Mini Review

Interventional Cardiology

Advancements in diagnosis of cardiac tumor: A personal reflection on my contributions

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Abstract

Tumor occurring in cardiac or major vessels is uncommon but life-threatening due to its direct influence on hemodynamics. Primary cardiac synovial sarcoma is one of them. In general, sarcoma frequently occurs in the limb of young people, and can be easily detected only by visual examination. However, it is very challenging to reveal sarcoma occurring in except body surface, because of its silent progression and rarity. The lack of detection of a specific maker can also delay diagnosis. Although the gold standard for definitive diagnosis is immunohistochemistry, imaging diagnosis is given priority in most cases. The effective treatment is only surgical resection for life expansion, and multimodality imaging is essential for early detection and diagnosis. This article explores the impact of multimodality imaging on clinical complex cases. In addition to that, this review highlights salient features and new insights into primary cardiac synovial sarcoma, and the importance of multidisciplinary approach.

Keywords: Cardiac synovial sarcoma • Diagnosis • Tumor • Heart • Epithelioid cells

Introduction

Primary cardiac synovial sarcoma was first reported in 1978 [1]. Primary cardiac tumors are uncommon, occurring less frequently than metastatic tumors (in a ratio of approximately 1:20~40) with an incidence in autopsy series ranging only from 0.001%-0.003%. Most of the primary cardiac tumors are benign and approximately 25% of them are malignant, of which the majority are sarcomas [2-4]. Primary cardiac synovial sarcoma accounts for approximately only 4.2% of primary cardiac sarcomas [5,6]. Synovial sarcoma tends to arise from the heart but also arise from the pericardium. The predilection site is the right atrium, with a right to left ratio of 2:1 [7]. It is usually discovered at the advanced stage, and most synovial sarcomas involve more than 1 anatomic compartment within the heart. In our case, complete resection was archived and an immunobiological examination was performed. The histological examinations of the tumor revealed bunch growth of spindle-shaped cells among epithelioid cells presented ductal structure. There were calcification and collagen fiber pigmentation in fibrosarcoma. A part of the tumor showed calcification and elastic fiber deposit. The immunohistochemistry results were positive for cytokeratin (AE1/3) in epithelioid, and diffuse positive cytokeratin (SS18-SSX, SSX) in both of epithelial and spindle cells. *SMARCB1* expression was weak compared with normal cells, this conformed to immunological characteristics of synovial sarcoma. In addition to immunohistochemistry, molecular pathology is useful for definitive diagnosis. Molecular genetic studies have shown a balanced reciprocal translocation, t (X; 18) (p11.2; q11.2), between the *SYT* gene on chromosome 18 and the *SSX1* or *SSX2* gene on chromosome X. This translocation is a specific cytogenetic abnormality that occurs consistently in synovial sarcoma. Its detection by fluorescence in situ hybridization or real-time PCR has therefore become useful for confirming the diagnosis of synovial

sarcoma. There are some reports which synovial sarcoma with complex karyotype has a greater tendency to metastasize.

Literature Review

Clinical symptoms of primary cardiac synovial sarcoma often relate to the location occupied by the tumor. Most synovial sarcoma patients are admitted with congestive heart failure symptoms or syncope, but it is often too late to treat when symptoms appear. The boundary of symptomatic or not, is unclear, but it is expected to appear symptoms when the tumor becomes above a certain size, affecting hemodynamics. The critical tumor diameter depends on individual cardiac size, so it is difficult to establish a reference. However, in cases of localized and small mass with no adhesion, complete resection is possible and long-term survival is expected. Resection of small (<2.5 cm) non-valvular or (<4.0 cm) pericardial synovial sarcomas allows survival beyond 3 years, but postsurgical survival is only 12-15 months with larger (>5.0 cm) intracardiac tumors. We found a few reports about primary cardiac synovial sarcoma with performed emergency operation [8,9]. One case is a primary synovial sarcoma of the right heart involving the tricuspid valve in an elderly Chinese woman. The woman underwent tumor resection surgery, and escaped death. Another case is in which synovial sarcoma obstructed the right atrium and prolapsed into the right ventricle in a young Egyptian man. He also underwent emergency tumor resection, and it was archived. However, the patient succumbed to hemorrhage from a brain metastasis [10]. As of 2004, the longest reported survival after surgery is 14 years. A combined modality therapy of surgery, chemotherapy, and radiotherapy was performed in that case [11]. In our case, the tumor diameter was more than 5.0 cm and it existed intracardiac (Figure 1). It is true that these were poor prognosis factors, but the tumor has not infiltrated yet. This is another important factor for the success of the operation. For that, quickly understanding the patient's hemodynamics was essential. Echocardiography revealed just a little residual flow through the tricuspid valve and decreased cardiac output due to the tumor. This meant the patient's hemodynamics were going to collapse, and emergency tumor resection was the only treatment. In addition, a CT scan timely provides the minimum information, such as the tumor size, location and positional relation with around tissue. It can provide general outline in a few minutes.

Discussion

Although we prioritized operation over imaging diagnosis due to hemodynamic deterioration in this case, Cardiac Magnetic Resonance Imaging (CMR) is useful for the characterization of tumors. Another case is presented where CMR proved valuable for differential diagnosis.

A 67-year-old-female presented with gradually worsening leg edema and dyspnea. Physical examination revealed Kussmaul's sign. A transthoracic echocardiography detected an enlarged right ventricle, which compressed the left ventricle, resulting in decreased cardiac output. The first diagnosis was pulmonary embolism. An enhanced Computer Tomography (CT) and pulmonary angiography revealed atypical findings, which was a massive filling defect in the right ventricular outflow tract, the main pulmonary artery and its branches (Figure 2). Although the mass was suspected to be the culprit, the diagnostic findings were not enough for differentiation. The patient's hemodynamics had been maintained, so CMR was performed. It showed that the mass was presented with bulging margins and higher T2 signal intensity, which were specific findings of primary Pulmonary Artery Sarcoma (PAS) [12]. The only treatment was to release the obstetrics. After conference with our surgeons, urgent tumor resection was performed. However, the tumor had already infiltrated to endothelium of the pulmonary artery and complete resection was not archived. The patient escaped death, but recurrence and metastasis were detected at 4 months after surgery.

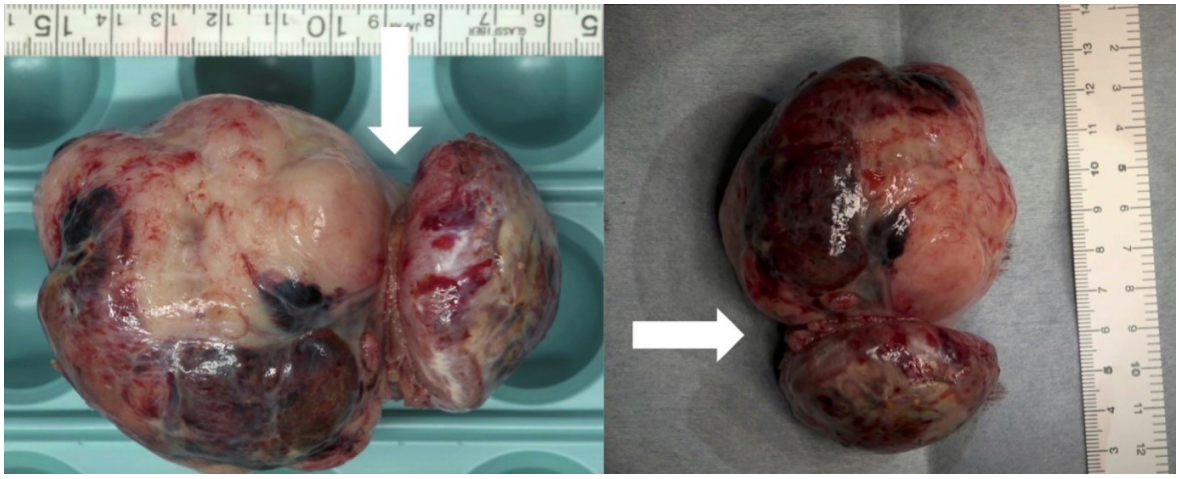

Figure 1: Synovial sarcoma arising from the right atrium wall. **Note:** White arrow is the dividing point from inside to outside of the right atrium.

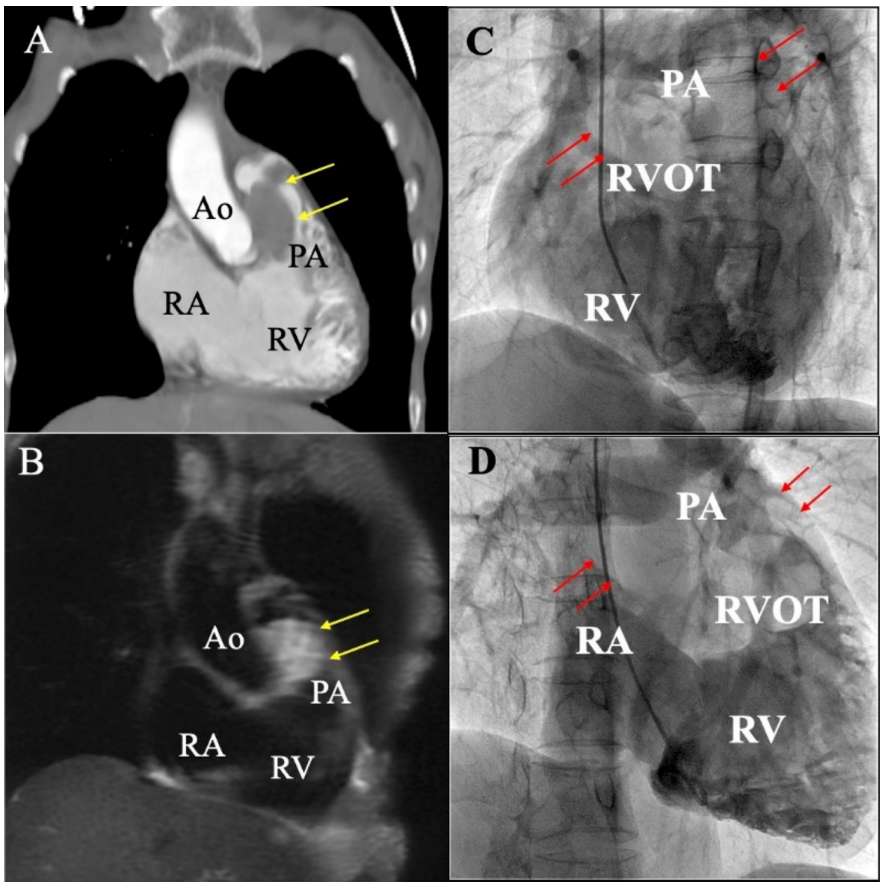

Figure 2: The imaging of pulmonary artery sarcoma. **Note:** A) Yellow arrow indicates enhanced CT axial view; B) Red arrow indicates the mass was presented with higher signal intensity on CMR T2 weighted image; C) Pulmonary Angiography (AP) and RAO; D) Red arrow indicates massive filling defect in RVOT and PA branches. Ao: Aorta; LA: Left Atrium; PA: Pulmonary Artery; RA: Right Atrium; RV: Right Ventricle; RVOT: Right Ventricular Outflow Tract.

Primary pulmonary artery sarcoma is a life-threatening tumor the same as primary cardiac synovial sarcoma. It develops within the inner or middle layer of the pulmonary artery, with an estimated incidence rate ranging from 0.001%-0.003% [13]. The survival period of primary pulmonary artery sarcoma is approximately 1.5 months for those who did not undergo surgery timely. Currently, surgical resection remains the main treatment for PAS [14,15]. Previous studies have reported that 85% of pulmonary artery sarcoma cases involve the main pulmonary artery, 71% involve the right pulmonary artery, 65% involve the left pulmonary artery, 32% involve the pulmonary valve, and 10% involve the right ventricular outflow tract [16]. In contrast, pulmonary embolism often occurs in the right lung, bilateral lower lungs, and peripheral pulmonary artery or other areas [17]. Thinking from the perspective of predilection site, pulmonary artery sarcoma more often occurs collapse of hemodynamics than thromboembolism. Early detection and diagnosis play a key role for expanding lifespan.

While there is a difference in which complete resection was archived or not, we successes lifesaving in described above two cases. There are three clinical learning points. First, the tumor should be recognized as an aggravating factor of hemodynamics. Tumors originating from cardiac or main vessels are very rare. However, it should be considered that physical obstacles may exist somewhere in systematic circulation, in case of heart failure of unknown etiology. Giving all considerable differentiation enables us to have a broad view and leads to proper diagnosis. Second, multimodality should be used based on their property. Echocardiography specializes in the comprehension of hemodynamics; therefore it is useful cases in which the patient's condition is unstable and quick evaluation is needed. Once detected that abnormal findings are disturbing circulation, understanding its location and positional relation around tissue will be needed. A CT scan gives this information and helps to make a surgical plan. When more detailed information is needed for decision of treatment, CMR should be performed. Such as in the second case, pulmonary artery sarcoma presents very similar imaging to pulmonary embolism. CMR is useful for distinguishing thrombosis or not. Fully understanding and combining multimodality according to their characteristics leads to early detection and lifesaving even if differential diagnosis is challenging. Third, a multidisciplinary approach involving oncologists, pathologists, surgeons, and cardiologists was essential, and this is the point which has should be emphasized most. Team medical care is more meaningful in such a complex clinical case, which have no established treatment.

Conclusion

Then, this is just a hypothesis, but there might exist a strong specific genetic or environmental factor of occurrence of cardiac sarcoma. A natural next step would be to detect a certain genetic mutation or environmental factor as this could enable early detection and primary prophylaxis. Hope this article will encourage further exploration of cardiac sarcoma. Its implications could lead to early detection and expanding life span, benefiting patients.

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