PRIMARY PERICARDIAL LIPOSARCOMA IN A YOUNG LADY

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ABSTRACT

Liposarcomas are tumors that appear to arise from precursors of adipocytes and are most commonly found in the extremities and retroperitoneum. They can metastasize to pericardium, but a primary liposarcoma in pericardium is very rare and generally has an insidious onset and often presents with complications. Treatment is primarily surgical; doxorubicin has a limited role in treatment. Now newer receptor directed therapies have shown benefit in liposarcomas. Primary pericardial liposarcoma due to its rarity is best treated at a high volume center. Here we present to our colleagues a case of primary pericardial liposarcoma in a young lady; whom we referred to a tertiary care center after diagnosis for optimum care.

Keywords: Liposarcoma, Pericardial mass, Primary pericardial tumour.

INTRODUCTION

While liposarcomas are commonly encountered tumors in adults, but a primary mediastinal liposarcoma is a very rare pathology which often tends to remain asymptomatic till it reaches a considerable size causing symptoms related to local invasion and pressure effects. Tumors of primary cardiac origin represent a rare occurrence themselves, an incidence of 0.17-0.19% is reported at autopsy; and only 10-19% of these tumors of the heart and pericardium are liposarcomas[1]. On the other hand, secondary neoplastic involvement of the pericardium is much more prevalent.

CASE REPORT

A 32 year old African-American female presented to the hospital with left sided sharp, non-radiating, pleuritic chest pain for 2 days but denied any aggravating or relieving factors. She complained of progressively worsening dyspnea for several weeks. No history of cough, fever, swelling or mass of the extremity. She did admits 15 pounds of unintentional weight loss in last one month. There was no significant past medical, surgical or social history.

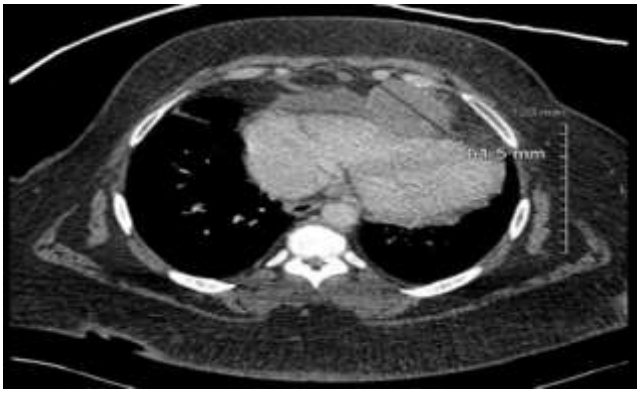

Figure-1: CT chest transverse section showing dominant pericardial mass.

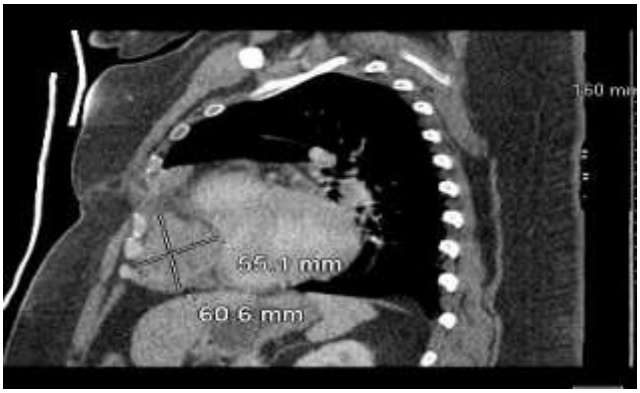

Figure-2: CT chest sagittal section showing dominant pericardial mass.

Blood complete examination revealed WBC count of 13.8 x 1012/dl, CMP, EKG and troponins were normal. She underwent CTA chest which

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revealed multiple epicardial masses, largest of which measured 6 x 5.5 x 6.1cm (fig-1 & 2), constrictive pericarditis was also suspected. Echocardiogram was done which revealed thickened and nodular pericardium with moderate to large size pericardial effusion without cardiac tamponade. Additional testing including CT head with contrast, CT abdomen/ pelvis with contrast and coronary angiogram were unremarkable. She underwent pericardial mass biopsy by interventional radiologist which revealed fibrous tissue but the sample was considered sub-optimal; this was then followed by mini-thoracotomy with pericardial mass biopsy along with random pericardial biopsy. Mass and pericardium both revealed well differentiated liposarcoma which was CDK 4 positive and ALK rearrangement was not seen. After detailed discussion at tumor board for treatment plan, decision was made to refer patient to a high-volume center for resection and evaluation for chemotherapy.

DISCUSSION

Primary Liposarcoma of pericardium is extremely rare. It has insidious presentation which leads to delayed diagnosis, and tends to present with local complications of the tumor. Transthoracic echocardiography is a useful initial investigation for detection and diagnosis of cardiac tumors including liposarcomas. CT scan evaluation also provide more details into morphology and extent of tumor. However, MRI is the optimum test especially for pre-op evaluation and planning, because it provides information into vascular invasion of the tumor[2], because of the great variety of histological presentations, it can sometimes be misdiagnosed. In our case initial biopsy only revealed fibrous tissue. Therefore, to make an appropriate histo pathological diagnosis adequate samples, optimum preparation and careful observation are essential[3]. Traditionally liposarcomas are treated with radical surgery due to being resistant to chemotherapy and radiation treatments; only doxorubicin has shown some benefit in past[4]. Since 90% of these tumors have CDK4 amplification, in the recent years Palbociclib (CDK4/CDK6 inhibitor) has shown benefit in progression of free survival in liposarcoma patients[5]. Unfortunately, due to stage at which many of these patients present, radical surgery is often difficult. Main cause of death is local recurrence peaking at sixth postoperative month on average. But fatal recurrence 5 to 10 years after the initial excision can also be seen[6].

CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

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