Cerebral Amyloid Angiopathy: Report of Two Cases

Serebral Amiloid Anjiyopati: İki Olgu Sunumu

ABSTRACT Cerebral amyloid angiopathy (CAA) is characterized by amyloid beta-peptide deposition in the media, adventitia of leptomeningeal, cortical, subcortical arteries and arterioles. No significant associations have been shown between CAA and other systemic/visceral amyloidoses or vascular risk factors, including hypertension. It is generally seen in elderly or demented patients. The amyloid in the vessel lead to fibrinoid necrosis, microaneurysms, cerebral infarction, intracerebral hematomas. Here we report two cases who presented with intracerebral hemorrhage. The histopathologic examination of the tissue obtained during hematoma evacuation demonstrated eosinophilic material accumulation in the walls of the leptomeningeal and cortical arterioles. The deposits which stained pink-red by Congo Red showed yellow-green birefringence by the polarizing microscope. In addition, vessels also showed fibrinoid necrosis. Both of the cases were diagnosed as severe grade of CAA. During the hematoma evacuation, a small parenchymal tissue is needed for determining the etiology of the hemorrhage including CAA.

Key Words: Amyloid angiopathy; cerebral hemorrhage; surgery


Anahat Kelmeler: Amiloid anjiyopati; serebral hemorajı; cerrahi


Cerebral amyloid angiopathy (CAA) is an important and common cause of spontaneous lobar hemorrhage in elderly and/or demented patients which goes undiagnosed in the absence of histological examination of the blood vessels in the clot. The patients are presented with stroke due to the intracerebral bleeding or cognitive impairment consequent to chronic ischemia. Amyloid deposition in neocortical regions is a hallmark feature of Alzheimer disease (AD). Caselli et al. confirmed that...
nondemented APOE ε4 carriers have much more cerebral amyloid pathology than noncarriers supporting the possibility that even in the absence of clinical impairment, neocortical and vascular amyloid represent an early step in AD pathogenesis.\(^2\) Clinical features and MRI findings are helpful for the diagnosis of probable CAA with leukencephalopathy and superficial siderosis.\(^3\) According to the Boston criteria probable diagnosis is made in elderly patients, with at least two acute or chronic lobar hemorrhagic lesions without any other cause of intracerebral hemorrhage like prior trauma, ischemic stroke, central nervous system tumor, vascular malformation or bleeding diathesis.\(^4\)

**CASE REPORTS**

**CASE 1**

A 74-year-old woman admitted to the emergency with sudden loss of consciousness. Her medical history did not reveal any risk factors for intracerebral hemorrhage (ICH) such as systemic hypertension, trauma and there was no history of cognitive impairment. Five years ago she was also hospitalized because of intracranial hemorrhage and got only conservative treatment. Blood pressure was 160/100 and was noted to have right sided hemiplegia. Cranial computed tomography revealed 6x4.5 cm hematoma in the temporoparietal region with midline shift and surrounding edema (Figure 1). Routine blood including coagulation were normal. She underwent emergent parietooccipital craniotomy and subdural, intraparenchymal hematoma was evacuated. The histopathologic examination of hematoma demonstrated the vascular thickening resulted from eosinophilic homogeneous material accumulation in the walls of the leptomeningeal arterioles (Figure 2). The superficial cortical parenchymal arterioles similarly showed round and thickened vascular contours. The deposits which stained pink-red by congo-red (Figure 3, 4) showed yellow-green birefringence when observed by polarizing microscope (Figure 5). There was a severe degree of vascular amyloid changes, characterized by extensive amyloid deposits, vessel wall fragmentation, fibrinoid necrosis (Figure 6).

**CASE 2**

A 73-year-old woman presented with impaired orientation. Her spontaneous speech was sparse and not fluent. Her medical history did not reveal any risk factors for ICH. There was no motor deficit. Cranial CT scan showed 4x4 cm hematoma in the left frontal lobe, periventricular multiple chronic lacunar infarction and ischemic signal alterations, atrophic cerebral and cerebellar hemispheres. She underwent emergent left frontal craniotomy and hematoma evacuation. On the first postoperative day, clinical picture was improved. Evacuated
hematomas showed blood clots and few hemorrhagic cortical fragments. There was eosinophilic homogeneous material accumulation in the walls of the leptomeningeal and cortical arterioles which was congophilic and birefringent.

**DISCUSSION**

Amyloid deposition restricted to the central nervous system, which was first described by Gustav Oppenheim in 1909, is a frequent form of localized amyloidosis. Its prevalence is strongly associated with increasing age and has been reported to be as high as 57% percent in case series of asymptomatic patients over 60 years of age with or without dementia. CAA occurs mostly sporadic condition and all of the sporadic cases are due to deposition of amyloid β-peptide (Aβ), derived from Amyloid Precursor Protein. The ApoE ε4 ε2 alleles are genetic risk factors associated with sporadic Aβ CAA. Hereditary forms are generally familial, more severe and earlier in onset. Vonsattel et al. graded CAA with respect to the severity of pathological changes in a blood vessel: mild, amyloid is restricted to the tunica media without significant destruction of smooth muscle cells; moderate, tunica media is replaced by amyloid and is thicker than normal; severe, extensive amyloid deposition
with focal wall fragmentation or microaneurysm formation, fibrinoid necrosis and leakage of blood through the blood vessel wall. Amyloid deposition can weaken cerebral vessel walls, causing rupture leading to asymptomatic microbleeds and lobar ICH or obliterate the vessel lumen leading to ischemia and infarction. Treatment or preventive strategy for CAA doesn’t exist at this time. Corticosteroid and other immunosuppressant treatments can ameliorate the symptoms associated with inflammation. Recent reported results suggest that controlling blood pressure is likely to reduce risk of CAA-ICH recurrence. Further researches are required to identify therapeutic targets and take caution. Spontaneous intracerebral hemorrhage, defined as hemorrhage in the brain parenchyma in the absence of trauma or surgery accounts for 8 to 15% of all strokes. Common causes include hypertension, amyloid angiopathy, coagulopathy, vascular anomalies. Examination of cortical tissue obtained surgically is an important tool for the diagnosis of CAA during life.

REFERENCES