Right atrial angiosarcoma presenting as supraventricular tachycardia

A 34-year-old woman was referred to our center for recurrent rapid palpitations which were usually self-terminating. She was found to have supraventricular tachycardia during one such episode (Figure 1, Panel A), which slowed down with carotid sinus massage and verapamil tablet. Subsequent ECGs brought out the mechanism to be atrial tachycardia with variable atrioventricular conduction, which reverted to sinus rhythm after 2 hours. She was otherwise healthy with no other symptoms and clinical examination was normal.

Her echocardiogram showed a mass attached to the lateral wall of the right atrium (RA) (Panel B), in close proximity to the tricuspid valve. No other abnormality was present. Our differential diagnoses were thrombus, fibroma, vascular tumor, and granuloma. We administered intravenous left ventricular opacification contrast, which showed a well-defined filling defect (Panel C). A cardiac magnetic resonance imaging was performed, which showed that the mass also extended to the RA appendage (Panel D). A small pericardial effusion without any pericardial thickening was also noted and the mass was suspected to be neoplastic. Hence, a whole-body positron emission tomography-computed tomography was done which showed the RA mass to have intense metabolic activity without any active disease elsewhere in the body. Hence, a primary cardiac malignant tumor was the most likely diagnosis.

With the help of echocardiography and fluoroscopy, an endomyocardial biopsy was performed using the femoral venous approach. A steerable 8F sheath was used through which the biopctome forceps was passed. Histopathology showed a remarkably cellular mitotically active malignant tumor composed of polygonal to spindle-shaped cells and pronounced vascularity, confirming the mass to be an angiosarcoma (Panels E and F). In view of infiltration over a wide area of the RA and involvement of the tricuspid valve, surgery was considered not feasible. After confirmatory immunohistochemistry, the patient was started on chemotherapy, with a plan of surgery at a later date, after reduction of tumor size.

Primary cardiac tumors are a rare entity in adults with only 25% being malignant.[1] Among primary malignant cardiac tumors, angiosarcoma is the commonest one. This is an endothelial cell tumor, nearly 90% of tumors occurring in the right atrium as a multicentric mass. The presentation is usually in the third and fourth decades and is varied—chest pain, dyspnea, myalgia, arrhythmias, pulmonary embolism, pericardial effusion, cardiac rupture, and distant metastasis.[2]

Figure 1: Panel A: Supraventricular tachycardia @ 160/minute. Panel B: Echocardiogram, apical four chamber view. There is clearly a mass in the right atrium, near the lateral wall and abutting the tricuspid valve. Panel C: Contrast echocardiogram. All four chambers are well opacified. The filling defect is clearly demarcated in the right atrium. Panel D: Cardiac MRI. The arrow points to the mass seen in the echocardiographic examination. The black dots show the extension into the right atrial appendage. Panel E: Pleomorphic plump ovoid to spindle shaped cells forming delicate dilated capillary channels (H&E x 400). Panel F: Some of the polygonal cells have formed intracytoplasmic lumina (arrow) which contain red blood cells (H&E x 400)
There is no standard protocol for their management and they mostly carry a grave prognosis, with a mean survival of a few months without surgical resection.\(^3\) Congestive heart failure, cardiac tamponade, obstruction to cardiac output and consequences of metastatic disease are the modes of death. Surgery is the commonly chosen therapy, with aggressive and complete resection in case of localized disease and possible debulking in case of advanced and metastatic disease. Moreover, adjuvant chemotherapy is almost essential and provides improved survival, which is yet only 1–4 years.\(^4\)

In our patient the initial presentation of a supraventricular tachycardia in a young healthy woman with a clinically normal heart, did not suggest anything sinister. The echocardiographic finding was surprising and led to the final diagnosis of a malignancy!

Declaration of patient consent
The authors certify that appropriate patient consent was obtained.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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Access this article online

Quick Response Code:  
Website:  
DOI: 10.4103/jpgm.JPGM_477_20  
PubMed ID: 33037166

How to cite this article: Bachani N, Bagchi A, Vaideeswar P, Lokhandwala Y. Right atrial angiosarcoma presenting as supraventricular tachycardia. J Postgrad Med 2020;66:222-3.