Cardiac Arteriovenous Malformation Involving Right Atrium Wall: Case Report of Rare Condition and Brief Review of the Literature

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Abstract

Cardiac arteriovenous malformation (AVM) is a rare finding that ranges in presentation based upon size and location. Whether congenital or acquired, they can result in a variety of cardiac complications, including sudden cardiac death. The classification of these lesions, through the International Society for the Study of Vascular Anomalies (ISSVA), has aimed to separate these vascular malformations from tumors and enhance our understanding. Yet, the separation of the two remains a challenge in some cases. In this paper, we will present a case of cardiac AVM involving the left atrium wall with a review of current literature.

Keywords: Arteriovenous Malformation, Cardiovascular, Right atrium

Abbreviations

AVM: Arteriovenous malformation; ISSVA: The International Society for the Study of Vascular Anomalies; MDCT: Multi Detector Computed Tomography; RA: Right Atrium

Introduction

Arteriovenous malformation (AVM) of the heart is a relatively rare finding due to the low incidence and sometimes asymptomatic presentation [1-3]. They can be congenital or acquired secondarily through trauma, surgery, or invasive procedures [3]. The effects of cardiac AVM are attributed to size, shape, organization, configuration, and location (e.g., endocardial, intramuscular, intramural, or epicardial) of the vascular malformation and range from asymptomatic to myocardial necrosis, bacterial endocarditis, and heart failure [1-4,8]. Large cardiac AVMs can result in severe myocardial ischemia and even death, due to the high volume of blood shunted from arteries to veins, depriving the myocardial tissue of oxygen [3,4]. Given the rarity of cardiac AVM and its limited clinical information, we aim to both present a review of current literature and to contribute to the field of research surrounding this topic, by presenting the unique case of cardiac AVM.

Case Presentation

A 68-year-old man presented with a cardiac mass. The mass was detected by transthoracic echocardiography during a routine medical checkup. The patient was asymptomatic and reported a history of controlled type-II diabetes for 9 years and controlled hypertension for several years. No other significant history or family history was reported. Electrocardiography was normal but transesophageal echocardiography showed a right atrium mass measuring 2.4 X 1.8 cm.

Imaging studies were inconclusive, and a benign cardiac tumor was suspected including possibly myxoma, lipoma, hemangioma and fibroelastoma, but a malignant vascular tumor was not ruled out. Initial management plan was to perform a tissue biopsy for definitive diagnosis before surgical intervention. However, a multidisciplinary meeting recommended a median sternotomy for the surgical resection of the mass to avoid the risk of pulmonary embolization and to determine the definitive diagnosis. The mass was surgically removed, and it was confirmed that the mass originated from the wall of the right atrium and an intraoperative transesophageal echocardiogram showed that the mass had been successfully removed.

Histopathological examination of the resected mass revealed an oval mass measuring 2.4 X 1.8 cm attached to the right atrium wall without infiltration into the surrounding structures (Figure 1A). The mass showed abundant branched vessels, thick-walled arteries, and thin-walled veins (Figure 1B). In addition, scattered abnormal arteries with irregularly thick wall were also identified (Figure 1C). The vascular walls were lined up with bland plumb endothelial cells with no evidence of pleomorphism, hyperchromasia or abnormal mitosis (Figure 1D). Elastic and trichrome (Figure 1E-F) stains were utilized to
identify the arteries and the veins. No nuclear malignant features were identified. The histomorphologic features were diagnostic of benign cardiac arteriovenous malformation.

No mass recurrence was shown by echocardiography during 18 months’ follow-up, after which the patient was lost to follow up.

Discussion

Arteriovenous malformation (AVM) is the result of aberrant connections between arteries and veins that can occur during embryonic development [4]. These abnormal connections result in vascular endothelial cell hyperplasia and absence of a capillary bed, eliminating vascular resistance and allowing for the direct flow of blood at high rates between arteries and veins [1,5]. While these vascular malformations can develop in almost any part of the human body, cardiac involvement is a relatively rare finding [1-5,8]. In the heart, these vascular malformations can involve the coronary arteries and/or adjacent vessels and one of the heart chambers (coronary-cameral fistula), or the heart wall itself [4]. In our case, this uncommon finding was localized to the right atrium (RA) wall.

In the past, cardiac vascular anomalies were termed hemangiomas and classified according to their location and vessel involvement [1,5]. Over the years, The International Society for the Study of Vascular Anomalies (ISSVA) has been widely used for the classification of these vascular anomalies into vascular tumors (mainly hemangiomas) and vascular malformations [1,5]. This focus was aimed to further separate vascular malformations from tumors and aid in our understanding of the clinical behavior and prognosis of these lesions [5].

In a study of 107 symptomatic vascular malformations conducted by Meijer-Jorna et al., [5], local tissue hypoxia due to malformation was deemed the cause of the vasoproliferative response in these angioproliferative masses. These mature AVM endothelial cells were cultivated and analyzed by Wautier et al., [9], showing expression of protooncogene, ets-1, a gene normally produced by immature endothelial cells of capillaries for proliferative and apoptotic processes [8]. These tissues also express genes that encode Ephrin B2/4, proteins normally involved in angiogenesis [8,9]. With medical advances and better understanding of the underlying genetic basis of these vascular lesions, potential molecular targets may shed light on new treatment options and pharmacotherapy benefits [1].

The diagnosis of AVM is made histopathologically through the visualization of mature but malformed tortuous arteries and dilated veins [5]. In the reported cases of cardiac AVM, these abnormal vessels were embedded in collagen-rich fibrous tissue with immature capillary vessels [5,8]. The arterial component showed abundant elastic lamellae in the thick portions of vessel walls, whereas the thinner portions displayed a single layer of external elastic lamina, indicating venous origin [5,8].

While histopathology is required for the final diagnosis of cardiac AVM, echocardiography, computed tomography (CT), and magnetic resonance imaging (MRI) of the heart are beneficial in ruling out other pathologies like myxomas, lipomas, fibroelastomas, and hemangiomas [1,5]. Localization of these lesions is also important in determining a differential. Despite the works of the ISSVA, separating true hemangiomas from
vascular malformations remains a challenge. Recent studies have shown hemangiomas to stain positive for Wilms Tumor-1 (WT-1) and erythrocyte-type glucose transport protein-1 (GLUT-1), immunohistochemical markers beneficial in the differentiation of the two [6,7,10]. Although these stains can be used to resolve a differential, echocardiography and Multi Detector Computed Tomography (MDCT) remain the primary method for the initial evaluation of vascular lesions [1,3,4]. Enhanced echocardiography and 18F-fluorodeoxyglucose-positron emission tomography can be used to better assess the vascular lesion, but MDCT can provide a more detailed anatomical analysis through post-processing image reconstruction algorithms that are useful for determining treatment options and management [1,3,4].

Symptomatic vascular malformations are often treated surgically through transcatheter embolization, a procedure designed to reroute blood away from the vascular malformation [3,8]. Surgical removal can also be performed if the location is easily accessible. While asymptomatic patients may be managed conservatively, the literature is unclear whether these patients should be treated surgically to prevent future cardiac complications.

Given the rarity of cardiac vascular malformations and limited reporting in the literature, it is our hope that this report raises awareness of this uncommon finding and what remains an unmet need in the understanding and differentiation of these vascular malformations.

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References


