Calcified amorphous tumor presenting with rapid growth in the ascending aorta

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ABSTRACT

A calcified amorphous tumor (CAT) is a rare, non-neoplastic cardiac mass frequently located in cardiac chambers, especially the mitral valve or annulus. Here, we report an exceedingly rare case of CAT as an atypical mobile mass in the ascending aorta in a 62-year-old man who was on hemodialysis for 11 years. The CAT grew rapidly within 3 months. We resected the mass, and he was discharged with no complications. This report shows that the CAT can grow rapidly, even in the aorta, and provides important information on the progression of this rare disease and its clinical features. (J Vasc Surg Cases and Innovative Techniques 2020;6:671-3.)

Keywords: Calcified amorphous tumor; Aorta; Cardiac tumor

A calcified amorphous tumor (CAT) is a rare hamartomatous tumor. In most cases reported in the past, CAT was detected mainly on the mitral annulus or mitral valve. Here, we report an exceedingly rare case of CAT in the ascending aorta of a patient on long-term hemodialysis. The CAT grew rapidly within a 3-month period. To the best of our knowledge, there is only one report in the literature that showed CAT in the ascending aorta. This case report improves our understanding of the progress of CAT, thereby contributing to a better understanding of the clinical features of the disease. Consent for publication was obtained from the patient.

CASE REPORT

A 62-year-old man with heart failure and poor left ventricle function presented to our hospital for a regular follow-up to monitor his condition. He underwent intermittent hemodialysis for 11 years because of a total nephroureterectomy for advanced urothelial carcinoma. He had no known familial or coagulation disorders.

Regular transthoracic echocardiography showed a highly echoic mobile linear projection with an acoustic shadow in the ascending aorta. To evaluate the condition more precisely, we performed computed tomography (CT) and transesophageal echocardiography. CT images showed a highly intense threadlike object protruding from a calcification on the ascending aortic wall. The calcification on the aorta was seen on the CT image 3 months before, but the protruding mass was not detected. Transesophageal echocardiography showed a swinging tumor measuring 3.5 × 1.0 cm in the ascending aorta and located 5 cm above the sinotubular junction (Fig 1). The coagulation profile of the man was found to be within the reference range.

We assumed that this tumor had a high probability of CAT because of his long history of hemodialysis, no signs of infectious endocarditis, and especially the total calcification of the mass on the CT image. This CT image helped us rule out fresh thrombus or vegetation.

Considering the high risk of fatal embolism, we performed emergent surgery through a median sternotomy on cardiopulmonary bypass. After aortic cross-clamping, the mass was approached through a transverse aortotomy. Next, the mass was taken out in one piece after resection of its foot in the aortic wall (Fig 2). On macroscopic observation, the mass appeared yellowish white, and it was partially calcified. Histopathologic evaluation revealed eosinophilic fibrin thrombus and multiple nodular amorphous calcifications (Fig 3). There were few giant calls and chronic inflammatory cells, consistent with the report of Reynolds et al,[1] and some neutrophils. We diagnosed it pathologically as a CAT. As the clinical course of the man was satisfactory, he was discharged 11 days after surgery. His follow-up period is 9 months to date, and there are no signs of recurrence or aorta-related complications.

DISCUSSION

The CAT was first reported in 1997 by Reynolds et al.[1] After their initial report, several case reports about CAT were published, and it has been considered a cardiac intracavitary pseudotumor. A systematic review by de Hemptinne et al[2] reported that CAT has been detected in all cardiac chambers, but it is most predominantly found on the mitral valve or annulus (36%), in the right atrium (21%), or in the right ventricle (17%). Watanabe et al[3] reported a case of CAT in the sinus of Valsalva,

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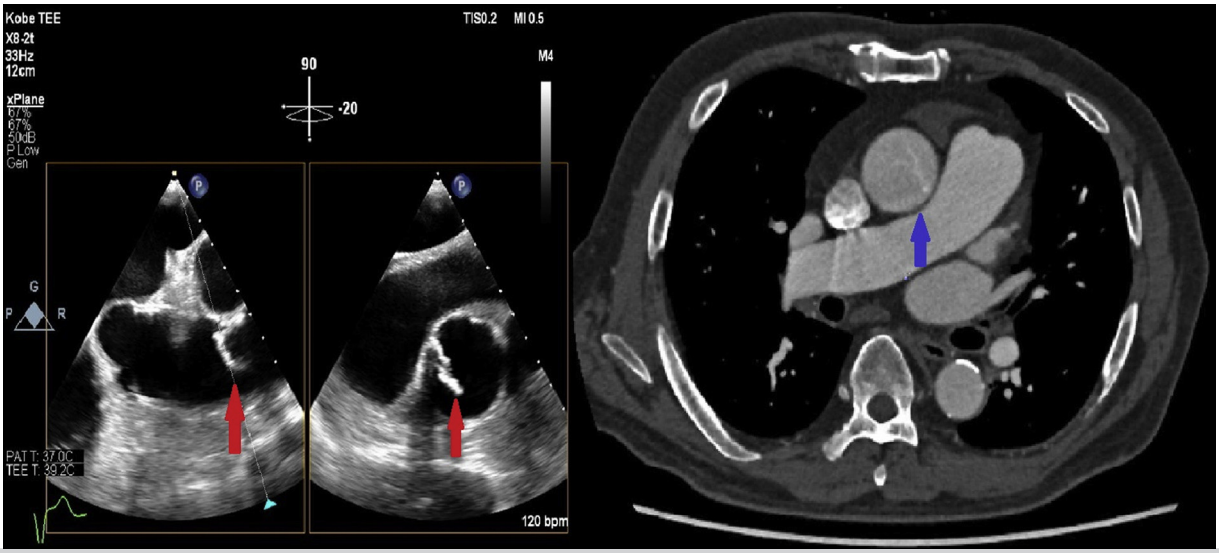

Fig 1. Transesophageal echocardiography shows the mobile mass with acoustic shadow in the ascending aorta (*red arrow*). This mass is swinging periodically with the heartbeat of the man. Computed tomography (CT) reveals the highly enhanced mass in the ascending aorta (*blue arrow*).

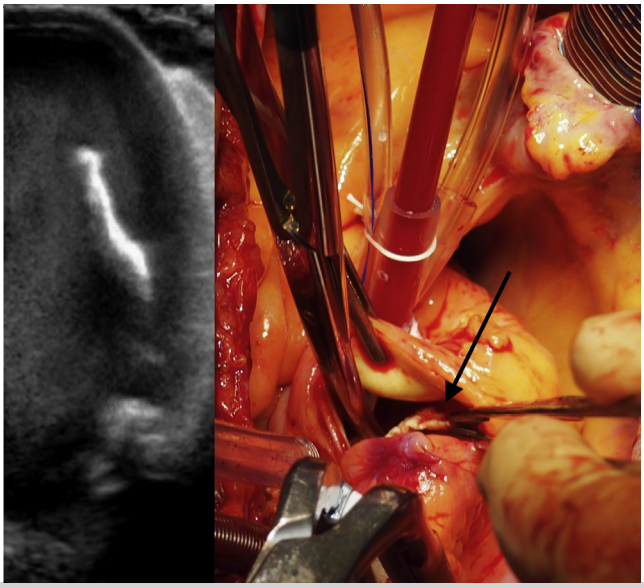

Fig 2. Epiaortic ultrasound image shows the mass protruding from calcification on the ascending aortic wall. After a transverse aortotomy, the mass is detected and resected (*arrow*).

and Ishida et al[4] reported the presence of a CAT in the ascending aorta. In this report, we present the second known case of CAT in the ascending aorta. The possibility of the emergence of a CAT in the ascending aorta has an important indication; because of the abnormal calcium-phosphate metabolism, the calcification on the aorta is common in hemodialysis patients, similar to the calcification on the mitral valve. One systematic review showed that 21% of CAT cases were detected in patients with end-stage renal disease.[2] These reports and the current report indicate that a certain number of undetected cases of CAT may arise from the calcified aorta, and there could be the possible origin of embolic events in hemodialysis patients. The best treatment strategy is controversial. However, there were several reports showing embolism from CAT,[5,6] so resection of the tumor as soon as possible is thought to be justified.

Preoperative differentiation of CAT from other diseases is challenging, and the final differential diagnosis relies on the histopathologic evaluation. This mass had unorganized fibrin, calcification, and giant cells. This patient had a long history of hemodialysis and no signs of infectious disease, expanding the possibility of CAT. In addition, thin-slice CT scan may help us diagnose CAT thanks to its total calcification. We can rule out fresh thrombus and fresh vegetation. Even though the growth rate of the tumor was so rapid, we can rule out a primary malignant tumor of the aorta because it is extremely rare, and there are no reports that show global calcification on primary malignant tumor of the aorta.[7] Differentiation of a primary benign tumor of the aorta, especially myxoma, seems to be much more difficult because some reports show both an aortic myxomatous tumor[8] and calcification of intracardiac myxoma.[9] These reports may imply the possibility of calcified myxoma in the aorta, but the surgical strategy for resection does not seem to differ for either tumor type. Because both CAT and myxoma have the possibility of recurrence, we resected the root of the tumor.

In this case, another notable point was the speed of growth of the CAT. Because of the limited evidence available on the progress of CAT, the growth rate of CAT is unclear. Kubota et al[10] reported the case of a CAT that originated from mitral annulus calcifications and

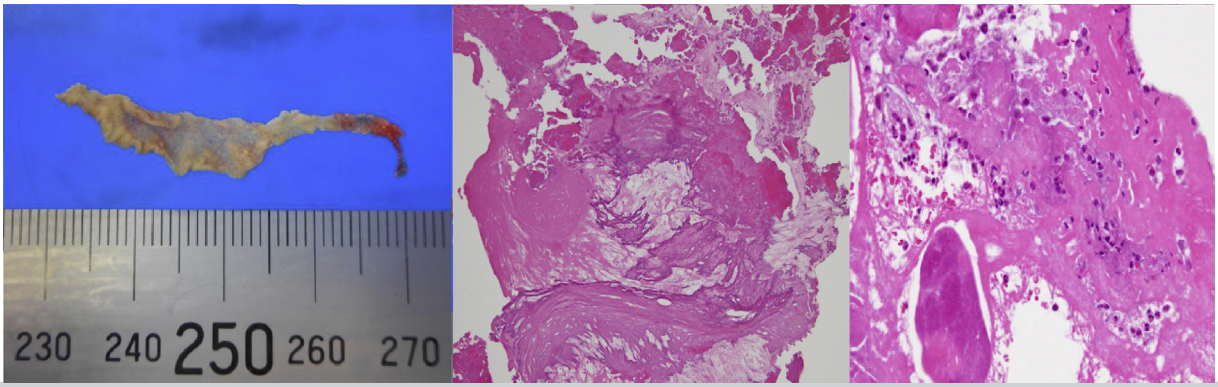

Fig 3. On macroscopic observation, the mass is yellowish white with some calcified regions. On microscopic examination, we can see the eosinophilic fibrin thrombus and multiple nodular amorphous calcifications (hematoxylin and eosin staining, magnification ×100). There are chronic inflammatory cells and neutrophils visible (hematoxylin and eosin staining, magnification ×200).

progressed rapidly within a 2-month period. The current case also shows the rapid progress of CAT in the ascending aorta within a 3-month period. Considering the previous report and the current case, we postulate that CAT can grow within a relatively short time, just a few months. Understanding the growth rate of CAT can significantly help in understanding the pathogenesis of the disease and thus can be potentially helpful in better management of the disease toward favorable outcomes. Our specimen contained neutrophils, one of the acute response inflammatory cells, showing there were mixed stages in one mass. This may be related to the growth rate of CAT; however, there are few reports that showed the cell types in CAT.

CONCLUSIONS

As the CAT is a rare tumor, its epidemiology and etiology are unclear. Further detailed pathologic findings and clinical cases should be reported to better understand how this rare tumor arises.

REFERENCES

1. Reynolds C, Tazelaar HD, Edwards WD. Calcified amorphous tumor of the heart (cardiac CAT). Hum Pathol 1997;28:601-6.
2. de Hemptinne Q, de Cannière D, Vandenbossche JL, Unger P. Cardiac calcified amorphous tumor: a systematic review of the literature. Int J Cardiol Heart Vasc 2015;7:1-5.
3. Watanabe Y, Naganuma T, Nakao T, Nakamura S. A calcified amorphous tumor originating in the sinus of Valsalva. Echocardiography 2016;33:796-8.
4. Ishida N, Shimabukuro K, Mitta S, Doi K. Thrashing petaloid flap in ascending aorta: aortic calcified amorphous tumour. Eur Heart J Cardiovasc Imaging 2019;20:244.
5. Aizawa Y, Nakai T, Saito Y, Monno K, Morikawa T, Kogawa R, et al. Calcified amorphous tumor-induced acute cerebral infarction. Int Heart J 2018;59:240-2.
6. Singu T, Inatomi Y, Yonehara T, Ando Y. Calcified amorphous tumor causing shower embolism to the brain: a case report with serial echocardiographic and neuroradiologic images and a review of the literature. J Stroke Cerebrovasc Dis 2017;26:e85-9.
7. Thalheimer A, Fein M, Geissinger E, Franke S. Intimal angiosarcoma of the aorta: report of a case and review of the literature. J Vasc Surg 2004;40:548-53.
8. Das AK, Reddy KS, Suwanjindar P, Fulmer A, Siquiera C Jr, Floten HS, et al. Primary tumors of the aorta. Ann Thorac Surg 1996;62:1526-8.
9. Lopez-Marco A, BinEsmael T, Rowlands G, Von Oppell U. Complete calcification of right atrial myxoma. Eur J Cardiothorac Surg 2015;48:171.
10. Kubota H, Fujioka Y, Yoshino H, Koji H, Yoshihara K, Tonari K, et al. Cardiac swinging calcified amorphous tumors in end-stage renal failure patients. Ann Thorac Surg 2010;90:1692-4.

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