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CASE REPORT

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Diagnostic imaging of cerebral cavernous angioma – case report and review of the literature

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Summary

Background: Cerebrovascular malformations are classified as cavernous malformations, arteriovenous malformations, developmental venous anomalies and capillary telangiectasias. Cavernous angioma is the second most common form of cerebrovascular malformations and constitutes up to 10–15% of the total incidence rate. Clinical features include epilepsy, focal deficits, headaches, or hemorrhages; cavernous angioma may also remain clinically silent.

Case Report: Presented is a rare case of multiple cerebral cavernous angioma enhanced in computed tomography and magnetic resonanse scans following intravenous infusion of contrast medium, also visible in angiography.

Conclusions: Magnetic resonance making use of the T2* GRE sequence with long TE is the best imaging tool of cerebral cavernous angioma. In rare cases, pathology may be revealed by cerebral angiography and angio-CT.

MeSH Keywords: Cerebral Angiography • Four-Dimensional Computed Tomography • Hemangioma, Cavernous, Central Nervous System

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Background

Angiomas account for 5–10% of the overall number of cerebrovascular malformations; they are traditionally classified as cavernous malformations, arteriovenous malformations, developmental venous anomalies and capillary telangiectasias [1,2]. Cavernous angioma (*angioma cavernosum*) is a benign vascular hamartoma consisting of a single chamber of multiple chambers of dumbbell shape. The interior of the angioma is sometimes compartmentalized by connective tissue partitions and filled with blood, thrombi or thrombus calcifications. The wall is devoid of muscle membrane and adventitia. The surrounding neural tissue is deformed by abnormal glia and hemosiderin; it may involve pathological vessels [1–4]. The incidence of cavernous angiomas is estimated at the level of 5–15% of all cerebral angiomas.[4]. Its prevalence within the central nervous system in human population, including autopsy examinations, is estimated at 0.02–0.5%. Cavernous angioma is a benign vascular lesion that may be found within the spine, optic chiasm, optic nerve, pineal gland, other arts of the brain as well as n other organs, such as liver and skin [5]. Most cavernous angiomas are single lesions, well-differentiated from the nervous tissue. Supratentorial locations are most common (80%), particularly within the frontal and temporal lobes. Less common are multiple lesions spread within the nervous system. This form was observed and described in Hispanic population, where familial incidence of cavernous angiomas was as high as 50%. It is believed that angiomas are inherited by the autosomal dominance pattern [6–8]. Most commonly, cavernous angiomas are manifested by epilepsy, focal deficits, or intracerebral hemorrhages [3,5,7]. Not all forms may lead to clinical manifestations [9,10]. Among focal lesions of vascular origins, cavernous angioma is increasingly often diagnosed as a cause of epilepsy [11]. The onset of pathological symptoms occurs between the third and the fourth decade of age, although reports were published that demonstrate that the disease may have a

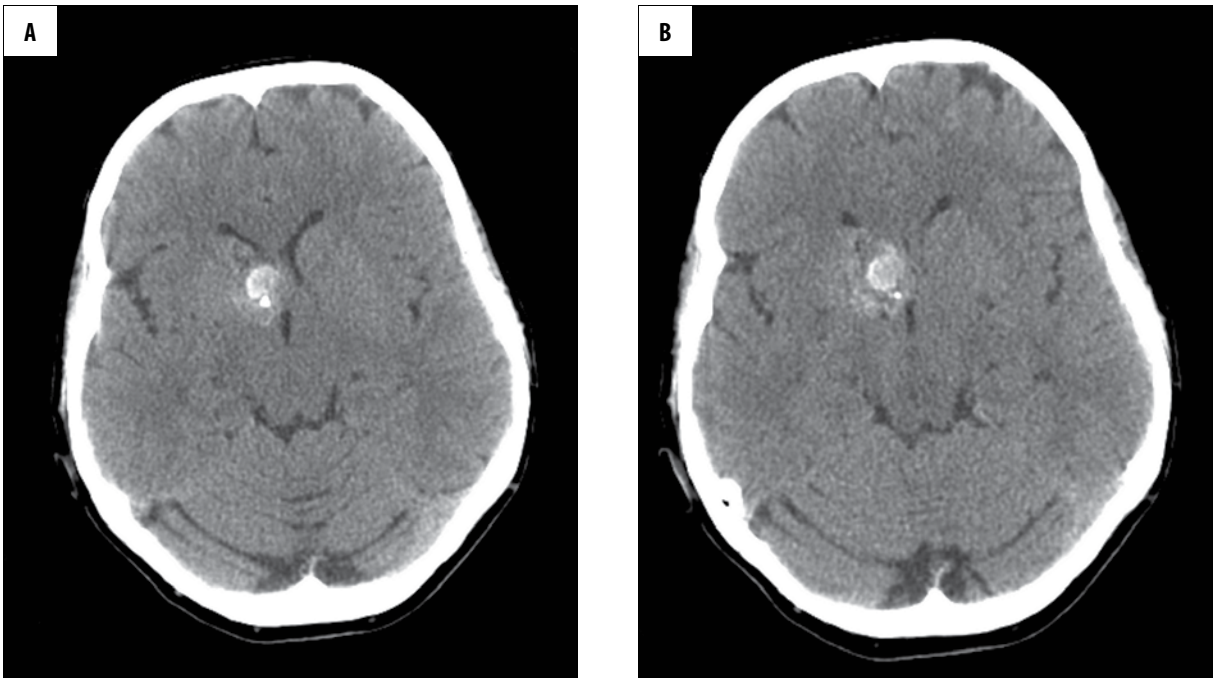

Figure 1. (**A, B**) Head CT, native scan – irregular hyperdense focus with calcifications and mass effect located in the right brain hemisphere, adjacent to the third ventricle and lower part of lateral ventricle frontal horn.

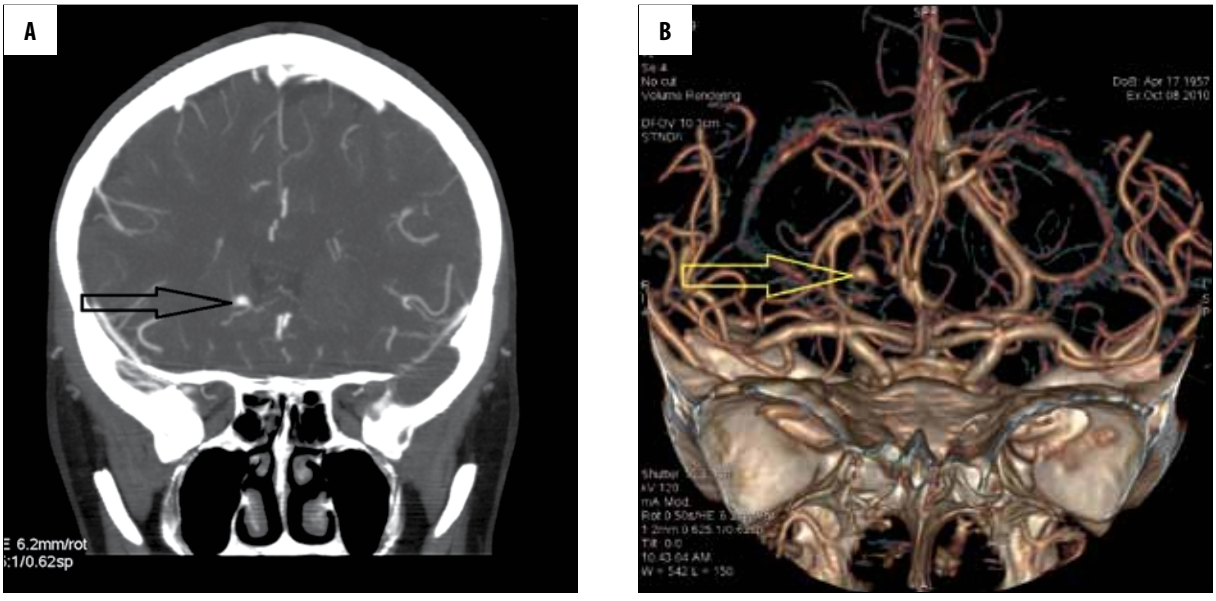

Figure 2. CT angiography. (**A**) MIP image – coronal plane, (**B**) 3D reconstruction – retention of contrast medium in cavernous hemangioma (arrow).

much earlier onset [2,4,11]. Magnetic resonance is the best method for imaging cavernous angiomas. T2-weighed GRE (gradient-echo) images with long echo times (TE) feature a characteristic "popcorn" image with hypointense hemosiderin hemlines. In computed tomography (CT) scans, cavernous angiomas are either not or poorly enhanced following intravenous contrast administration. Similarly, the lesion signal in magnetic resonance imaging (MRI) is usually not subject to pronounced enhancement following administration of contrast agents. Angiomas are usually impossible to visualize in angiographic examinations (DSA) due to the free flow of blood within the caverns. In exceptional cases, "discoloration" due to contrast administration may be observed at the site of the angioma [12]. Surgical excision id the treatment of choice in case of cavernous angiomas. This holds true in both pediatric and adult populations [13–15].

Case Report

A female patient, aged 53, was admitted to the Department of Nephrology due to moderately intense headaches within the right temporal and occipital region persisting for several days and accompanied by numbness of the right upper limb. Neurological examination revealed psychomotor retardation, hypoethesia in the right side of the face, mild muscle weakness within the right upper limb, weaker

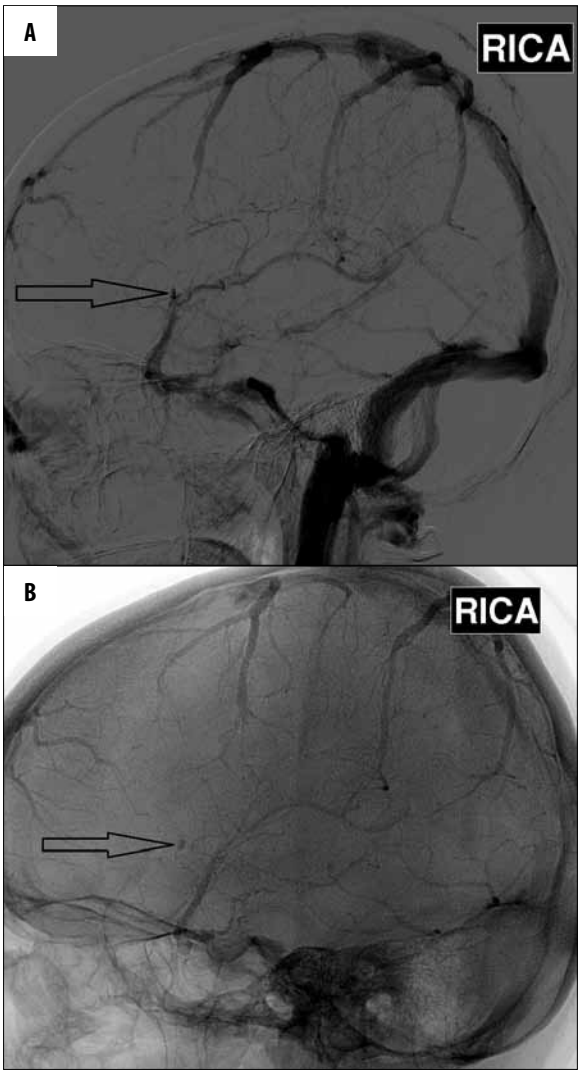

Figure 3. Angiography of the right common carotid artery, lateral projection. (**A**) subtraction image, (**B**) no subtraction image. Retention of contrast medium in cavernous hemangioma (arrow).

plantar reflex on the right side. A CT scan revealed an irregular hyperdense focus with calcifications adjacent to the third ventricle and lower part of lateral ventricle frontal horn (Figure 1). Contrast enhancement of the lesion signal was observed following intravenous administration of a contrast agent. Angio-CT, angiography and cerebral MRI scan were performed as part of more detailed diagnostics. Venous phase CT angiography revealed a presence of an oval, well-differentiated site of contrast agent retention within the cavernous angioma in the frontal horn of the right lateral ventricle (Figure 2). Venous phase DSA scan revealed an oval, well-differentiated site of contrast agent retention within the cavernous angioma in the region of the frontal horn of the right lateral ventricle, quite well visible without subtraction and due to the slow flow of blood within the lesion (Figure 3). An MRI scan revealed a non-homogeneous focal lesion sized 28×21×15 mm, of mixed signal intensity in both T1- and T2-weighted images, corresponding to partially coagulated blood at different stages of aging (Figure 4). Following administration

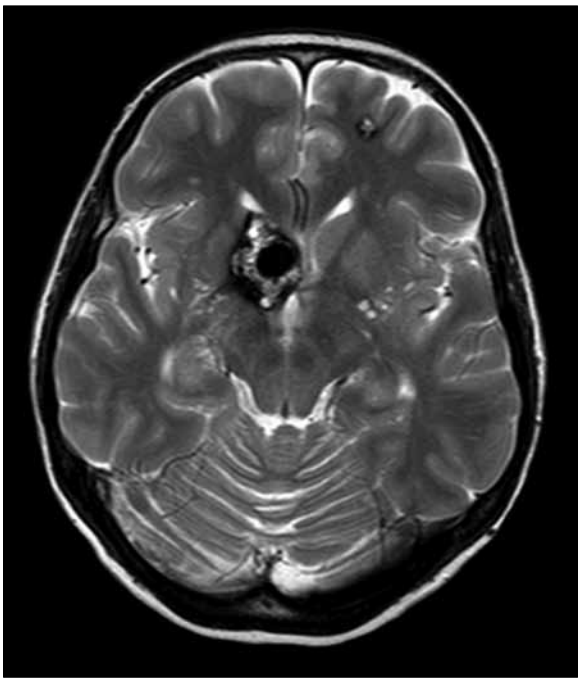

Figure 4. Head MRI scan, T2-weighted image, transverse plane. Non-homogenous foci of mixed signal intensity with blood, at different stages of aging, in the right brain hemisphere, adjacent to the caudate nucleus head and in the left frontal lobe.

of a contrast agent, the signal was locally enhanced in T1-weighted images (Figure 4). The focus gave rise to mass effect with remodeling of the septum pellucidum and the third ventricle accompanied with partial involvement of the caudate nucleus, striatum, and thalamus. A focus of similar type, 7 mm in diameter, is also visible within the white matter of the left frontal lobe (Figure 5).

Patient was discharged in good overall condition, without neurological deficits, with recommendation to report for a follow-up MRI scan. Follow-up cerebral MRI scan was performed after three months, revealing no significant differences compared to the previous scan.

Discussion

After anteriovenous angioma, cavernous angioma is the second most common type of nervous system angiomas [1–4]. The sizes of cavernous angiomas are differently assessed by different authors. The approximate diameters range from several millimeters to several centimeters [11]. Supratentorial locations are most common (80%), particularly within the frontal and temporal lobes. In the reported case, the angioma image remained unchanged in the follow-up scan after three months; however, cavernous angiomas present a tendency to grow in the long term, probably due to recurrent bleedings into the angioma itself as well as into its adjacent nervous tissue [16–18]. Clinical symptoms of cavernous angioma include epileptic seizures (22–50% of patients), neurological deficits (20–45% of patients), headaches (6–34% of patients), and intracerebral hemorrhages (ca. 20% of patients) [3,4,12]. Less common isolated symptoms of cavernous angiomas include optic disc swelling, internal

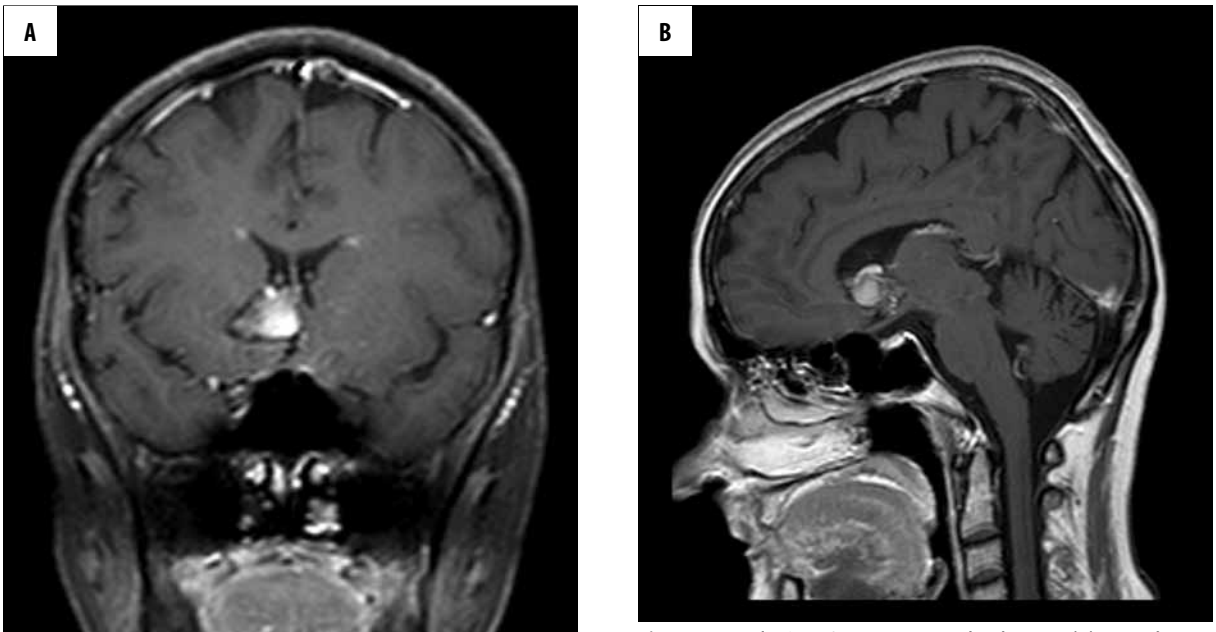

Figure 5. Head MRI + CM scan, T1-weighted image. (**A**) coronal plane, (**B**) sagittal plane. Central enhancement of hemangioma.

hydrocephalus or cranial nerve injuries [17,18]. In the reported case, neurological deficits manifested as hypoesthesia within the right side of the face, mild muscle weakness within the right upper limb and weaker plantar reflex on the right might be due to the small angioma within the left frontal lobe. The angioma located within the right hemisphere was most probably clinically asymptomatic. Magnetic resonance imaging is considered to be the method of choice in imaging diagnostics of cavernous angiomas [10,13]. Computed tomography images are not characteristic and require differentiation from teratoma, craniopharyngioma, ischemic stroke or developing inflammation. Cerebrovascular angiography is usually incapable of imaging cavernous angiomas due to the free flow of blood within the lesion; however, is allows to differentiate cavernous angiomas from other types of cerebral angiomas characterized by dynamic blood flow [6,13]. Only one case of the retention of contract medium within the angioma was revealed in the study material. In the remaining cases (90%), the angiographic image was unremarkable.

Conclusions

Magnetic resonance making use of the T2 GRE sequence with long TE values is the best imaging tool of cerebral cavernous angioma. Only in rare cases, pathology may be revealed by cerebral angiography and angio-CT.

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