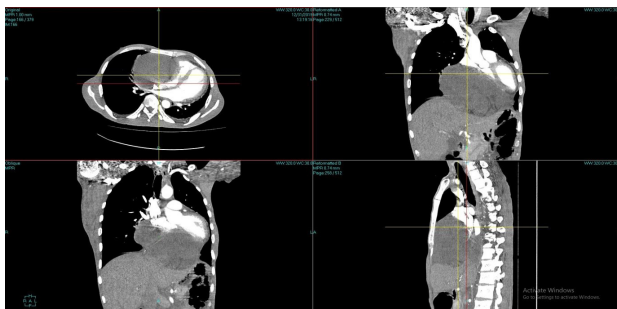
Hb (Hemoglobin), WBC count (White blood cell count), MCV (Mean Corpuscular Volume), ALT (Alanine Transaminase), ALP (Alkaline Phosphatase), ESR (Erythrocyte Sedimentation Rate), INR (International Normalized Ratio), PT (Prothrombin Time) APTT (Activated Partial Thromboplastin Time), WNL (within normal limits), HBsAg (Hepatitis B surface antigen), anti HCV (anti Hepatitis C Virus)

**Table 1: Differential diagnoses of a mediastinal mass**

	<b>Lesions</b>	<b>Fluid</b>	<b>Fat</b>	<b>Vascular</b>
Anterior	Thymic Lymphoma Germ Cell Goiter	Thymic Cyst Thymoma Pericardial Cyst Germ Cell Lymphoma	Germ cell-b Thymolipo- ma Fat pad	Thyroid Cardiac Coro- nary arteries
Middle	Lymph nodes Du- plication Cyst Arch anomaly	Duplication Cyst Necrotic nodes Pericardial recess Retroperitoneal	Lipoma Esophageal Fibro Vascular polyp	Arch anomaly Azygous vein Vascular nodes
Posterior	Neurogenic Bone and marrow	Neuroenteric Cyst Schwannoma Menin- gocele	Extramedullary Hemato- poiesis	Descending aorta
>1 com- partment	Infection Hemorrhage Lung Cancer	Lymphangioma Mediastinitis	Liposarcoma	Hemangioma



**Fig 1: Distended neck veins and raised JVP**



**Fig 1: Distended neck veins and raised JVP**

Pleomorphic liposarcoma is asymptomatic at the early stage, but compression and visceral invasion develops as the tumor grows. Tumours located in mediastinum present as edema, raised JVP, breathlessness, distended neck veins, ascites and pleural effusion. Pleomorphic liposarcomas recur and metastasize frequently. The five-year survival rate is reported as 60%<sup>5</sup>. Imaging such as CT/MDCT, Magnetic Resonance Imaging (MRI), and ultrasound are important before surgery to stage and grade the tumor<sup>4</sup>. Morphologically, these tumours are defined by the presence of multivacuolated pleomorphic lipoblasts, and immunostaining usually identifies S100 protein as the major protein in lipoblasts. Molecular features comprise of complicated karyotypes, and commonly TP53 and RB1 gene inactivation. To confirm the diagnosis, biopsy samples are taken from adipose and non-adipose tissues.

Complete surgical excision is the mainstay of treatment. Tumours that are unresectable are debulked to relieve symptoms. Recurrence of deep seeded mass is very common after surgery so radiotherapy is also given in some cases<sup>7</sup>. In certain aggressive cases, adjuvant chemotherapy is employed in addition to surgery. However, there is no consensus on the use of routine radiotherapy and chemotherapy, and proper guidelines are not available to date<sup>8</sup>.

## CONCLUSION

It can thus be concluded that not all patients presenting with symptoms attributable to involvement of cardiovascular system actually have cardiac conditions. Rarely extra cardiac masses can also manifest as dyspnea and chest pain. Advanced investigations and involvement of interventional radiologists for image guided biopsy help in reaching a diagnosis in such cases.

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