A Rare Case of Cardiac Malignancy: Myxoid Liposarcoma

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A 53-year-old female, who is a known hypertensive for 8 years, presented with difficulty in breathing for 1 week. Associated with orthopnea, the patient also complained of dry cough and fever. Fever was low grade, intermittent, and not associated with chills and rigors. No history of chest pain, palpitations, giddiness, syncopal attacks, or pedal edema was found.

On examination, patient was conscious, coherent, and oriented. Tachypnea and tachycardia were noted. Oxygen saturation was 98% at room air. JVP (jugular venous pulse) was elevated. No pallor, icterus, cyanosis, lymphadenopathy, or pedal edema was found. Cardiovascular examination revealed muffled heart sounds. Examination of other systems was unremarkable.

ECG (electrocardiogram) showed tachycardia, low voltage complexes, and nonspecific ST-T changes. Emergency echocardiogram was done, which showed massive pericardial effusion with homogenous mass attached to the right atrium with normal LV (left ventricular) function. Patient was taken up for emergency pericardiocentesis using a pig tail catheter, and 550 mL of hemorrhagic pericardial fluid was drained and sent for analysis.

Pericardial fluid was negative for malignant cells, AFB (acid-fast bacillus), and gene expert for MTB (mycobacterium tuberculosis). Protein was 5.7, sugar 91, ADA (adenosine deaminase) 61.1, LDH (lactate dehydrogenase) 3,069, and culture sensitivity showed no growth.

Once the patient was stabilized, a CECT (contrast-enhanced computed tomogram) Thorax was done for evaluating the cardiac mass. CECT showed minimally enhancing mass lesion in right atrial wall extending into pericardium. (computed tomography) CT-guided biopsy was done from the right atrial mass.

Histopathology showed lesion predominantly composed of atypical adipocytes and lipoblasts exhibiting moderate...
pleomorphism. Numerous signet ring lipoblasts were also noted admixed with delicate thin-walled arborizing and curvilinear capillaries. Background showed a myxoid matrix, shown in ►Fig. 1. The lipoblasts were focally positive for S-100 and negative for cytokeratin, immunohistochemically. With H&E (Hemotoxylin and Eosin) and IHC (immunohistochemistry) features, a diagnosis of myxoid liposarcoma was rendered.

PET-CT (positron emission tomography-computed tomography) was done to rule out metastasis. PET-CT showed mild FDG (fluorodeoxyglucose) avid hypodense lesion measuring 58 x 50 mm along the lateral wall of right atrium without any other significant lesions. Now, patient is planned for complete excision.

Primary cardiac tumors are a rare entity whose incidence, according to surgery and autopsy reports, is 0.3 to 0.7% of all cardiac tumors. Metastasis to heart from other primary sites is more common. Of this rare group, 75% are benign with most common entity being myxoma.

Liposarcomas are often asymptomatic. Symptoms may be due to compression or direct invasion of adjacent organs after attaining a considerable size, which may delay the diagnosis of liposarcomas.

Limited data are available for the management of cardiac sarcomas. The available data show that the prognosis is generally bleak. Complete resection is not of much benefit for malignant cardiac sarcomas. Very few studies addressed the use of chemotherapy in cardiac sarcomas. Isambert et al found that chemotherapy improved survival, but only in patients who were not operated. Abu Saleh et al studied the use of neoadjuvant chemotherapy. This study concluded that increased completed resection rates were seen in the neoadjuvant group, which translated into increased survival. Most commonly employed chemotherapy regimens were doxorubicin and ifosfamide.

In conclusion, myxoid liposarcoma arising from heart is an extremely rare entity. Surgical resection combined with chemotherapy or radiotherapy remains the main treatment strategy to date. Novel treatment strategies for the treatment of cardiac sarcomas, such as molecular targeting therapy, need to be explored further.

Conflict of Interest
None.

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References

Fig. 1 CMLS (cardiac myxoid liposarcoma) histology.