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Case Report

Primary Left Atrial Leiomyosarcoma; An Elusive Tumor Revisited

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Introduction:

Primary cardiac tumors are exceedingly rare with an incidence of 0.0017 to 0.33 % of all cardiac tumors according to surgical and autopsy reports. Metastatic tumors from other primary sources are 30 times more common. Only 25% of all primary tumors are malignant and sarcomas constitute 75% of them.

Left atrial leiomyosarcoma is extremely rare. It is elusive as it masquerades as other more common tumor such as myxoma. Multimodality imaging with echocardiography and MRI can be very helpful in identifying its location, its morphology, relation to valves such as mitral in our case and relation to adjacent cardiac structures such as pulmonary veins.

In our presentation of this case report we intend to take the reader on a journey of varied facets of this extremely rare entity. In the process, we review the literature and embark on a learning experience specially with regard to multimodality imaging for all concerned in the care of this patient.

Case Report:

49 years old female without any past medical history of cough and shortness of breath for past few months came to ER with increasing dyspnea for last couple of days on 6/20/19. CT chest showed 4.5x5.2x5.4 cm mass within left atrium obscuring the orifice of left inferior pulmonary vein. Findings could be related to thrombus or neoplasm. Echocardiogram (TTE, TEE) showed a large mass in LA (video clips 1, 2) causing severe MS (Fig 1) and severe pulmonary hypertension. Patient went into acute pulmonary edema while waiting in ER. She underwent emergency surgical procedure on 6/24/19.

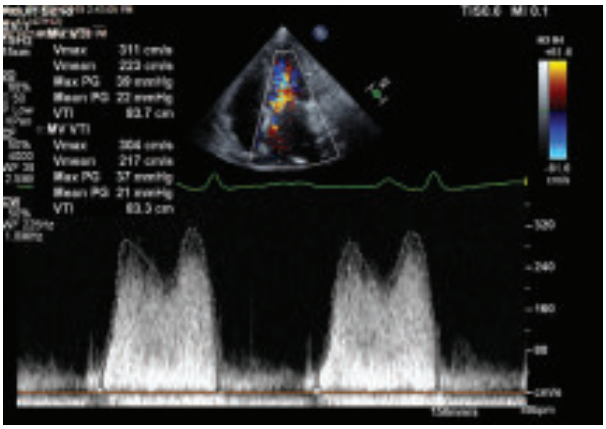

Fig.-1: *Echocardiographic feature consistent with mitral valve obstruction by LA mass*

Two large well circumscribed masses were identified. One mass had a stalk on the lateral wall; second mass had stalk in the medial wall. Both masses were resected completely and sent to pathology. The mitral valve was found to be normal. Large pericardial effusion and a pleural effusion bilaterally were drained. Post-op echocardiogram showed no residual masses, peak pressures were significantly lower and the heart had good function

Pathology: Leiomyosarcoma, low grade. Sections demonstrate a tumor with epithelioid to spindle cells, frequent bizarre nuclear atypia, cytoplasmic vacuoles focally positive for PAS and mitotic activity elevated to approximately 5 mitoses per 10 high

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power fields. No definite tumoral necrosis is identified.

By immunohistochemistry, tumor cells are weakly positive for SMA; focally positive for HHF-35 and desmin. Negative for myogenin, S100, CD34,31, AE1,3 and calretinin. Approximately 10% positive for Ki-67.

Although Leiomyosarcoma can arise as a primary cardiac tumor, clinical correlation is advised to exclude possible metastasis from the uterus or other primary site.

Post-surgical scans: CT chest/abd/pelv (7/23/19) shows no evidence of metastatic disease within the chest, abdomen, pelvis. Small pericardial effusion measuring 8 mm. TTE (8/23/19) demonstrating normal left ventricular systolic function, normal right ventricular function, no significant valve disease and no pulmonary hypertension. There is no pericardial effusion. No residual masses were noted in left atrium.

Within few months patient returned to ER with shortness of breath. Repeat echocardiogram revealed recurrent tumor. Cardiac MRI revealed several masses in the left atrium. A trilobed mass was noted attached to the lateral and inferior walls measuring approximately 4.2x2.2 cm and in close proximity to left lower pulmonary vein ostium. Another mass was noted attached to interatrial septum and atrial aspect of the anterior mitral leaflet measuring 1.7x1.4 cm that partially prolapses across the mitral valve. A third mass is attached to the left atrial roof extending to left upper pulmonary vein measuring 2.7x1.6 cm. A 9x7 mm area of the mass attached to the mitral leaflet is hyperintense with short inversion times and hypointense with long inversion times finding consistent with partial thrombus/necrotic tumor. Normal biventricular systolic function. No significant valve disease. No evidence of myocardial scar or necrosis. (Fig 2).

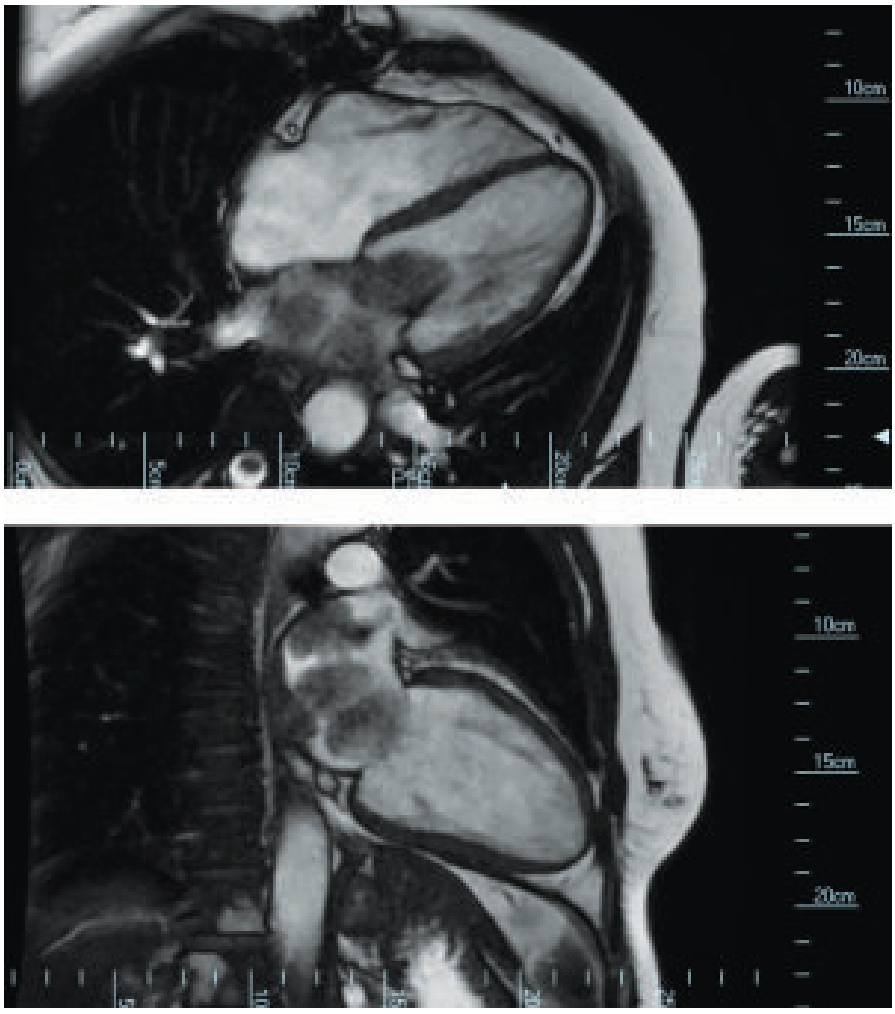

Fig.-2: *Cardiac MRI showing left atrial mass attached to mitral leaflet.*

She was deemed to be not a surgical candidate and was waiting for chemotherapy when she had another episode of shortness of breath and was admitted to hospital where work-up revealed recurrent leiomyosarcoma without any distant metastasis. She was started on chemotherapy with Doxorubicin. While on Doxorubicin, repeat echocardiogram showed left atrial masses are dramatically increased in size causing severe mitral stenosis, worsening tricuspid regurgitation and severe pulmonary hypertension. MRI also revealed increase in tumor size. Due to these findings, she was started on combination chemotherapy with Gemcitabine/Docetaxel. She remains relatively asymptomatic and tolerating this regimen well.

Discussion:

Primary cardiac Leiomyosarcoma is an extremely rare tumor with a dismal outcome. It occurs in less than 1% of malignant cases preceded by angiosarcoma, rhabdomyosarcoma, mesothelioma and fibroma.[1]

Primary site of origin is left atrium whereas mitral valve is extremely rare (2 in a 15 case series).[2] Because of the rarity of the tumor and its left atrial location, it is misinterpreted as myxoma in most of the cases by initial echocardiographic evaluation as was the case with our patient. Cardiac MRI plays a major role in localizing the tumor, in delineating its location in adjacent to neighboring structures such as pulmonary veins thereby assessing the extent of the spread of the tumor in and around the atrial chamber and somewhat in tissue characterizing. Ultimately pathological analysis of the excised tumor only clinches the final diagnosis.

Although various case reports have been published, this tumor with its rare occurrence and its myriad ways of presentation makes it worth revisiting it in a slightly different format than before. In our case, patient presented with acute pulmonary edema due to mitral valve obstruction caused by the tumor. After the tumor was resected, the mitral leaflets had no other intrinsic pathology of mitral stenosis and hence it was a truly pseudo-stenosis of the mitral valve. This is in contrast to other case report where the mitral valve was found to have intrinsic mitral stenosis at surgery and underwent mitral valve replacement.[3] The longest survival so far in adult cases has been 26 months.[4] Our patient is still alive after 16 months and doing well with chemotherapy without any distant metastasis.

Although surgery still remains the gold standard of treatment specially in case like ours where it causes mitral valve obstruction with acute pulmonary edema, post-surgical recurrence is a problem as exemplified by our case and some others.[4]

Although certain surgical techniques such as resection of all atrial myxoid tumors with at least 1 cm wide margin has been proposed as complete surgical resection appears to correlate with prolonged survival in few reported cases of Leiomyosarcoma.

Multiple chemotherapeutic agents have been tested in treating this tumor mostly with distant metastases although long term survival benefit of none of these agents have been fully established

The rare entity of primary cardiac leiomyosarcoma manifests in multiple different ways. Sometimes it appears as valvular disease as mitral valve mass resulting in vascular emergency as a peripheral vascular embolism at origin of common iliac arteries. At other times it masquerades as pseudo-stenosis of mitral valve only to be proven as adherent to true mitral stenosis postoperatively. It could also present as an aggressive tumor with distant metastasis to multiple organs and can be lethal. It exhibits recurrence after surgical excision and can present with therapeutic challenge in absence of any definitive treatment.

Since this disease entity is exceedingly rare and preserving cardiac functionality is of vital importance, a multidisciplinary approach towards its management is absolutely necessary. Considerations for palliative radiation therapy were discussed at the multidisciplinary tumor board but it was decided to be held off as she remained asymptomatic from cardiac perspective.

Conclusion:

Our case illustrates one other enigmatic presentation of this rare tumor with novelties in presentation, causing pseudo-stenosis of mitral valve, having post-surgical recurrence without any distant metastasis and still ongoing prolonged survival. We intend to share our experience through this case report how the use of

multimodality imaging such as echocardiogram, cardiac CT and MRI and the close collaboration between cardiologist and oncologist can be of immense help in taking care of patients presenting with this enigmatic cardiac tumor.

Conflict of Interest - None.

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