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Case Report

A case of cerebral amyloid angiopathy associated with cerebral venous thrombosis

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ABSTRACT

Cerebral amyloid angiopathy is a clinical picture which is commonly seen in elderly and progressing with the deposition of amyloid in the cerebral arteries without systemic amyloidosis. We report the first case in the literature, a 71 year-old patient having an association of cerebral vein thrombosis and cerebral amyloid angiopathy presenting with recurrent cerebral hemorrhages. The cause-and-result relationship of this association of cerebral vein thrombosis and cerebral amyloid angiopathy should be investigated.

Keywords: Cerebral amyloid angiopathy, Cerebral vein thrombosis, Recurrent cerebral hemorrhages

INTRODUCTION

Cerebral Amyloid Angiopathy (CAA) selectively involves the leptomeningeal arteries and small arteries of the cerebral cortex. This is a rather lobar localized clinical condition which leads to recurrent parenchymal hemorrhage, mainly in the frontotemporal regions. It is characterized by the accumulation of amyloid, an abnormal protein in the vessel walls, and degeneration in the smooth-muscle cells. As a result of hypoperfusion caused by the vessels narrowed with accumulation of amyloid, hemorrhage is accompanied also by leukoencephalopathy. It may show genetic transition, as well as being sporadic. Staining of amyloid in the vessels with Congo red allows the establishment of the final diagnosis.[3] Since patients may develop recurrent cerebral hemorrhage, antiagregant and anticoagulant therapies should be given carefully.[3] In elderly patients, CAA, which is the most common cause of lobar intracerebral hemorrhage, can be diagnosed without need for pathological examination via the Boston criteria (whose validity has been proven.[4]

Boston criteria for CAA

Definite CAA: Full postmortem examination demonstrating

- Lobar, cortical, or corticosubcortical hemorrhage
- Severe CAA with vasculopathy
- Absence of other diagnostic lesion

Probable CAA with supporting pathology: Clinical data and pathological tissue (evacuated hematoma or cortical biopsy) demonstrating

- Lobar, cortical, or corticosubcortical hemorrhage
- Some degree of CAA in specimen
- Absence of other diagnostic lesion

Probable CAA: Clinical data and MRI or CT demonstrating

- Multiple hemorrhages restricted to lobar, cortical, or corticosubcortical regions (cerebellar hemorrhage allowed)
- Age >55 years
- Absence of other cause of hemorrhage

Possible CAA: Clinical data and MRI or CT demonstrating

- Single lobar, cortical, or corticosubcortical hemorrhage
- Age >55 years
- Absence of other cause of hemorrhage

Cerebral Venous Thrombosis (CVT) is a clinical condition which is an infrequent cause of stroke, and which has various causes, including coagulopathy, intracranial infections, malignity, dehydration, and pregnancy, each of which play a role in the etiology and are also among the infrequent cause of strokes.[2] Many signs and symptoms may be seen during the course of CVT, ranging from headache to focal neurological deficits, seizures, impaired consciousness, and coma.

This report focuses on a case present in the current literature of a patient having an association of cerebral vein thrombosis and cerebral amyloid angiopathy presenting with recurrent cerebral hemorrhages.

CASE REPORT

A 71-year-old male patient presented at the emergency department with acute-onset weakness in the right arm and leg, epileptic seizures, and loss of consciousness. The brain Magnetic Resonance Imaging (MRI) scan showed a wide intraparenchymal hemorrhage area at the left frontal level opening to the ventricular system and multiple blooming subcortical hypointensities (Figure 1). The patient had a history of first- and second-cerebral hemorrhagic attacks in 2012, with an interval of one month for one in the left frontal and another in the right basal ganglia. Upon a T2W gradient scan of the brain MRI, multiple punctate hypointensities signal records suggesting amyloid angiopathy were observed in the cerebral subcortical white matter areas and late subacute-process hematoma areas were seen in the right insular lobe-basal ganglia and the left frontal subcortical white matter areas (Figure 2). The T2W scan also detected hyperintense signal records compatible with thrombosis in the left transverse sinus and sigmoid sinus (Figure 3). Since the first attack of cerebral hemorrhages was followed-up in a different center, we could not access the follow-up and treatment information. However, upon a cerebral MRI, parenchymal hematoma and mild edema was reported in the left anterior frontal region. Furthermore, it was described in the patient report that there were chronic ischemic gliotic lesion areas in wide patch form in the periventricular white matter of both cerebral hemispheres that were developed due to hyperintense chronic small vessel ischemic disease. In the MRI report, amyloid angiopathy was stated as a possible cause to the etiology of the hemorrhage. At the contrast-enhanced examination, no meningeal and parenchymal enhancement was monitored in the brain stem, cerebral, and cerebellar hemispheres. Upon an MR-angiogram, no finding compatible with vascular pathology was found.

In terms of patient history, there was no significant disease except hypertension, and no use of alcohol or amphetamines. It was learned, however, that the patient's cousin (the daughter of his aunt) died from cerebral hemorrhage. The patient was using warfarin and amlodipine but no other drugs. He was hospitalized with the diagnoses of CAA and cerebral vein thrombosis, and put on anti-edema therapy with dexamethasone and mannitol. The hemogramme and routine biochemical analyses yielded normal outcomes. The thrombosis panel was normal and vasculitis markers were negative. He was administered anticoagulant therapy for cerebral vein thrombosis for one year. Prothrombin Time (PT) values were found to be continuously monitored and in the therapy range. On his last admission, İnternational Normalized Ratio (INR) value was 2.6. Cerebral Arteriovenous Malformation (AVM), aneurysms, and tumors were ruled out with the completion of the imaging. MRI outcomes were compatible with "probable CAA." The patient developed pulmonary infection on the 10th day after his admission, did not respond to antibiotherapy, and was lost due to sepsis.

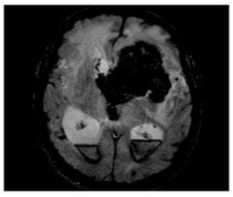

Figure 1: Brain magnetic resonance imaging (MRI) showing hemorrhage in the ventricles and left frontal lobe (the third hemorrhagic attack).

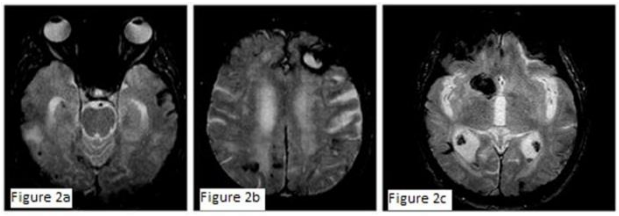

Figure 2: Upon T2W gradient examination of the brain MRI, punctate hypointensies signal records suggesting amyloid angiopathy were observed in the bilateral cerebral subcortical white matter areas (Figures 2a, b), and late subacute-process hematoma areas were seen in the insular lobe-basal ganglia and the left frontal subcortical white matter areas (Figures 2b, c) related to the second hemorrhagic attack.

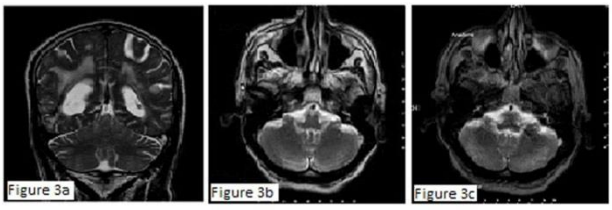

Figure 3: Brain magnetic resonance imaging (MRI) showing hyperintense signal records compatible with thrombosis in the left transverse sinus and sigmoid sinus related to the second (Figures 3a, b) and the third hemorrhagic (Figure 3c) attacks.

DISCUSSION

Cerebral Amyloid Angiopathy (CAA) is a clinical condition commonly seen among the elderly and which progresses via the deposition of amyloid in the cerebral arteries without systemic amyloidosis. CAA incidence is between 4% and 10% in the patients with spontaneous cerebral hemorrhage.[1] A definite incidence rate is difficult to provide since histopathological examinations cannot fully be carried out, but it is known that the incidence increases with age. Today, there is not a specific therapy, but is known that antiplatelet and anticoagulant therapies should be avoided.[3]

The 71-year-old patient reported on in this paper had been examined due to three attacks of hemorrhage and was diagnosed with CAA and cerebral venous thrombosis. This association has not been reported so far in the literature examined for this report.

There is no direct association between CAA and hypertension. However, hypertension is shown in 66.7% of CAA patients, and there was a history of hypertension in the patient examined here.

CAA affects the neocortex more than the basal ganglia, cerebellum, and brain stem, with the most commonly involved area demonstrated to be the frontal, temporal, and occipital lobes.[1] A hemorrhage was seen in the frontal and basal ganglia area in the 71-year-old patient, who also had a history of recurrent cerebral hemorrhage, with the first two attacks developing with an interval of one month, and a third attack occurring one year later.

According to the Boston Criteria, which are used as a standard in diagnosis of CAA, existence of two or more hemorrhages limited within the lobar, cortical, or corticosubcortical areas are classified as probable CAA unless another structural brain pathology is present in patients age 55 and over.[4] Due to possessing these features, the patient also was compatible with the probable CAA diagnosis.

The 71-year-old patient had concurrent sinus vein thrombosis with the second attack of cerebral hemorrhage and was taking anticoagulant medication for that pathology. In a recent study, it was reported that amyloid accumulation was seen in the cerebral vein walls in addition to the cerebral arteries, and the incidence for this reached 78% in the patients having CAA.[5] We believe that amyloid angiopathy that might be accumulated in the vein walls could contribute to the development of cerebral vein thrombosis, although it might also be considered that the anticoagulant therapy the patient had been receiving after the development of the second attack of hemorrhage or hypertension could be the reason for the development of the third attack. The different areas affected in the hemorrhagic attacks and the patient's previous history of having two attacks of cerebral hemorrhage before the use of anticoagulant led us to the diagnosis of CAA.

In conclusion, the patient is the first to be reported for the association of cerebral vein thrombosis and amyloid cerebral angiopathy, which is demonstrated by the recurrent cerebral hemorrhages and the cause-and-result relationship of this association, should be investigated.

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REFERENCES

1. Cosgrove GR, Leblanc R, Meagher-Villemure K, Ethier R. Cerebral amyloid angiopathy. Neurol. 1985;35(5):625-31.
2. Einhäupl K, Stam J, Bousser MG, De Bruijn SF, Ferro JM, Martinelli I, et al. EFNS guideline on the treatment of cerebral venous and sinus thrombosis in adult patients. Eur J Neurol. 2010;17(10):1229-35.
3. Kase CS. Intracerebral hemorrhage. In: Bradley WG, Daroff RB, Fenichel GM, Jankovic J, eds. Neurology in Clinical Practice: The Neurological Disorders. 4th ed. Heinemann-Philadelphia: Butterworth; 2004: 1251-1267.
4. Knudsen KA, Rojand J, Karluk D, Greenberg SM. Clinical diagnosis of cerebral amyloid angiopathy: validation of the Boston criteria. Neurol. 2001;56(4):537-9.
5. Mendel T, Wierzba-Bobrowicz T, Stępień T, Szpak GM. β-amyloid deposits in veins in patients with cerebral amyloid angiopathy and intracerebral haemorrhage. Folia Neuropathol. 2013;51(2):120-6.

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