Case Report

Calcified Amorphous Tumor of the Mitral Valve Causing Stroke: A Rare Case of Heart Tumor

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Abstract

Calcified amorphous tumor (CAT), first described in 1997, is a rare primary cardiac tumor with a lifetime incidence of 0.0017%–0.02%, accounting for approximately 2.5% of all primary cardiac tumors. CAT can be discovered incidentally or following neurological events. Here, we report a poststroke patient diagnosed with filamentous formation on the mitral valve. After the surgery, the biopsy identified it as a CAT. There was a debate among colleagues regarding the indication for surgery due to the small mass. The prevalence of valvular strands is from 5.5% in the general population to 40% in patients with unexplained stroke. The incidence of systemic embolism, including cerebral, retinal, and transient ischemic attacks, is reported at 53%. Considering the significant risk of cerebrovascular accidents associated with calcified amorphous or similar masses, especially in patients with a history of stroke, surgery remains the treatment of choice.

Keywords: Calcified amorphous tumor, cardiac tumor, stroke

Introduction

Primary cardiac tumors are rare, with a lifetime incidence of 0.0017%–0.02%. Among primary cardiac tumors, calcified amorphous tumor (CAT) is estimated to account for about 2.5%. CAT is a nonneoplastic primary tumor of the heart, first described by Reynolds *et al*. in 1997.[1] It is usually discovered during cardiac imaging studies or following neurological events.[2] We present a case of a poststroke patient in whom a formation was discovered on the mitral valve. The case remains unique because CAT is encountered very rarely. Patients with a history of cerebral accident and presence of mitral strand have a high incidence of stroke, and surgery remains the only way for the differential diagnosis with other heart tumors and treatment.

Case Report

A 62-year-old woman was admitted to the clinic with the diagnosis of the mitral strand, probably fibroelastoma. She complained of nontypical anginal chest pain and referred a history of a cerebrovascular accident 6 months before. Trying to identify the cause of her stroke, a filamentous formation (mitral strand) was discovered on the mitral valve, probably a fibroelastoma, for which urgent surgery was recommended. In the interim, she underwent 48-h Holter rhythm monitoring and computed tomography of the head and carotid arteries to determine the cause of the cerebrovascular accident, but the results were all normal. Transesophageal echocardiography revealed fibrotic mitral valve leaflets, mild-to-moderate mitral regurgitation, and a very mobile filamentous formation about 10 mm in length, attached to the posterior leaflet of the mitral valve on the atrial side, suggestive of fibroelastoma [Figure 1].

The patient underwent coronary angiography, which revealed two-vessel coronary artery disease. The intervention was performed under cardiopulmonary bypass and included tumor resection, two coronary artery bypass grafts, left internal mammary artery to left anterior descending artery and a saphenous venous graft to first obtuse marginal artery, and mitral valve annuloplasty with an Edwards ring No. 26. The milky-colored filamentous formation, approximately 10 mm in length, was fixed in the middle of the P2 scallop of the posterior

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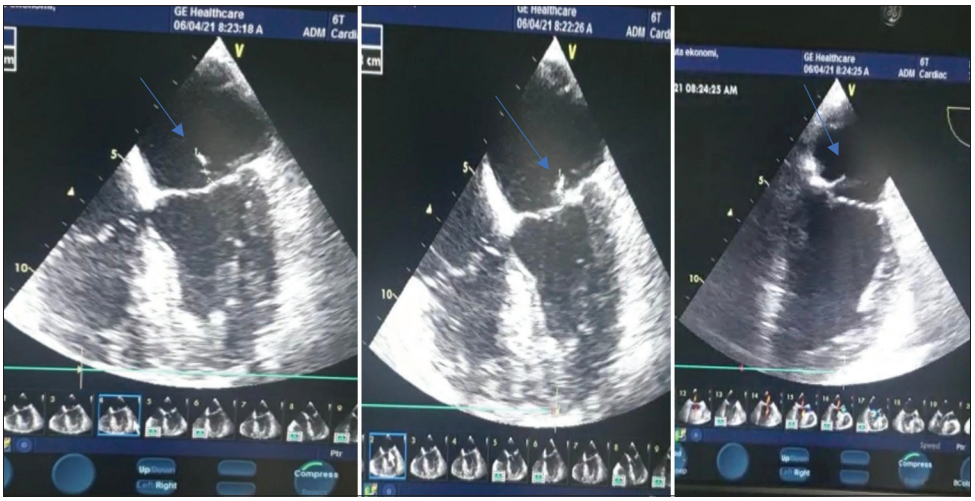

Figure 1: Transesophageal echocardiography. The arrow indicates the filamentous formation

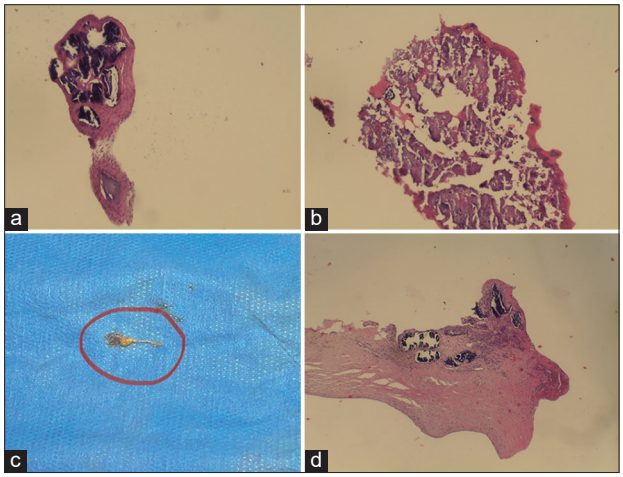

Figure 2: Macroscopy and biopsy. (a) Microscopic findings showing heterogeneous calcium deposits with surrounding amorphous eosinophilic and fibrinous material (magnification × 40). (b)Microscopic findings showing acellular eosinophilic tissue with nodular and diffuse central basophilic calcium deposits. Rare inflammatory multinuclear giant cells adjacent to the deposits (magnification × 100). (c)Macroscopic image of the lesion. (d)CAT microscopic with acellular tissue with degenerated fibrin at the periphery of the lesion (magnification × 100)

leaflet valve. It was removed along with a 5-mm portion of P2 at the fixation site. The defect was closed with continuous 5/0 prolene suture. The formation [Figure 2c] was immediately fixed in formalin and sent forbiopsy. The intervention and the postoperative period were uneventful. The histopathologic examination revealed acellular eosinophilic tissue with nodular and diffuse central basophilic calcium deposits and rare inflammator multinuclear giant cells adjacent to the deposits. Degenerated fibrin was observed at the periphery of the lesion [Figure 2a, b and d]. The diagnosis was confirmed as a CAT.

Discussion

While primary cardiac tumors are rare, CAT accounts for a small part of them. Histologically, it is characterized by calcified nodules and amorphous fibrinous material. CAT is a nonneoplastic primary tumor of the heart, first described as a special entity by Reynolds *et al*. in 1997.[1] It is usually discovered casually during cardiac imaging or following a neurologic event, as in our case.[2]

de Hemptinne *et al*.[3] reported that valvular disease, concomitant with mitral annular calcification, end-stage renal disease, diabetes mellitus, and coronary artery disease (CAD) are the most frequently associated conditions. Our patient had concomitant critical stenosis of the left anterior descending and first obtuse marginal arteries. The echocardiography showed significant mitral annulus calcification. Tumor size ranges from small punctate lesions (1.7 mm) to very large masses (20 mm × 90 mm). CAT can be located in all cardiac chambers, aortic valve,[4] and left ventricle outflow tract,[5] although it most frequently grows on the mitral valve or annulus, as in our case.[2,3]

Pulmonary or systemic embolization was reported in 31% of the cases.[3] Kimura *et al*. reported multiple cerebral infarctions from a posterior cusp of the mitral valve first treated with apixaban and aspirin. Follow-up showed newly developed multiple cerebral infarctions.[6] Mitral valve localization of the tumor is strongly associated with neurological events. The smaller the measures of the mass, the higher the incidence of cerebral events, but the reason for this phenomenon remains unclear. One hypothesis is the detection of postembolic CAT remnants. Surgery was performed in most cases to affirm the histopathologic diagnosis and was generally used as the treatment of choice with good results.[2] Nishiguchi *et al*. in a review on cerebral infarction due to CAT revealed that cerebral infarction can occur even with small CAT. Surgical treatment is recommended to prevent cerebral infarction recurrence, especially when CAT is accompanied by mitral annular calcification or has marked mobility.[2] In our case, the mass was referred to as the mitral strand, suggesting a differential diagnosis with fibroelastoma. The patient had a history of stroke 6 months ago and underwent several examinations to

determine the cause of the neurological event. There is no discussion that the patient with atrial fibrillation (AF) is "high risk" for cerebrovascular events. Meanwhile, the influence of CAD is not clear. There are some studies that report CAD as a univariate predictor of stroke, and in the other hand, multivariate analysis did not find the presence of CAD as an independent predictor of stroke.[7] There is a strong evidence of no casual association of AF and CAD. CAD is an independent risk factor for AF. CAD in the general population is estimated to have an incidence of about 12%–14% that is significantly lower than in patients with nonvalvular AF which goes more than 50%.[8]

Atherosclerotic carotid artery disease is one of the major causes of ischemic stroke and transient ischemic attack, responsible for approximately 10%–15% of cases.[9]

We considered the arguments above taking into account the fact that the possible cause of stroke needed to be investigated and moreover because CAD and AF may reinforce each other related to the risk of cerebrovascular events.

We have mentioned above that the patient underwent the protocol of examinations following the cause of stroke and between them 48-h rhythm Holter and computed tomography-angiography of carotid arteries which resulted in normal. In addition, the patient even during postoperative period had no episodes of AF.

Despite current risk scores being based on traditional cardiovascular risk factors, a significant number of cardiovascular events still remain unexplained. Considerable research has focused in inflammatory responses. However, the potential role of inflammatory markers such as cytokines in the risk of cerebrovascular events has yet to be thoroughly explored.[10] We did not conduct laboratory tests of inflammatory markers, an argument that I think we should consider in the future.

Given that the mass was small (2 mm ×10 mm), there was a debate among colleagues about the indication for intervention.

The prevalence of valvular strands ranges from 5.5% in the general population to 40% in patients with unknown causes of stroke.[11] The risk of embolism due to fibroelastoma, which was addressed for differential diagnosis, is high. The incidence of systemic embolism could reach 53%, including cerebral, retinal, and transient ischemic attacks. There was no difference in the thromboembolism risk between mitral and aortic valve locations.[12] Thus, it was decided to proceed with the intervention. The intervention and the postoperative period were uneventful.

Conclusion

Considering that CATs or similar masses harbor significant risks of cerebral events, surgery remains the treatment of choice. We have to be careful in investigating as much as possible the causes of cerebrovascular events with the main goal to reach the most appropriate solution for the patient case after case.

Author contributions

Selman Dumani designed the study and wrote the first draft of the manuscript with Ermal Likaj, Arben Baboci, and Altin Veshti. Laureta Dibra was part of the team, made the diagnosis and postoperative follow-up and contributed to the definitive manuscript, Majlinda Ikonomi is the anatomo-pathologist and contributed in the definitive manuscript Saimir Kuci was part of intervention team and contributed to the definitive manuscript. Each coauthor contributed to either the delivery of the study or helped to devise the protocol. All authors have given final approval for the current version to be published.

Ethical statement

Ethical statement is not applicable for this article as it is a case report. This study was conducted per the ethical principle of the Declaration of Helsinki.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Data availability statement

All data generated or analyzed during this study are included in this published article.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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