SIR 2005 Film Panel Case: Peripheral Embolization From Cardiac Myxoma

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HISTORY

A 39-year-old man presented with the acute onset of a cold, painful, and numb left leg. Physical examination in the emergency room demonstrated a palpable left femoral pulse and absent left popliteal, dorsalis pedis, and posterior tibial artery pulses. The left foot and lower leg were pale and cool to touch, and the patient exhibited diminished sensation over the anterior foot and decreased mobility at the ankle joint. Vascular examination of the right lower extremity was entirely normal. Past medical history was remarkable only for recreational drug use consisting mainly of cocaine. An electrocardiogram showed normal sinus rhythm and angiography was requested.

Runoff arteriography was performed via a right femoral approach (Fig 1) and then a thoracic arteriogram was obtained (Fig 2).

RADIOGRAPHIC FINDINGS

The runoff arteriogram displays abnormalities at multiple levels all confined to the left lower extremity. There is a well-circumscribed lobulated filling defect in the distal left common femoral artery extending into the proximal superficial femoral (SFA) and profunda femoris arteries consistent with a saddle embolus (Fig 1d, arrow). The profunda and SFA fill normally beyond the occlusion. The left SFA and popliteal arteries are normal but there is abrupt occlusion of the tibioperoneal trunk at the origin of the peroneal (Fig 1f, arrow) and abrupt occlusion of the anterior tibial artery in the mid-calf (Fig 1g, arrow). There are no collateral vessels around any of the occlusions indicating that these findings are acute in nature. By comparison, the right lower extremity runoff vessels are entirely normal.

The thoracic and abdominal aorta are normal for the patient’s age without evidence of significant vascular disease (Fig 2). Specifically, there are no abnormalities in the thoracic or abdominal aorta to indicate a source for distal emboli.

TREATMENT AND FOLLOW-UP

The patient underwent systemic anticoagulation and taken urgently to the operating room. A left femoral embolectomy was performed through a groin incision. Well-organized, gelatinous material was removed from the bifurcation of the common femoral artery. A longitudinal arteriotomy was then created in the distal popliteal artery and the anterior and posterior tibial arteries were cleared of the same gelatinous material. On completion of the procedure, there was massive swelling in the lower extremity, and a four-compartment fasciotomy was performed.

Transesophageal echocardiography performed in the operating room revealed normal results and did not demonstrate any cardiac abnormalities. Similarly, postoperative thoracic magnetic resonance (MR) imaging was normal without evidence of thrombus or masses within the heart. The left-sided chambers were normal in morphology and signal intensity, and the cardiac wall motion was normal.

On pathologic examination, specimens from the left femoral artery bifurcation and the calf vessels demonstrated similar findings. The gelatinous material consisted of a multi-lobulated mass with focal frond-like structures. Histologically, the mass was comprised of bland spindle and stellate cells with eosinophilic cytoplasm and small nuclei without atypia embedded within an abundant myxoid matrix. The periphery of the mass and vascular channels within it were lined by a single layer of endothelial cells. Immunohistochemical staining was positive for calretinin and negative for desmin within the spindle cells. The endothelium was highlighted by a positive CD31 stain.

DIAGNOSIS

Peripheral embolization from cardiac myxoma.

DISCUSSION

The clinical presentation and angiographic findings in this case are pathognomonic for acute arterial embolism to the left leg (1). Clinically, the patient manifested the cardinal signs of acute
arterial ischemia (referred to in the vernacular as the “5 P’s”): pulselessness, pain, pallor, paresthesia and paralysis (1). Angiographically, the vascular abnormalities were characteristic of embolic disease in both location and morphology. Occlusions occurred predominantly at vessel bifurcations and collaterals were absent. A well-circumscribed filling defect was present at the level of obstruction but the surrounding arteries were otherwise normal. Angiographic abnormalities were limited to a single extremity. All of these findings suggest that an embolus initially lodged at the left common femoral artery bifurcation and then showered smaller embolic fragments downstream.

The unusual aspect of this case is not the presence of peripheral embolism but rather the cause of the embolism. Peripheral emboli are usually thrombotic in nature. Most (80%–90%) originate in the heart in patients with atrial fibrillation or myocardial infarction; a minority (5%–10%) originate in the thoracic or abdominal aorta. Occasionally, thromboembolism is caused by venous thrombus that has embo-

Figure 1. Preoperative aortogram and runoff arteriography shows multiple abnormalities isolated to the left lower extremity. (a) Abdominal aortography shows normal results. (b, c) Bilateral pelvic oblique views reveal a filling defect in the left common femoral artery. (d) A well-circumscribed filling defect is seen in the distal common femoral artery extending into both the SFA and profunda. The profunda and SFA fill normally beyond the occlusion. (e) The left SFA and popliteal arteries are normal. (f) There is abrupt occlusion of the tibioperoneal trunk at the origin of the peroneal artery. (g) The reconstituted posterior tibial artery is patent to the foot but there is abrupt occlusion of the anterior tibial artery in the mid calf.

Figure 2. Thoracic aortography shows normal results without abnormalities to suggest a source for distal emboli. (a) Arch aortogram in left anterior oblique projection. (b) Lower thoracic aortogram in anteroposterior projection.
lized through the heart (paradoxical embolism). All of these conditions usually occur in older patients. Thromboembolism in a young patient may suggest the presence of an underlying hypercoagulable state.

Our patient possessed no specific risk factors for thromboembolic disease (ie, no history of atrial fibrillation, myocardial infarction, venous thrombosis or hypercoagulable state). He did have a history of cocaine abuse. Cocaine can cause vasoconstriction resulting in myocardial infarction, stroke, and peripheral ischemia but is not a common cause of thromboembolism. In a young patient without obvious risk factors for peripheral thromboembolism, other less common causes of embolic disease could be considered. Chief among these are primary vascular tumors or metastatic tumors invading the vascular system.

Cardiac atrial myxomas, although rare, are the most common primary benign tumor of the heart, with an estimated incidence of 0.5 per million population per year (2). They occur more frequently in women, and most commonly between the ages of 30 and 60 years (3,4). About 75% of cardiac myxomas are located in the left atrium, and left atrial myxoma is a rare but known cause of stroke or acute limb ischemia (5). Clinical presentation is related to obstruction of the valvular or venous orifices of the heart (dyspnea, atrial fibrillation, dizziness, syncope, palpitations, pulmonary edema, or congestive heart failure), constitutional symptoms from the elaboration of humoral substances (ie, interleukin-6), and embolization (6,7). Interleukin-6 has been detected in tissue cultures from cardiac myxoma cells, and several studies have shown a relationship between IL-6 plasma levels and the occurrence of a constitutional syndrome that disappeared once the myxoma had been excised (7–9). Published figures for embolism range from 6% to 43% (11,12), with most studies demonstrating a 28% to 29% rate of embolization (5,6,12). Embolism is much more common in cerebral arteries than in the visceral or peripheral circulation (5,6).

The simplest and most reliable diagnostic evaluation of cardiac myxoma is transthoracic two-dimensional echocardiography. Transthoracic echocardiography is as sensitive as 97% sensitive for the detection of cardiac
myxomas, and transesophageal echocardiography approaches 100% sensitivity (13–17). In cases of massive embolism, the cardiac tumor can disappear. Myxomas frequently arise from a narrow stalk and have a predilection for the interatrial septum. The most important distinguishing feature of a myxoma at echocardiography is the characteristic narrow stalk, followed by tumor mobility and distensibility (18). Myxomas demonstrate variable internal echocardiographic features. They may be homogeneously, have central areas of hyperlucency representing hemorrhage and necrosis, or have echogenic foci of calcification (18).

Computed tomography (CT) and MR imaging may be useful modalities if echocardiography is not diagnostic. CT and/or MR imaging often cannot distinguish the thin delicate stalk of myxomas, but a generally narrow base of attachment can usually be seen. A cardiac myxoma can be diagnosed with a high degree of confidence on CT or MR when a mass is detected arising from the interatrial septum from a narrow base of attachment (18). Myxomas usually have heterogeneous low attenuation on CT imaging because of the gelatinous nature of these tumors, and calcification is frequently seen. Myxomas tend to have markedly increased signal intensity on T2-weighted MR images. However, because of calcification and the presence of hemosiderin, they may have areas of decreased signal intensity or magnetic susceptibility artifacts (18,19).

Pathologically, cardiac myxomas can be classified on the basis of gross characteristics. Myxomas with villous, papillary, or frond-like structures, as in this case, or those with a roughened or filiform luminal surface are prone to embolize (Fig 3) (20). In contrast, tumors with a smooth, firm surface are less likely to embolize. Cardiac myxomas are true neoplasms and arise from primitive stromal cells that have the capacity to differentiate along many cell lineages. Recently, various cardiomyocyte-specific transcription factor genes have been identified, suggesting the cells of origin to be mesenchymal cardiomyocyte progenitor cells (21). Calretinin is a calcium-binding protein expressed in fetal heart cells, thought to function as a calcium modulator (22). Antibody to calretinin can be used as a marker to distinguish a cardiac myxoma embolus from a myxoid thrombus (23).

The treatment of choice for cardiac myxomas is complete surgical removal (24). Even in asymptomatic patients, embolic complications or sudden death can occur. Special care at the time of operation should be taken to avoid intraoperative systemic or pulmonary embolization of the myxoma. Postoperative complications are comparable to other cardiac operations, with similar morbidity and mortality rates. The prognosis for patients with solitary myxomas after surgical resection is excellent (6,24,25).

The risk of recurrence in sporadic tumors is approximately 1% to 3% (3). Recurrences have been reported from as soon as a few months to as long as 22 years after excision of the myxoma (26). The cause of recurrence is not clear, but risk factors most likely include inadequate or incomplete resection, intracardiac implantation, intraoperative spillage of tumor material, and multicentricity of the tumor (5). Patients with familial cardiac myxomas have a higher recurrence rate and are more likely to have multifocal tumors (27). All patients require life-long routine surveillance echocardiography after surgical resection, probably at 5-year intervals. Patients with multifocal, atypical, or familial myxomas should be followed at shorter time intervals.

SUMMARY

Although myxomas are rare, they should be considered in the differential diagnosis of peripheral embolic disease, especially when an embolic event occurs in a young adult without evidence of endocarditis or an arrhythmia. Echocardiography is the modality of choice for diagnosis and follow-up of the tumor. Complete surgical removal is required and when embolectomy is performed, the removed material should always be sent for pathologic examination. Patients require lifelong monitoring for recurrence.

References

16. Obeid AI, Marvasti M, Parker F, et al. Comparison of transthoracic and-