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Oncotic Cerebral Aneurysms in a Case of Left Atrial Myxoma, Role of Imaging in Diagnostics and Treatment

Madan Mohan Gupta^{1A}, Neha Agrawal^{2B}

¹ Department of Interventional Neuroradiology, Jaslok Hospital and Research Center, Mumbai, India

² Department of Critical Care Medicine, Jaslok Hospital and Research Center, Mumbai, India

Author's address: Madan Mohan Gupta, Department of Interventional Neuroradiology, Jaslok Hospital and Research Center, Mumbai, India, e-mail: madan_gupta05@yahoo.in

Summary

Background:	Myxomatous cerebral (oncotic) aneurysms following atrial myxoma is a rare neurological complication.
Case Report:	We report an 11-year-old boy with left atrial myxoma and multiple cerebral oncotic aneurysms. The characteristics of these aneurysms are indefinite and variable. The "Metastasize and Infiltrate" theory may be the key mechanism in the formation of these aneurysms.
Conclusions:	Magnetic resonance imaging (MRI), computed tomography (CT) and angiography are useful in the diagnostics while digital subtraction angiography (DSA) is the best option. There are no definite guidelines for therapy of these aneurysms. Resection of cardiac myxomas, chemotherapy, radiotherapy, coil embolization and surgical treatment could be helpful.
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Background

Cardiac myxoma is the most common primary benign tumor of mesenchymal origin in the heart. More than 80% of these tumors are found in the left atrium. Patients with cardiac myxomas usually present a variety of neurological syndromes even in the absence of cardiac symptoms. Systemic embolus is the second most frequent presentation (29%) in patients with left cardiac myxomas [1]. There were reports of the embolisms of cerebral vessels, coronary and renal arteries, spleen, mesenteric arteries, and arteries of the lower limbs [2]. The emboli are most frequently related to cerebral vessels with a stroke (21%). The mobility, but not the size of myxomas may be related to embolic potential [1]. Although cardiac myxoma-related cerebral emboli are well documented, there are also other rare neurological complications such as cerebral aneurysms, myxomatous metastasis [3] and cerebral cavernous malformations [4]. Here we describe a patient with multiple cerebral aneurysms associated with cardiac myxoma and we review the existing literature to describe imaging and angiographic findings for diagnosis and therapy of the disease.

Case Report

An 11-year-old boy presented with a history of a sudden loss of consciousness followed by vomiting, the day before. His past medical history revealed right-sided hemiplegia and UMN facial palsy one year earlier. On examination, the patient was conscious, oriented to time, place and person. He had hemiplegic gait, increased muscle tone and decreased power on the right side.

During last hospitalization, an MRI study was performed to define the extent of his neurological lesions. An acute infarct was noted on T2, FLAIR and DW images in the left gangliocapsular region (Figure 1A, 1B). MR Angiogram showed complete cut-off of segment M1 of the left middle cerebral artery (MCA) and an aneurysm in the intradural segment of the right vertebral artery (Figure 2). After one month, follow-up MRI with MRA showed gliotic area in the left gangliocapsular region and a partially recanalized, tortuous, narrowed left MCA (Figure 3A, 3B).

On digital subtraction angiography (DSA), left MCA appeared irregular, narrowed, and tortuous, and multiple

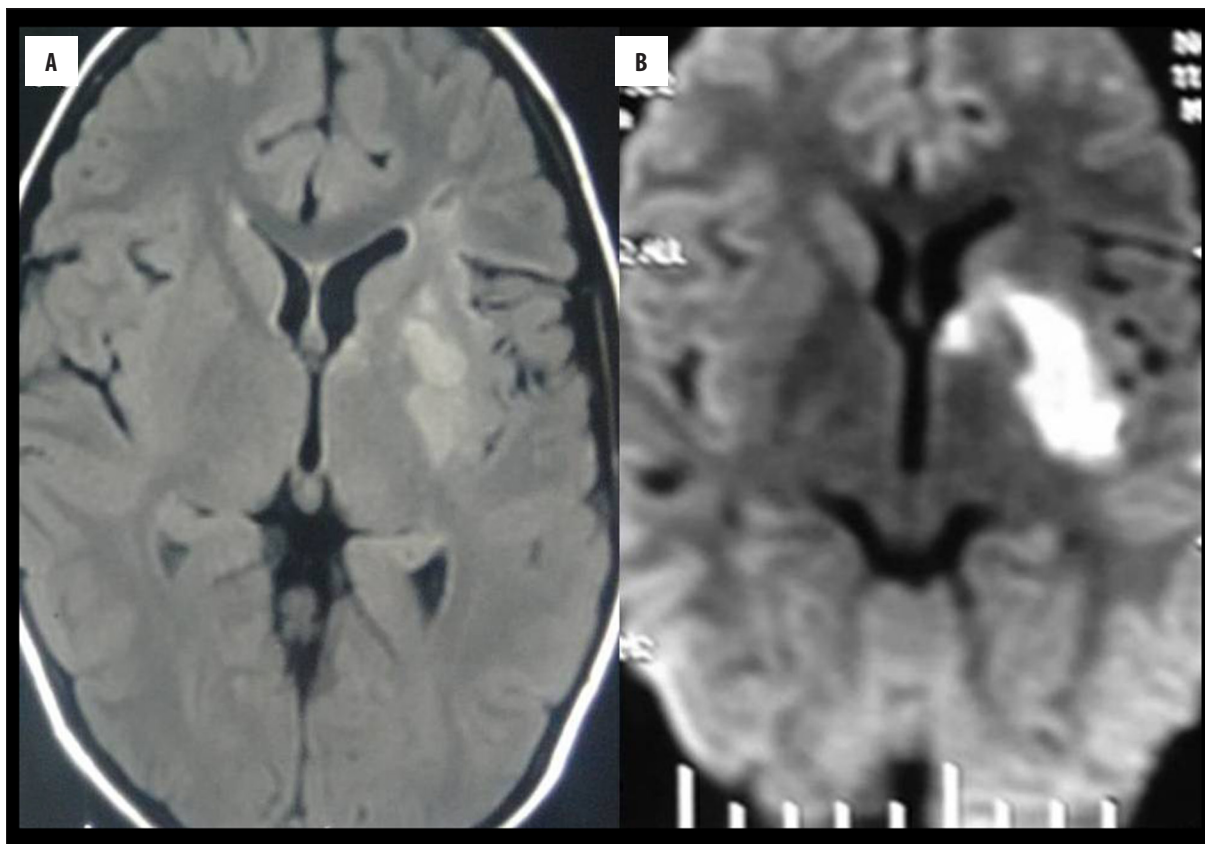


Figure 1. An 11-year-old boy left atrial myxoma with oncotic aneurysms; (A) FLAIR image of the brain shows acute infarct in the form of hyperintensity in the left gangliocapsular region. (B) Diffusion weighted image shows restricted diffusion in the corresponding area.

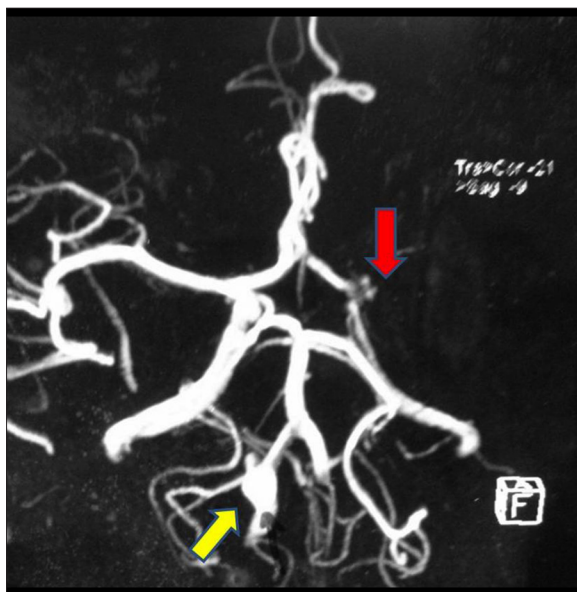


Figure 2. An 11-year-old boy with left atrial myxoma and oncotic aneurysms; MR Angiogram (Time of flight sequence) shows complete cut-off of M1 segment of the left middle cerebral artery (MCA) (red arrow) and an aneurysm in the intradural segment of the right vertebral artery (yellow arrow).

aneurysms were seen in distal branches of bilateral MCA (right>left) (Figures 4, 5) and in the intradural segment of the right vertebral artery (Figure 6A, 6B). The lesions

had focal fusiform morphology suggestive of myxomatous aneurysms. Further on the patient was sent for 2-D echocardiogram. Doppler echocardiography showed a left atrial myxoma, 5.2×5.1×2.1 cm in size, which originated from the inferior inter-atrial septum and prolapsed into the left ventricle (Figure 7A). On the basis of the above findings, the final diagnosis was left atrial myxoma with multiple myxomatous cerebral (oncotic) aneurysms. Then the patient was sent for cardiac surgery for removal of that myxoma. The histopathological examination of the specimen revealed findings of myxoma in the form of spindle-shaped lipidic cells in a loose myxoid stroma (Figure 7B). The patient was lost for follow-up, so control echocardiogram was not performed.

Now, one year later, the patient presented with syncope. Recent computed tomography showed infarcts (not visible one year before) in the head of the right caudate nucleus and the right high parietal region, and chronic infarct in the left gangliocapsular area (Figure 8A, 8B). CT angiography showed tortuous, dilated and fusiform left MCA (Figure 9A) and multiple aneurysms in bilateral MCA and both vertebral arteries (Figure 9B).

Discussion

Cardiac myxomas are neoplasms of endocardial origin and are mostly found in the left atrium. About 10% of all patients with a myxoma of this location remain completely asymptomatic. Echocardiographic examination revealed a

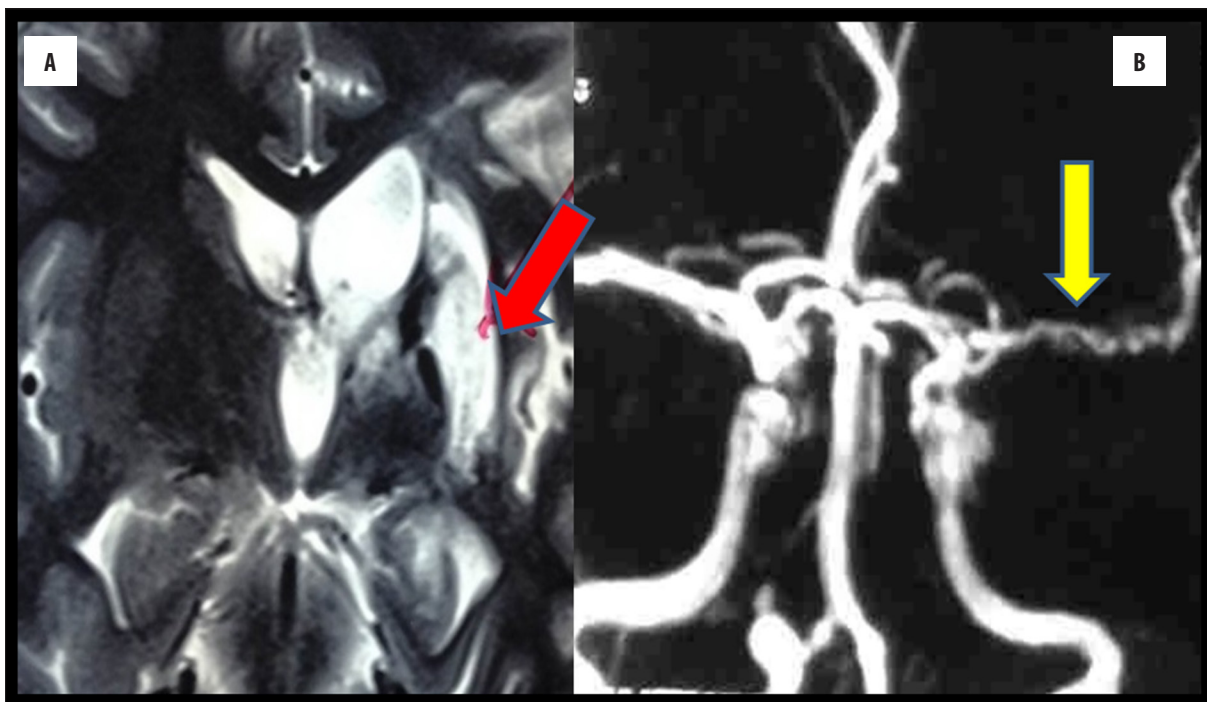


Figure 3. An 11-year-old boy with left atrial myxoma and oncotic aneurysms; Follow-up MR, (A) shows gliotic area in the left gangliocapsular region (red arrow). (B) MR Angiogram (Time of flight sequence) shows partially recanalized, tortuous, narrowed left MCA (yellow arrow).

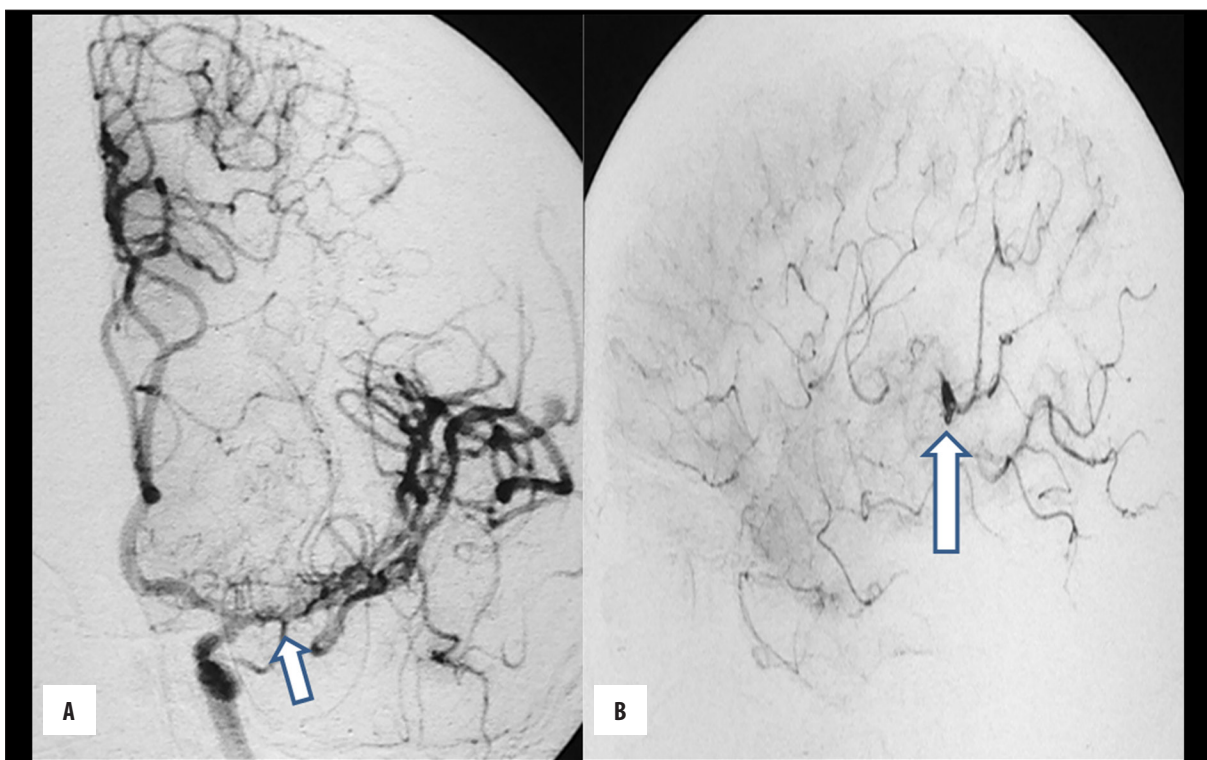


Figure 4. An 11-year-old boy with left atrial myxoma and oncotic aneurysms; Digital subtraction angiography (DSA), Left ICA angiogram shows – (A) irregular, narrowed and tortuous left MCA (small white arrow). (B) An aneurysm in the distal branch of left MCA (large white arrow).

large tumor that almost completely obliterated the mitral valve [1–3].

Intracranial aneurysms are rarely associated with cardiac myxoma. Up to a half of cardiac myxomas produce

systemic emboli. Emboli from cardiac myxomas can lead to cerebral ischemia, infarction, and aneurysm formation. Of these complications, ischemic stroke is the most common, and it has been estimated that 0.5% of all strokes are caused by myxomatous emboli. Strokes caused by

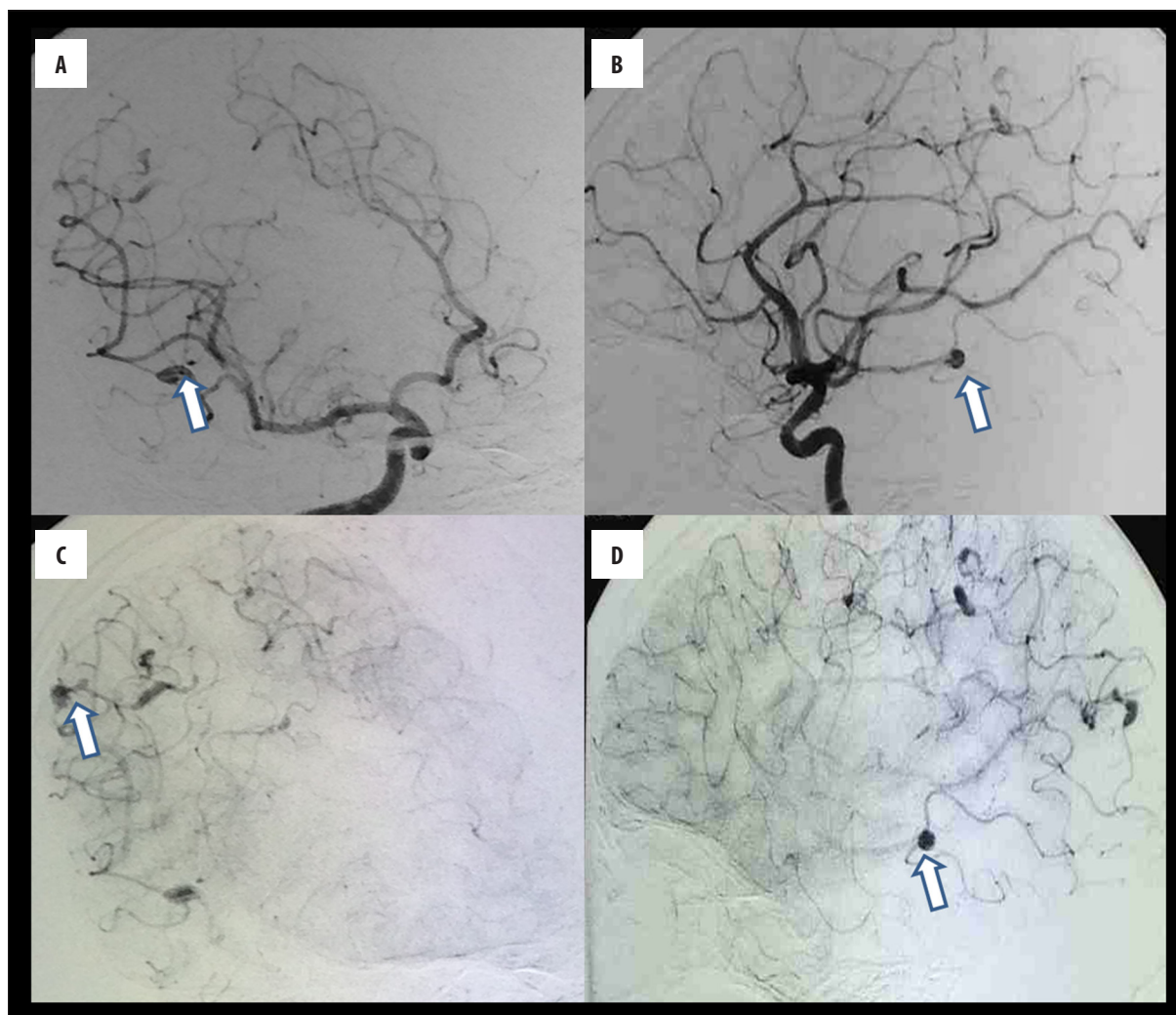


Figure 5. An 11-year-old boy with left atrial myxoma and oncotic aneurysms; Digital subtraction angiography (DSA), Right ICA angiogram (A–D) – show multiple aneurysms in distal branches of the right middle cerebral artery (white arrows).

cardiac myxoma most commonly occur in the middle cerebral artery [5]. In strokes secondary to cardiac myxoma, mechanical thrombectomy might represent a safe and effective treatment option [6]. In the setting of acute paediatric stroke due to atrial myxoma emboli, mechanical thrombectomy may be the first-line therapy [7].

These aneurysms are usually multiple with fusiform or saccular shape, but mostly fusiform (91%). In the present case, fusiform and saccular aneurysms were both observed. Most of the aneurysms are located in the distal branches of both sides of the middle cerebral artery. The mechanism of the disease has not been well understood. The theory “Metastasize and Infiltrate” is mostly recognized. Fragments of myxomas can metastasize to the brain. Then the myxomatous cells may infiltrate the vessel walls via mitotic and proliferating activity within the vessel and interrupt the elastic lamina, leading to vessel dilatation and aneurysm formation. Patients with multiple cerebral aneurysms should prospectively receive echocardiography for myxoma. CT, MRI and cerebral angiography are elective methods in the diagnostics of myxomatous cerebral aneurysms [8].

On MRI, the lesions may be characterized by tubular dilations of arteries within the sulci. The lesions appeared as contrast-enhancing focal dilations of distal segments of intracranial arteries on T1-weighted images. That may be a result of slow flowing inside the aneurysms or possibly enhanced myxoma tissue within the aneurysm wall. On T2-weighted images, they are characterized by low signal intensity flow voids, sometimes associated with cerebral infarctions [9]. Magnetic resonance angiography (MRA) may confirm the nature of these lesions. Myxoma-related cerebral aneurysms may appear as hyper-density on CT. CTA in a high resolution CT scanner is necessary when MRA is likely insufficient to rule out a myxomatous aneurysm. Cerebral DSA may be a better choice comparing with CT or MRI. DSA is free from the disturbance of edema or hemorrhage, demonstrating the dilated vessel and peripheral distribution of the cerebral arteries clearly. Cerebral aneurysms can appear as irregular fusiform outpouchings or saccular dilations on angiography [10,11]. The greatest part of aneurysms are located on the middle cerebral artery (74.2%), then anterior cerebral artery (13%), cerebellar arteries (7%), posterior cerebral artery (5%) and the least is basilar artery (1%).

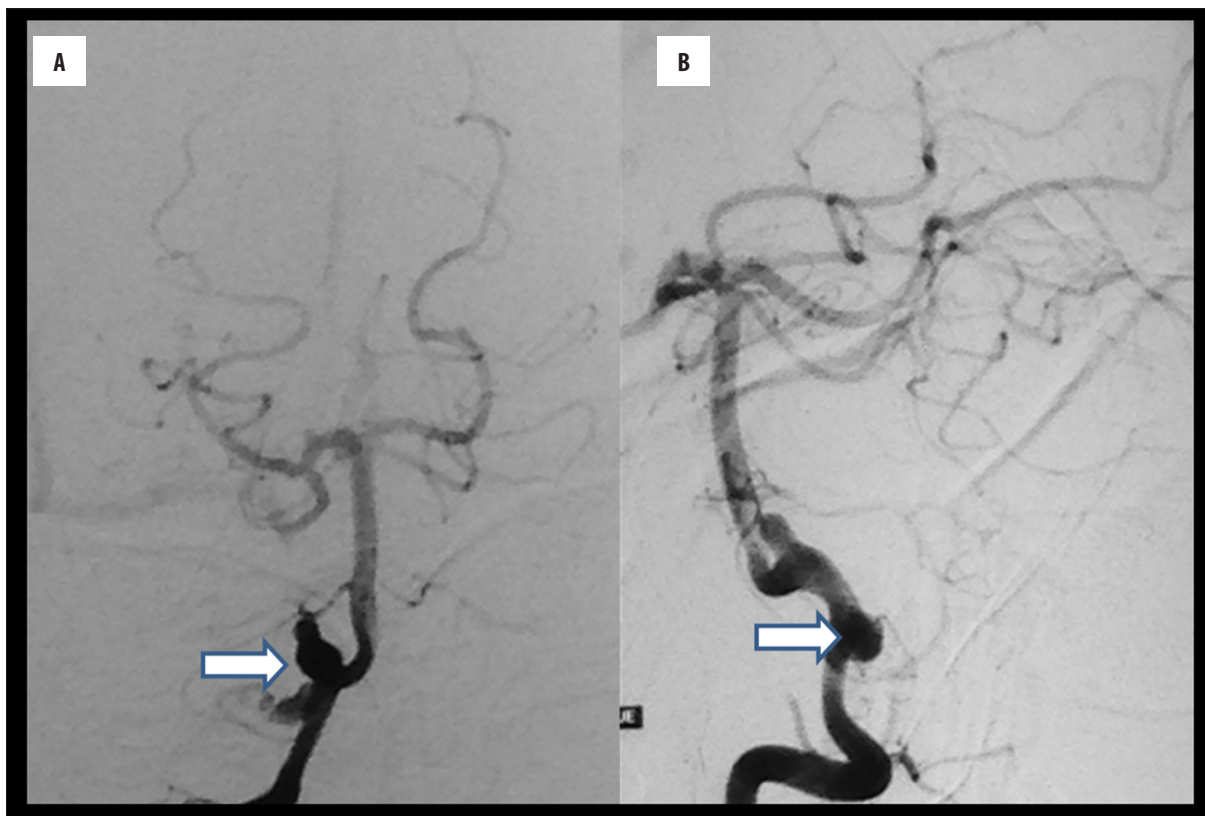


Figure 6. An 11-year-old boy with left atrial myxoma and oncotic aneurysms; Digital subtraction angiography (DSA), Right vertebral angiogram (A, B) – show an aneurysm in the intradural segment of the right vertebral artery before its junction with the basilar artery (arrow).

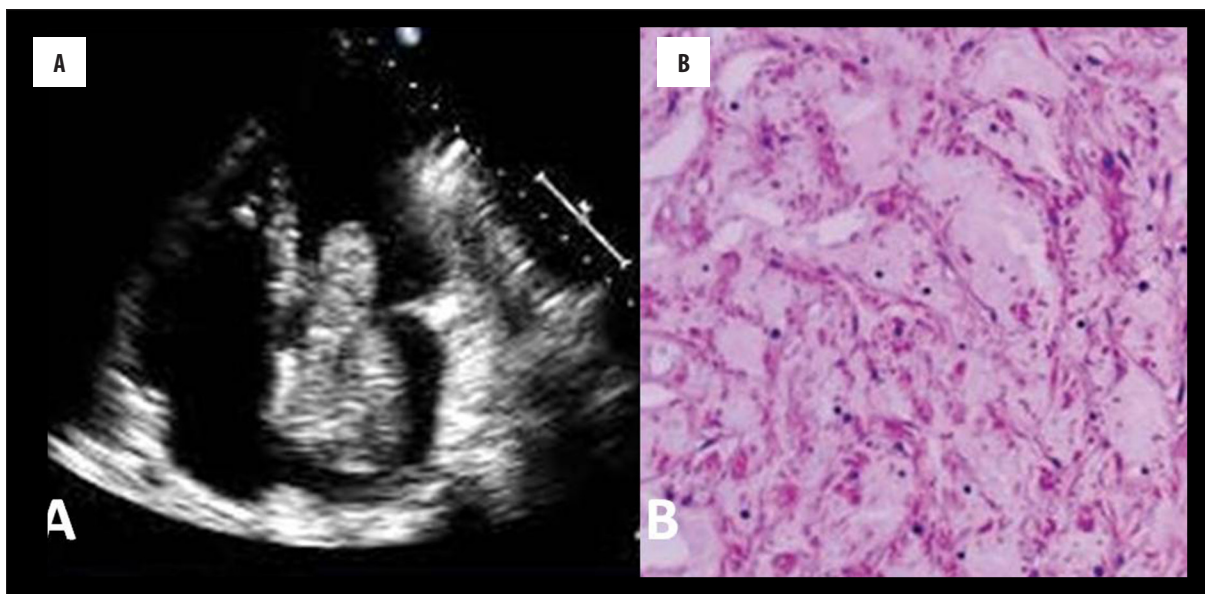


Figure 7. An 11-year-old boy with left atrial myxoma and oncotic aneurysms; One year after non-contrast computed tomography shows – (A) – infarcts (not visible one year before) in the head of the right caudate nucleus (small white arrow) and (B) – right high parietal region (small white arrow). In addition (B) – also show an old infarct in the left gangliocapsular area (large white arrow).

Since small peripheral aneurysms, which are more frequent, can only be detected on DSA by delayed washout of contrast relative to the arterial phase, a finding that cannot be appreciated on MRA or CTA, DSA may be the most sensitive imaging method in the diagnosis of small peripheral aneurysms [12].

No definite guidelines for therapy of the disease have been formulated. Resection of the cardiac myxoma is useful in eliminating early neurological symptoms but cannot completely abolish the risk of delayed cerebral aneurysm formation. Chemotherapy results were equivocal [13]. Radiation therapy in combination with chemotherapy has

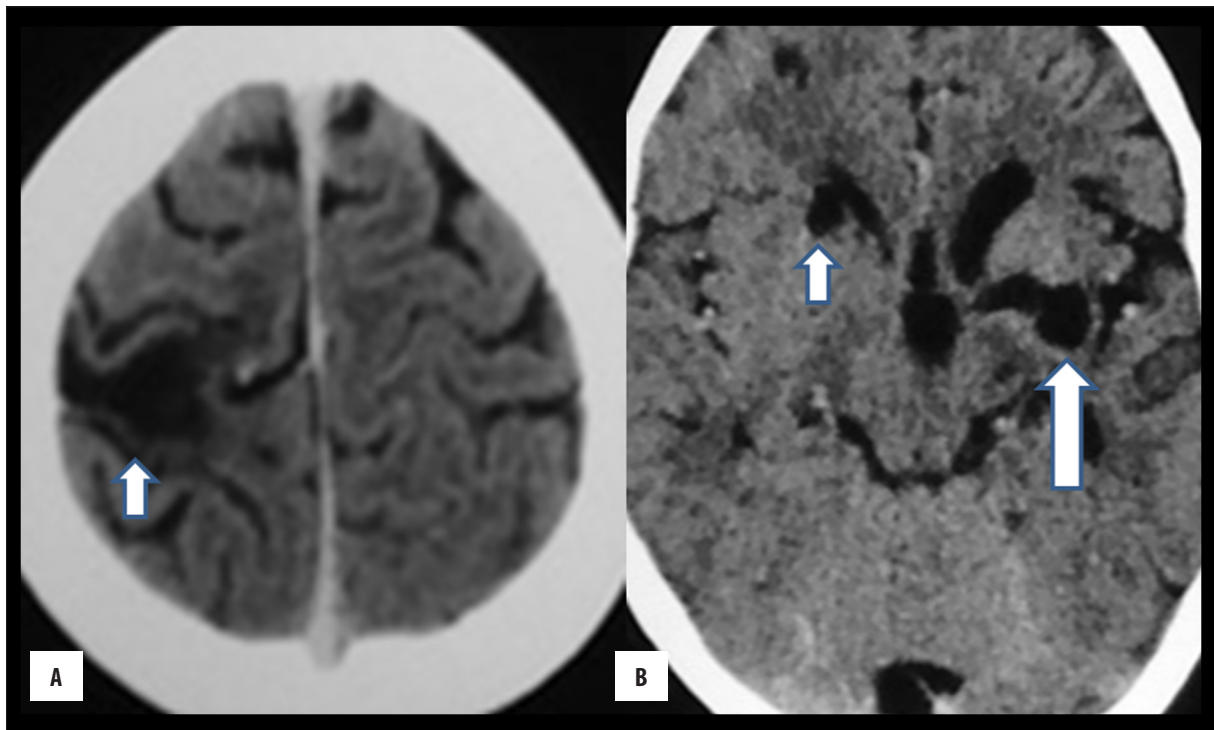


Figure 8. An 11-year-old boy with left atrial myxoma and oncotic aneurysms; One year after CT angiogram shows – (A) – tortuous, dilated and fusiform left MCA (large white arrow). (B) show multiple aneurysms in the distal branches of bilateral middle cerebral and vertebral arteries (small white arrows).

been reported as an effective way in degradation of myxoma metastasis, therefore it may be useful in preventing the myxomatous aneurysms [14].

Conclusions

Patients with multiple cerebral aneurysms, especially young patients, should be alerted to cardiac myxoma or the

resection history. Similarly, patients with cardiac myxoma should be observed for myxoma-related cerebral aneurysms as well as a cerebral embolism and the tumor must be resected as soon as possible to prevent further complications. Long-term regular follow-up of these patients is recommended.

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