Giant Intracardiac Smooth-Muscle Cell Tumor Presenting as Superior Vena Cava Syndrome

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We report the case of a 53-year-old man presenting with a superior vena cava syndrome secondary to a giant intracardiac mass occupying the majority of the right-side cavities of the heart. A mass measuring 10.5 × 9.5 × 4.0 cm originating from the superior vena cava and occupying most of the right atrium extended through the tricuspid valve into the right ventricle. The mass was resected. The patient was discharged on postoperative day 11. The pathology report revealed the mass to be a smooth-muscle cell tumor of unknown malignant potential.


Primary cardiac tumors are a rare entity with a prevalence of 0.01% to 0.03% compared with secondary cardiac tumors with a prevalence of 1% based on postmortem studies [1, 2]. On thorough review of current literature, this is the first case describing an intracardiac smooth muscle cell tumor of unknown malignant potential (STUMP).

A 53-year-old man with a history of a testicular nonseminomatous tumor treated with orchiectomy and chemotherapy two years earlier presented with a chief complaint of progressive stabbing chest pain and exertional dyspnea. Physical examination was notable for significant jugular venous distention along with edema and cyanosis of the face and bilateral upper extremities, suggestive of superior vena cava (SVC) syndrome. Laboratory study was significant for elevated pro-B-type natriuretic peptide of 1,598 ng/mL. As part of his workup, the patient underwent a transthoracic echocardiography, and it became evident that he had a multilobulated intracardiac mass. The four-chamber view (Fig 1A) shows the tumor protruding through the tricuspid valve into the right ventricle (RV), and the bicaval view (Fig 1B) shows the tumor entering the right atrium (RA). Transeosophageal echocardiography also showed a dilated RV with hypokinesia of the free wall (Video 1 and Video 2). Computed tomography imaging of the chest confirmed a large heterogenous mass that spanned from the SVC to the RV, partially obstructing the RV outflow tract (Fig 2). There were no significant findings in the metastatic workup, which included abdominal and pelvic computed tomography, and tumor markers including lactate dehydrogenase, human chorionic gonadotropin, and alpha-fetoprotein were within normal limits. Given the significant strain on the patient’s hemodynamics, the decision was made to proceed with a resection of the mass.

A median sternotomy approach was used and the patient was placed on cardiopulmonary bypass. The cardiac chamber was accessed through a right atriotomy. A large fibrous mass was found extending from the SVC through the RA and into the RV (Fig 3A). The tumor was adherent to the endothelial surface of the SVC and the endocardium of the RA. Once the mass was dissected from the SVC and RA, it was able to be retrieved en bloc out of the RV through the tricuspid valve. The tricuspid valve annulus was dilated secondary to the mass, causing tricuspid insufficiency and therefore DeVega annuloplasty was performed. The patient was successfully decannulated from cardiopulmonary bypass, extubated, and transferred to the intensive care unit for recovery. His postoperative course was significant for a brief episode of hemodynamic instability requiring pressors; however, that resolved within 24 hours. The patient was discharged on postoperative day 11.

Pathology reported a diagnosis of a smooth-muscle cell tumor of unknown malignant potential (STUMP) that tested negative for estrogen and progesterone receptors (Fig 3B). At the 4-week follow-up, the patient exhibited resolution of his cardiopulmonary symptoms as well as normalization of his pro-B-type natriuretic peptide. His follow-up chest and abdomen computed tomography scan did not show any additional masses or lymphadenopathies. Pericardial fluid cytology and prostate biopsy were negative for malignancy.

Comment

Primary cardiac tumors are exceedingly rare in comparison with metastatic disease. The STUMP tumors are a group of tumors that cannot be clearly classified as benign or malignant, and are most commonly found in women in association with uterine tumors. Our patient had been previously diagnosed with testicular nonseminomatous tumor two years earlier, yet this disease had been in remission after orchiectomy and chemotherapy. In addition, the histologic analysis of the resected cardiac tumor did not have any resemblance to testicular cancer to suggest metastatic disease.

The Videos can be viewed in the online version of this article [https://doi.org/10.1016/j.athoracsur.2017.11.065] on http://www.annalsthoracicsurgery.org.
which is indicative of a primary process. To our knowledge, this is the first intracardiac STUMP reported, and also the largest intracardiac tumor reported in the literature.

The histopathologic analysis of the resected tumor was strongly positive for Caldesmon in more than 95% of the neoplastic cells, which pointed to the diagnosis of a smooth muscle cell tumor. The diagnosis for STUMP is based on histologic findings of (1) focal moderate to severe cytologic atypia, no tumor cell necrosis, and fewer than 5 mitosis events per 10 high-power fields; or (2) no or mild cytologic atypia, tumor cell necrosis, and fewer than 10 mitosis events per 10 high-power fields [3]. Analysis of this tumor complied with these diagnostic criteria for STUMP. However, the tumor was negative for estrogen and progesterone receptors, unlike other STUMP reported in literature, while strongly positive for p53 and Ki-67; that pointed toward the characteristics of a leiomyosarcoma, which can also be seen or confused with STUMP. The diagnosis of STUMP versus
leiomyosarcoma was a point of discussion, and therefore multidisciplinary meetings were held. Based on serology, follow-up imaging, and no evidence of a primary site of origin, the diagnosis of primary intracardiac STUMP was confirmed.

Treatment recommendation for STUMP is not well defined because of its rarity, unknown malignant potential, and its often incidental diagnosis. Current consensus on treatment and follow-up of STUMP is based on uterine leiomyosarcoma data. If diagnosed, the patient undergoes surgical resection with close follow-up by annual imaging for as long as 5 years for possible recurrence or malignant conversion. There are no current recommendations for chemotherapy or radiation [4].

References