

pected distal occlusion [3]. Said and colleagues [5] simply ligated the proximal ends of the fistula and sutured the distal ends through the pulmonary artery. A surgical strategy or technique must be selected in accordance with the size and anatomy of the aneurysm to perform safe and effective corrections, and the contribution of imaging is essential for preoperative evaluation.

Our patient was suffering from restrictive cardiac dysfunction not only because of a giant aneurysm oppressing the right ventricle, but also because of the intramyocardial calcification of the left ventricle wall due to calcified circumflex arteries. We believe that this is the first report of this complicated hemodynamic physiology in a hemodialysis patient. We believe that complete removal of the aneurysm would be necessary to release the ventricle restriction; therefore, we performed a resection of the aneurysm and closed the fistula under direct vision through the aneurysm during cardioplegic arrest. The complete removal of the aneurysm was effective in improving the patient's hemodynamics during hemodialysis, even though we left the calcified left ventricle as it was before the surgery.

In summary, we have reported a successful surgical treatment of a rare giant calcified coronary artery aneurysm with a pulmonary artery fistula in a chronic hemodialysis patient. Surgical resection should be considered for giant coronary artery aneurysm with a restrictive cardiac dysfunction.

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Pedunculated Endocardial Left Ventricular Fibroma Presenting With Cerebral and Bilateral Peripheral Embolization

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Left ventricular fibroma is a rare benign tumour of the heart. We present the case of a 24-year-old man with left

hemiplegia and bilateral popliteal artery occlusion associated with left ventricular mass. The patient underwent successful excision of a pedunculated mass attached to the trabeculae of left ventricular cavity. Histopathologic examination confirmed the presence of fibroma associated with septic thrombus. The association of fibroma and embolization is rare.

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Cardiac fibromas are considered to be the second most common benign tumors in children [1]. Under rare circumstances, they appear in adults [2]. These tumors commonly arise from the left ventricle, but any cardiac chamber can be involved. They typically grow intramurally and attain a huge size [3]. We report an unusual case of a pedunculated left ventricular fibroma in an adult arising from the endocardium of the posterior wall of the left ventricle. The tumor was associated with infected thrombus formation that resulted in central and peripheral embolization.

A 24-year-old man presented for management of a left ventricular mass. The problem dated back 3 months when the patient sustained a syncopal attack on two separate occasions. In between the attacks, he was completely symptom free. Seventeen days before his recent admission, he sustained a sudden loss of consciousness associated with left sided hemiplegia, impaired speech, and a high fever (40°C).

The result of a brain computed tomography scan was normal. He was kept under observation for 4 days and treated conservatively. On day 5, an acute onset of severe pain in the right calf associated with swelling resulted in a diagnosis of acute right lower limb ischemia, confirmed by urgent duplex ultrasound scanning. It showed bilateral limb ischemia due to occlusion of the distal half of the right popliteal artery extending to the proximal two-thirds of the infrapopliteal arteries and occlusion of the left posterior tibial, peroneal, and distal part of anterior tibial arteries. Successful right popliteal artery embolectomy was promptly performed.

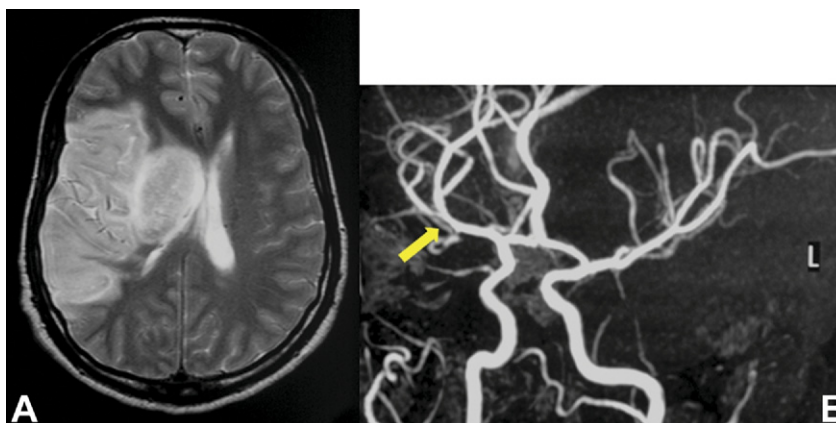
Further evaluation by magnetic resonance imaging (MRI) and angiography of the brain showed an acute right cerebral infarction in the territory of middle cerebral artery. The right middle cerebral artery appeared patent but was shifted upwards and medially by the infarcted area (Fig 1). There was no evidence of aneurysm or other vascular malformations of intracranial vessels.

A transthoracic echocardiogram showed a highly mobile echogenic mass 4 cm long and 0.6 cm at its narrowest diameter attached to the interventricular septum near the apex extending down to midleft ventricular cavity (Fig 2). There was no evidence of any valvular affection or segmental wall motion abnormalities or any cardiac chamber enlargement. The appearance of the mass was further confirmed by chest MRI. The patient received heparin, aspirin, Plavix (Bristol-Myers Squibb, Princeton, NJ), and intravenous antibiotics.

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Fig 1. (A) Magnetic resonance imaging shows an acute right cerebral infarction. (B) Magnetic resonance angiography shows the right middle cerebral artery is patent but is shifted upwards and medially by the infarcted area (arrow).



On admission, the patient was cooperative, and slightly restless because of the tingling and numbness along his right calf. Examination revealed a regular pulse of 80 beats/min, a blood pressure of 120/65 mm Hg, and a temperature of 37.5°C. Results of a chest and heart examination were normal. Femoral and popliteal pulsations were felt equally on both sides. Posttibial pulsations were felt bilaterally but were weaker on the right side. Dorsalis pedis pulsations were absent bilaterally. The right calf was slightly edematous.

Motor power examination showed affection along the left lower limb (motor power score 3/5), with complete paralysis of the left arm (score 0/5). The right side was completely normal. Sensation was intact along both limbs with hyperesthesia along the right side.

A chest roentgenogram was normal, and the electrocardiogram showed nonspecific ST-T wave changes in leads V3-V5. Laboratory investigations showed elevated levels of liver function tests, lactic dehydrogenase, and total creatine kinase. Repeated blood culture was negative. The patient received anticoagulants and antiplatelets for another 5 days.

Surgical intervention proceeded with standard cardio-

pulmonary bypass and moderate hypothermia and blood cardioplegia. The mass was exposed through a left ventriculotomy incision. An elongated 4-cm mass was found attached to the trabeculae of the left ventricle near the apex. The main bulk of the mass was firm, and whitish in color with a smooth surface, whereas the rest of the mass was irregular and reddish brown in color. The mass was attached by a narrow stalk that was easily excised (Fig 3). The rest of the ventricular cavity was free of tumor. The patient had uneventful recovery and was discharged from the hospital receiving oral aspirin and anticoagulants.

Histopathologic examination revealed a ventricular fibroma formed of proliferating fibroblasts with bland nuclei and mature collagen. The tumor was adherent to ventricular muscle and showed an adherent septic thrombus. Staining by Masson trichrome showed a positive blue stain for fibroblasts and red stain for cardiac muscle. No elastic fibers were seen by orcein and von Gieson special stain (Fig 4).

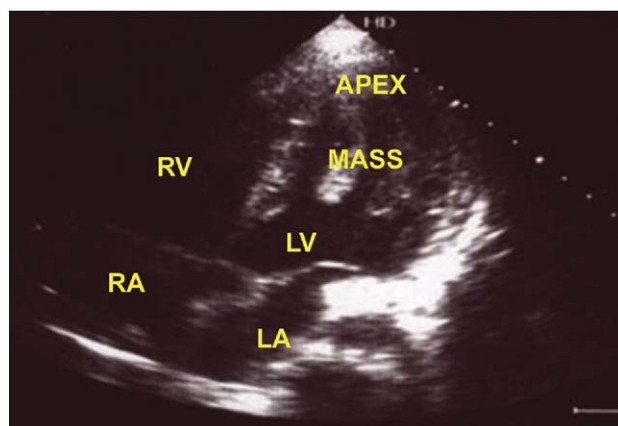


Fig 2. Echocardiography shows a left ventricular (LV) mass attached to the interventricular septum and extending to middle left ventricular cavity. (LA = left atrium; RA = right atrium; RV = right ventricle).

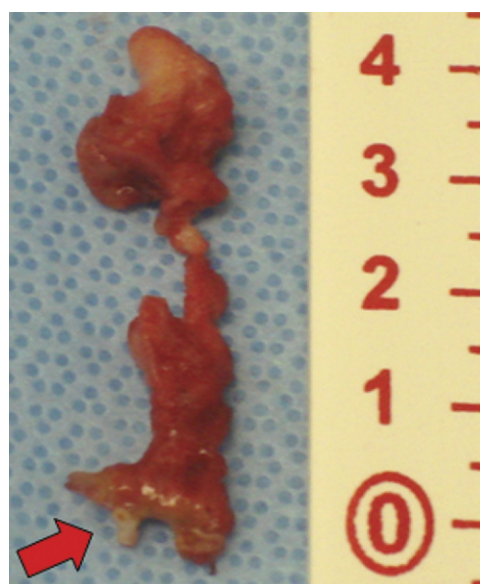


Fig 3. The excised elongated left ventricular mass was attached by narrow stalk (arrow).

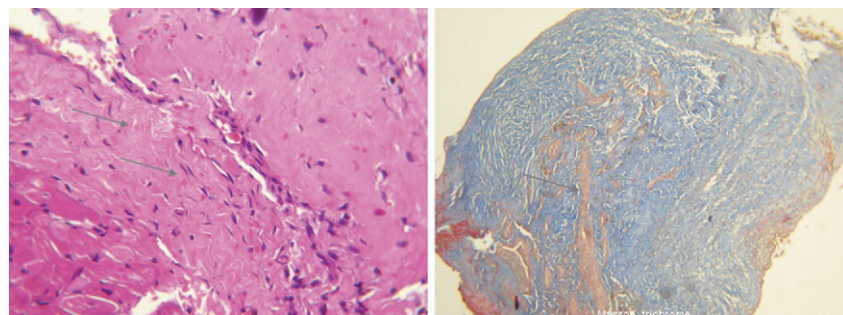


Fig 4. Histopathology. Left photomicrograph shows proliferating fibroblasts and mature collagen by hematoxylin and eosin stain (×400). Right photomicrograph shows fibroblasts (positive blue stain) and cardiac muscle (red stain) by Masson trichrome (×100).

Comment

Cardiac fibromas are rare benign tumors. They are considered the second most common benign tumors in children after rhabdomyoma [1]. The incidence varies from 2% to 5% of primary benign cardiac tumors [4]. They most commonly arise from the left ventricle, occupying the free wall or interventricular septum [3, 5], but any cardiac chamber and even the pulmonary artery can be involved [3, 6]. These tumors originate from fibroblasts and myofibroblasts; this explains the predilection to the left ventricle, which is rich in fibroblast interstitium [3].

More than 80% of these tumors occur in children. Adult cases are exceptionally rare [2]. These tumors usually arise within the myocardium, very rarely from endocardium [5, 7, 8]. Intramural tumors usually grow rapidly and attain a huge size. Those arising from the endocardium are smaller in size and pedunculated in shape [5, 8]. Morphologically, the tumors have clear, defined borders, are usually solitary, firm, grey-white in color, and do not have a distinct capsule [3]. Because these tumors grow rapidly, they compress and entrap islands of myocardium, simulating infiltration [3, 9].

Most fibromas produce symptoms due to obstruction, interference with contraction, or arrhythmias depending on the location and size of the tumor. Sudden death has been reported in 23% due to fatal ventricular tachycardia. Symptoms often include dyspnea, syncope, easy fatigability, and chest pain [3, 7], but 36% of patients may remain asymptomatic and are discovered during operation [3, 8]. The main presentation of our patient was systemic embolization both to the central nervous system and peripheral arteries.

The association of fibroma and embolization is rare. The atypical morphology of fibroma in our case regarding its endocardial origin, free mobility, pedunculated appearance, and the association of infection all predispose to thrombus formation and subsequent embolization.

The diagnosis of the left ventricular mass in our patient was based primarily on echocardiography. Further MRI studies did not help to define the tissue characteristics of the mass. The pedunculated appearance of the tumor and the association of embolization led us to think about more common lesions such as myxoma, fibroelastoma, or left ventricular thrombus. Definite diagnosis was made only after excision of the mass. Absence of elastic tissue and myxoid matrix excluded the presence of such conditions. The presence of an organized thrombus was also

excluded because the patient had no predisposing factors leading to thrombus formation. His echocardiography showed neither valvular affection nor segmental wall motion abnormalities. His pulse rate was regular all through. The evaluation for a coagulation defect showed no abnormalities.

The behavior of cardiac fibromas is unpredictable [3]. Some patients may remain asymptomatic, with no change in the size of the tumor during follow-up; others may die suddenly due to rhythm disturbances. It is very difficult to predict the consequences; thus, surgical excision appears to be the optimal treatment for symptomatic and asymptomatic patient. In most patients the resection is complete, with excellent prognosis. In larger tumors with unusual location, the resection is difficult and cardiac transplantation may be required [3]. Our patient had an excellent result due to small size of the tumor with an approachable site and no evidence of any valvular involvement.

In conclusion, we believe that atypical forms of cardiac fibromas can be associated with embolization and should be considered in the differential diagnosis of masses arising from the left ventricle and presenting by embolization as myxoma, fibroelastoma, and left ventricular thrombus.

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