INTRODUCTION

A 36-year-old male presented with dyspnea for 1 year, progressively increasing in severity, with recent onset of orthopnea. He had recurrent episodes of syncope, bilateral lower limb swelling, distended abdomen in the past 1 month. The patient did not have any history of a cough or fever. Furthermore, there was no history of any congenital heart diseases and respiratory ailments such as tuberculosis or pneumonia. The patient was a nonsmoker and nonethanol user.

On examination, he had tachypnea, tachycardia with a stable blood pressure. He was not cyanosed. There was no clubbing, lymphadenopathy, and icterus, but bilateral pedal edema was present along with abdominal swelling. He was hypoxic with a room air saturation of 90% with further desaturation on exercise. Electrocardiogram was suggestive of cor pulmonale. His chest radiograph revealed bilateral dilated pulmonary arteries with multiple nodules in upper, mid, and lower lung zones, bilaterally [Figure 1]. The two-dimensional echocardiography was suggestive of severe pulmonary artery hypertension with an estimated pulmonary artery systolic pressure of 90 mm of mercury, dilated right atrium and right ventricle (RV), with a mass in the right ventricular outflow tract. His prothrombin time/activated partial thromboplastin time, International Normalized Ratio was normal, also protein c, protein s, and homocysteine levels were within normal limits.

DIFFERENTIAL DIAGNOSIS

With the above history and investigation strong possibility of chronic thromboembolic pulmonary hypertension (CTEPH) with the right ventricular clot was considered.

In view of mass in the right ventricular outflow tract possibility of primary cardiac tumors with multiple lung metastasis was also suspected.

Hence, the patient was further evaluated with computerized tomography pulmonary angiogram (CT-PA).

RADIOLOGY FINDINGS

Dr. Raj: CT-PA was performed after intravenous contrast administration. This demonstrated dilated right atrium and RV. There was hypodense nonenhancing filling defect within the RV. Multiple hypodense filling defects were present in segmental and subsegmental pulmonary arteries bilaterally. There was secondary dilatation of the pulmonary arteries with reasonable flow distally. Few bronchial artery collaterals were also seen. Overall, CT features were those of chronic pulmonary artery hypertension with multiple filling defects [Figure 2]. This was not entirely typical of chronic pulmonary thromboembolic disease in view of dilatation of vessels. Therefore, a working diagnosis of RV neoplasm with tumor emboli in the pulmonary vasculature was considered.
Dr. Syed/Dr. Tiyas/Dr. Muralimohan (pulmonology). The case was discussed in a multi-disciplinary meeting and it was decided to subject the patient for a positron emission tomography (PET) scan in view of the CT-PA findings, to rule out primary cardiac tumor.

Dr. Raj. To evaluate this further a fludeoxyglucose (FDG) PET/CT study was performed. This demonstrated non-FDG avid hypodense filling defects within the RV, the pulmonary arteries and the multiple “metastatic” pulmonary nodules [Figure 3]. No other primary neoplastic lesion was identified, further strengthening the possibility of the right ventricular myxoma.

Dr. Syed/Dr. Tiyas/Dr. Muralimohan (pulmonology). Case was again discussed in the multidisciplinary meeting and the consensus was right ventricular myxoma with multiple lung metastasis.

Literature review at this moment was done, according to the various studies available the most common primary cardiac tumors are myxomas accounting for about 25–40% of all cardiac masses.[1,2] Although myxomas most commonly affect the left atrium, the RV can also be rarely involved. The incidence of right-sided myxomas is around 5%,[3,4] An intracavitary mass that obstructs the right ventricular outflow tract is about 300 times more likely to be malignant than benign.

**MANAGEMENT**

Dr. Binoy. Operative intervention is mandatory to achieve complete relief from symptoms and to prevent dramatic complications that can occur in untreated patients. There have been some reports since the 1960s where successful surgical removal of right ventricular myxoma has been carried out.[5,6]

Hence, the patient was taken up for surgery with the intention of debulking the right ventricular mass. The right ventricular mass was removed and sent for histopathology. Later, the patient developed postoperative complications, and he succumbed.

**PATHOLOGY**

Dr. Preeti. The histopathology showed laminated membrane with focal giant cell response on one aspect. No nucleated germinal membrane was seen. Also seen in the same field was fibrocollagenous tissue with marked myxoid change, mostly probably could be an endocardial degenerative change or a valvular tissue showing degeneration.

Impression-hydatidosis of the heart most likely Echinococcus multilocularis as there was no nucleated germinal membrane seen [Figure 4].

**DISCUSSION**

Cardiac hydatid cyst remains one of the major differential diagnoses for myxomas in an endemic area. Primary cardiac hydatid cyst, though rare, is a well-known entity. The frequency of cardiac echinococcosis ranges only between 0%–5% and 2% according to various authors.[7] The continuous contractile activity of the heart makes it an unsuitable environment for the persistence of the hydatid cyst within it.[8] Echinococcosis commonly affects the ventricular myocardium and most of the time it is the left ventricle that is affected.[9] In the atria, however, there is no preference for either side. When the hydatid cyst is present in the right ventricular myocardium, then the chances of its rupturing into the ventricular cavity is higher than the chances of its perforating outside the cavity as occurs in the left ventricle.[9] Patients may present with acute pulmonary thromboembolism when the cyst perforates into, or daughter cysts migrate into the right ventricular cavity and are swept into the pulmonary arteries.[10] As these cyst components or daughter cysts cannot undergo fibrinolysis, there is clearly a greater risk of progression to CTEPH.

![Figure 3: Positron emission tomography - computerized tomography scan shows right ventricular hypodense mass lesion](image)

![Figure 4: Myocardium with a hydatid cyst with giant cell response](image)
E. unilocularis is much more common than E. multilocularis. Even though E. multilocularis can affect any organ, the liver and lungs are more frequently affected by it. E. multilocularis affecting the heart, to the best of our knowledge, has not been reported. E. multilocularis causes tissue invasion and destruction of surrounding tissue. The proliferation of the germinal layer and its invasion of blood vessels can cause distant metastasis to any organ, but the lungs and the brain are more frequently involved. Due to its aggressively invasive nature and multifocal involvement, it may mimic a malignancy with metastases.

In view of potential complications such as cyst rupture, cerebral embolism, and ventricular wall damage, surgical debulking becomes the mainstay of the treatment.

CONCLUSION

Cardiac echinococcosis should be included in the differential diagnosis of CTEPH in areas, endemic for echinococcosis. The lack of specific diagnostic tests for echinococcosis and its aggressive behavior may pose difficulties in differentiating it from malignancies. Strong suspicion of hydatidosis should be raised when ventricular myocardium in particular is involved. This may be especially true for E. multilocularis infections.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Reynen K. Cardiac myxomas. N Engl J Med 1995;333:1610-7.
2. Hirota J, Akiyama K, Taniyasu N, Matsuoka K, Kobayashi Y, Sakamoto N, et al. Injury to the tricuspid valve and membranous atrioventricular septum caused by huge calcified right ventricular myxoma: Report of a case. Circ J 2004;68:799-801.
3. Gopal AS, Arora NS, Messineo FC. Right ventricular myxoma. N Engl J Med 2000;342:295.
4. Kishimoto M, Sakai S, Saito S. Primary tumor of the heart: A report of a case with myxofibroma of the right ventricle. Am Heart J 1959;57:769-74.
5. Sakakibara S, Okawa M, Konno S, Hashimoto A, Gomi H, Miyamoto AM, et al. Myxoma of the right ventricle of the heart: A report of a case with successful removal and review of the literature. Am Heart J 1965;69:382-91.
6. Wada J, Ikeda T, Komatsu S, Ikeda K, Endo T, Sato T. Right ventricular myxoma. Report of a case diagnosed preoperatively and removed successfully. Ann Thorac Surg 1965;1:84-8.
7. Calamai G, Perna AM, Venturini A. Hydatid disease of the heart. Report of five cases and review of the literature. Thorax 1974;29:451-8.
8. Ulgen MS, Yazici M, Kayrak M, Düzenli MA, Koç F. Three-year follow-up of recurrent cardiac echinococcosis simulating myxoma: Report of a rare case. Anadolu Kardiyol Derg 2007;7:442-3.
9. Shevchenko YL, Travin NO, Musaev GH, Morozov AV. Heart echinococcosis: Current problems and surgical treatment. Multimed Man Cardiothorac Surg 2006;2006:mmcts. 2005.001115.
10. Leila A, Laroussi L, Abdennadher M, Msaad S, Frihka I, Kammoun S. A cardiac hydatid cyst underlying pulmonary embolism: A case report. Pan Afr Med J 2011;8:12.
11. Canda MS, Guray M, Canda T, Astarcioğlu H. The pathology of echinococcosis and the current echinococcosis problem in Western Turkey (A report of pathologic features in 80 cases). Turk J Med Sci 2003;33:369-74.
12. Czermak BV, Unsinn KM, Gotwald T, Waldenberger P, Freund MC, Bale RJ, et al. Echinococcus multilocularis revisited. AJR Am J Roentgenol 2001;176:1207-12.