INTRACRANIAL VENOUS ANGIOMAS – CASE REPORT

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SUMMARY – Two cases of intracranial venous angiomas, one in a 42-year-old woman and another one in a 27-year-old-man, are presented. Intracranial venous angiomas, also known as developmental venous anomalies, represent one of four cerebral vascular malformations. Usually they are incidentally discovered on cerebral imaging. These lesions have a characteristic appearance in the venous phase of angiography, a starburst pattern or umbrella sign, described as *caput medusae*. Most of the lesions are clinically silent, however, they can be symptomatic with clinical presentation including seizures, headache, dizziness and focal neurologic deficits. They are sometimes associated with cerebral hemorrhage. In general, no treatment is required. Surgery is only indicated if seizures attributed to the lesion or hemorrhage occur.

Key words: Central Neuro System Venous Angioma; Case Report

Introduction

Intracranial venous angiomas, also known as developmental venous anomalies (DVAs)1-3 are one of four cerebral vascular malformations^{1,4} usually expected in regions supplied by the middle cerebral artery or in the region of the vein of Galen⁵. The cerebral venous angiomas are actually embryologic variants of venous drainage and form an alternative venous drainage of the surrounding nervous tissue6, which is composed of anomalous veins separated by normal neural parenchyma⁷ in which embryologic medullary venules persist and cluster in a central, large, draining venous trunk. This collector drains intracerebrally into the deep or superficial venous system⁸⁻¹⁰. One third of these lesions are located in the cerebellum and in the brainstem; the remaining two thirds are supratentorially located. These lesions usually are incidentally discovered on cerebral imaging^{1,11} and have a characteristic appearance in the venous phase of

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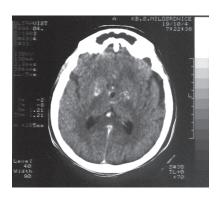
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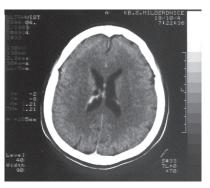
angiography, a starburst pattern or umbrella sign¹² that is described as *caput medusae*. Most venous angiomas are clinically silent, however, they can be symptomatic with clinical presentation including seizures, headache, dizziness and focal neurologic deficits¹³. They are sometimes associated with cerebral hemorrhage⁸. In general, they require no treatment. Surgery is only indicated if seizures attributed to the lesion or hemorrhage occur⁴.

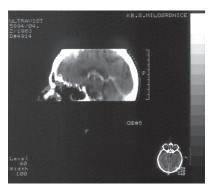
Case Reports

Case 1

A 42-year-old, previously healthy woman was admitted to the Hospital Department of Neurology for grand mal seizures. At the time of admission, she had no neurologic deficit. Computer tomography (CT) scan of the head performed immediately upon admission showed hyperdensities in the region of lenticular nucleus and internal capsule bilaterally, near the third ventricle and in the area of the splenium corporis callosi. Upon contrast application, there was enhancement in these areas as a radiologic sign of arteriovenous malformation (Fig. 1). Therefore, magnetic resonance imaging (MRI) and MR angiography were performed, which







A = axial scans;

B = axial scans

 $C = sagittal \ reconstruction$

Fig. 1. Contrast enhanced CT scan of the brain shows characteristic finding for arteriovenous malformation in the area of the lenticular nucleus and internal capsule on both sides nearby the third ventricle and the splenium corporis callosi.



A = T2 weighted axial scan;



B = MR angiograms



C = MR angiograms

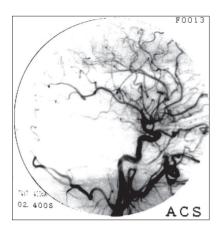


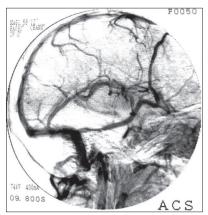
D = MR angiograms

Fig. 2. MR with MR angiography showed the same findings as contrast enhanced CT scan. Clearly visible arteriovenous malformation.

showed the same findings (Fig. 2). The patient was treated conservatively and transferred to Department of Neurosurgery. Cerebral angiography showed venous angioma involving both sides of the mid-brain region, with drainage into

the superior sagittal sinus, straight sinus and sigmoid sinus (Figs. 3,4,5). There were no signs of intracerebral hemorrhage, so we decided to treat the patient conservatively for the low risk of hemorrhage.





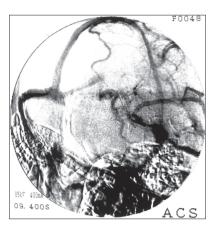
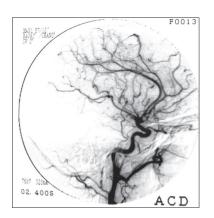
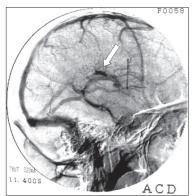


Fig. 3. Digital subtraction angiography: left common carotid artery (CCA) catheterized. Arterial phase (A) without pathomorphologic changes. Late venous phase (B) and (C): extensive venous malformation in the mid-brain region.





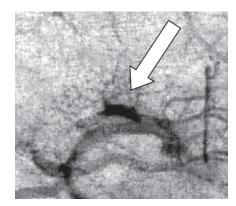


Fig. 4. Digital subtraction angiography: right common carotid artery (CCA) catetherized. Arterial phase (A) without pathologic changes. Late venous phase (B) and magnified detail from previous figure (C): extensive venous angioma with characteristic drainage.

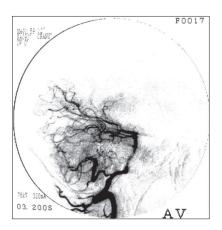
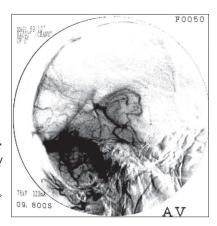


Fig. 5. Digital subtraction angiography: left vertebral artery (VA) catheterized. Arterial phase (A) without pathomorphologic changes. Late venous phase (B) shows venous drainage of the mid-brain as venous malformation.



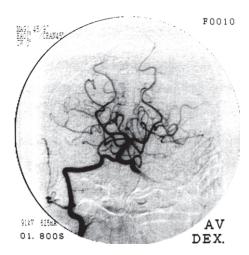


Fig. 6. Digital subtraction angiography: right vertebral artery (VA) catheterized. Middle arterial phase in AP projection with tube inclination. Normal finding.

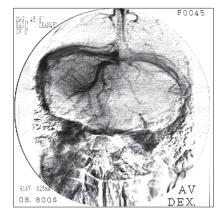
Case 2

A 27-year-old-man was admitted to the Hospital Department of Neurosurgery for headache and cerebellar symptomatology. He had been treated for epilepsy from age 10. Initially, cerebellar symptompatology was present, with postural instability and left lower extremity weakness. CT scan of the brain showed no pathologic finding. MRI of the brain showed Arnold Chiari I malformation. In the right cerebellar hemisphere there was a thin zone of intensity on both T1 and T2 weighted images that looked like a venous angioma. Cerebral angiography showed typical venous angioma in the right cerebellar hemisphere, formed from a number of tortuous smaller veins draining into one central vein (Figs. 6,7). The patient was operated on for Chiari I

malformation. Decompressive suboccipital craniectomy was performed. Venous angioma was treated conservatively.

Discussion

Two cases of intracranial venous angioma are reported. Venous angiomas, also called developmental venous anomalies^{2,3} are composed of anomalous veins separated by normal neural parenchyma and represent congenital anomalies of the intracranial venous drainage. The malformations may be composed of a single, greatly dilated, tortuous vein or a number of smaller veins coalescing at a single point without direct arterial input. Histologically, venous angiomas are characterized by a composition of sometimes thickened and hyalinized veins with interspersed normal neural parenchyma. Although once considered rare, they are now thought to be one of the most frequent cerebral vascular abnormalities¹³⁻¹⁵. Very often these anomalies are incidentally discovered on cerebral imaging¹¹. On angiography they present typically as deep, medullary veins during the early or middle venous phase accompanied by a single large draining vein. On CT they are rarely detectable without contrast enhancement. After application of contrast medium the draining veins appear as a linear focus of enhancement¹⁶. On MRI venous angiomas have a characteristic flow void on both T1 and T2 weighted images¹⁵. Administration of MR contrast material considerably improves demonstration of the draining vein and often allows visualization of the medullaryveins. However, they can also be symptomatic with clinical presentation including seizures, headache, dizziness and focal neurologic deficits^{13,17}. They may be associated with a cavernous malformation and are somettimes associated with



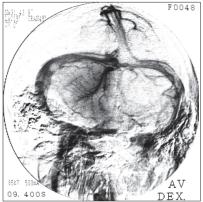




Fig. 7. Digital subtraction angiography: late venous-sinus phase with normal venous sinuses and clearly visible typical venous malformation (A). Very late venous-sinus phase with venous malformation and delayed contrast flow and extensive drainage vein, which ends in confluence of sinuses (B). Enlarged detail from previous figure (C).

cerebral hemorrhage¹⁴. The clinical importance of these lesions is as potential causes of cerebral hemorrhage and cause of obstructive hydrocephalus¹⁸. The authors present two cases of venous angiomas, one in a 42-year-old woman with grand mal epilepsy. Since surgery is indicated if seizures are attributed to the lesion, in this case there was a relative indication for operative treatment. Yet, we did not indicate it due to the location of the lesion and probably massive postoperative neurologic deficit. The other patient was operated on for Arnold-Chiari I malformation, the probable cause of his symptoms. However, venous angioma was not treated operatively. There are opinions that there is no indication for surgery in patients with venous angioma unless they present with an extensive intracerebral hematoma, since venous angiomas drain normal cerebral tissue within a functionally normal arterial territory and resection may lead to venous infarction17.

We think that there are many factors that should be considered, e.g., clinical symptomatology, patient age, localization and size of angioma. However, because of the low risk of new neurologic events, we propose conservative management in most cases. The risk associated with surgical intervention greatly exceeds the low risk of morbidity related to venous malformation hemorrhage. The other treatment option, radiosurgery, also remains contraindicated^{4,19}.

Although cerebral venous malformations have been reported to cause epilepsy, progressive neurologic deficits, and hemorrhage, their clinical significance still remains controversial¹³.

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Sažetak

INTRAKRANIJSKI VENSKI ANGIOMI – PRIKAZ SLUČAJA

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Prikazana su dva slučaja intrakranijskih venskih angioma, jedan u 42-godišnje žene i drugi u 27-godišnjeg muškarca. Intrakranijski venski angiomi, također poznati kao razvojne venske anomalije, čine jednu od četiri cerebralne malformacije. Obično se slučajno otkrivaju neuroradiološkom obradom. Ove lezije se znakovito prikazuju u venskoj fazi angiograma kao zvjezdasti uzorak ili poput kišobrana, što se opisuje kao *caput međusae*. Većina venskih angioma su klinički asimptomatski, iako mogu biti i simptomatski te se tada klinički očituju epileptičnim napadajem, glavoboljom, vrtoglavicom i žarišnim neurološkim deficitom. Ponekad su povezani s intracerebralnim krvarenjem. U većini slučajeva ne zahtijevaju liječenje. Operacijsko liječenje je indicirano samo u slučajevima dokazane povezanosti angioma i epileptičnih napadaja odnosno u slučaju krvarenja.

Ključne riječi: Venski angiom središnjeg živčanog sustava, Prikaz slučaja