Multiple Cerebral and Systemic Aneurysms in a Case of Left Atrial Myxoma – Are They Related?

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Abstract

Cardiac myxomas are known to be associated with cerebral aneurysms, however the mechanism of aneurysm formation is still debatable. Myxomatous tumor emboli to the vessel wall have been propounded as the plausible mechanism for aneurysm formation. Recent literature also implicates raised interleukin-6 (IL-6), secreted by atrial myxomas, in the genesis of aneurysms. We describe a patient who presented with mild persistent headache, later diagnosed to be harbouring multiple unruptured cerebral and systemic aneurysms. Our case highlights the importance of radiological modalities in timely diagnosis of myxoma with its cerebral and systemic embolic complications.

Keywords

Atrial myxoma, Aneurysm, Angiography, Interleukin-6

Introduction

Cardiac myxoma is a mesenchymal tumour and is the most common primary cardiac neoplasm [1]. As it usually arises in left atrium, systemic emboli from myxoma can spread to both cerebral and systemic vasculature, lead to ischemic, haemorrhagic complications and rarely aneurysms formation [2–4]. Aneurysms associated with myxomas are usually multiple, fusiform and peripheral [5, 6]. The embolization of tumor particles to vessel wall has been ascribed in weakening of subintimal tissue resulting in aneurysm formation [7]. Recent reports have also implicated the role of IL-6 in aneurysm genesis in patients harbouring atrial myxomas. IL-6 is a pro-inflammatory cytokine that has also been implicated in the development of cerebral aneurysms [8, 9]. Neurological manifestations in a patient presenting initially, may lead to discovery of primary tumor i.e. cardiac myxoma in the course of investigations and diagnosis, as in our case. We report a case of an adult male presenting with headache. Imaging and angiography revealed multiple cerebral & systemic aneurysms along with left atrial myxoma on echocardiography.

Case Report

A 38-year-old adult male patient presented with complaint of intermittent headache. NCCT (Figure 1a) and CECT (Figure 1b) were done, which showed iso to hyperdense globular lesions along bilateral sylvian fissures and other sulcal
spaces showing homogenous post contrast enhancement. MRI was done for further evaluation, showing corresponding T1 hyperintensity (Figure 1c) and T2 hypointensity (Figure 1d) within the lesions with homogenous post contrast enhancement (Figure 1e) raising the suspicion for vascular lesions. Patient was posted for diagnostic cranial angiography using biplane digital subtraction angiography (DSA). Angiography (Figure 1f-j) showed numerous (upto 20) globular and fusiform outpouchings and focal vascular ectatic segments in bilateral anterior and posterior circulation suggestive of aneurysms. To rule out the possibility of mycotic aneurysm from valvular vegetations, echocardiography was done. Echocardiography (Figure 2a) showed a large broad based ~ 4.8 x 3.4 cm sized homogenously echogenic lesion in left atrium. CT angiography was done to rule out any systemic complications. CT angiography shows a large hypoenhancing mass in left atrium (Figure 2b), few (upto 3) peripheral hepatic artery aneurysms (Figure 2c), distal superior mesenteric artery branch aneurysm (Figure 2c) and single left renal artery aneurysm at hilum (Figure 2d). Surprisingly the patient had normal serum IL-6 levels (6 pg/ml), as opposed to many literature documentations.

Discussion

Cardiac myxoma is a mesenchymal tumor and accounts for half of all primary cardiac neoplasms, predominantly in (75%) left atrium [10]. Patients with cardiac myxoma usually presents with cardiac (60%), constitutional and embolic symptoms. Constitutional symptoms include anemia, weight loss, fever, elevated erythrocyte sedimentation rate, hypergammaglobulinemia and leukocytosis. Cardiac symptoms can be ascribed to impediment of myocardial function by left atrial tumor and comprise of dyspnea, palpitations and syncope [11]. Due to their proclivity for left atrium, systemic embolism is quite common. The neurological signs and symptoms in a patient of left atrial myxoma results from embolization, out of which cerebral ischemic & haemorrhagic changes, brain parenchymal metastasis and oncotic aneurysm formation are most common [12]. Ischemic infarcts due to vascular occlusion from embolised myxomatous particles have also been documented in various literature reports [13, 14]. First case of cerebral aneurysm associated with cardiac myxoma was reported by Stoane et al. [15]. Cerebral aneurysms can be diagnosed with CT and MRI angiographies, but catheter angiography is gold standard. Cerebral vascular aneurysms associated with myxoma are usually fusiform (may have saccular element) and peripherally located [16].

Cerebral aneurysms dilatations and vessel irregularities in patients of cardiac myxomas have been linked to neoplastic properties of a myxoma and embolism of tumor particles to vessel wall. The embolic oncotic particles deposit in the vessel wall, penetrate the vasa vasonum, causing weakening, ectasia and aneurysmal dilatation of vessel wall as has been demonstrated by various studies [17, 18].

Many studies have supported the fact that atrial myxoma cells are capable of producing IL-6. IL-6 is a cytokine which causes differentiation & proliferation of cells along with overexpression of several proteolytic enzymes that degrade extracellular matrix (ECM), promoting fragmentation of tumor and acceleration of embolic event. Formation of cerebral aneurysm may be related to overproduction of IL-6 by tumor emboli causing degradation of ECM in cerebral vessels [19-21].

Available literature suggests that the treatment of choice for atrial myxomas is surgical removal. Surgery should be conducted at the earliest so as to prevent the possible risk of further tumour embolism, cardiac valve obstruction and improving the cardiac function. Few literary evidences have also suggested presumably decreased number and size of aneurysms following myxoma resection. However, myxoma resection does not eliminate the risk of delayed aneurysm formation regardless the mechanism of development either by embolization or metastasis [22]. For this reason, follow-up monitoring for the development of aneurysms, using non-invasive imaging (MRI/MRA) is recommended after intervention for myxoma resection [23]. No definitive treatment has been advised, as of now, for aneurysms due to cardiac myxomas except for active observation.

Our patient has been under follow up of cardiac surgery department for atrial myxoma, under interventional neuroradiology and neurosurgery department for intracranial aneurysms. Up to recent OPD follow up, patient has been stable, been advised for myxoma resection and detailed about the prognosis. For size and numbers of intracranial aneurysms the patient is being followed up with MRA every 6 months.
under neurosurgery and neuro radiology department.

Multimodality radiological evaluation should be done in the patients presenting with multiple cerebral aneurysms, especially echocardiography to rule out the mycotic (valvular vegetations) or oncotic (atrial myxoma) etiologies and angiography (CTA, MRA or catheter angiography) to define the location and number of cerebro-systemic aneurysms. Patients with multiple cerebral aneurysms should routinely be assessed for cardiac myxoma and vice-versa.

References


