中文題目：一種不常見的自發性腦出血發生在一位患有末期腎臟病及高血壓的女性

英文題目：An uncommon spontaneous intracerebral hemorrhage in a female with end stage renal disease and hypertension

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Background:

Cerebral Amyloid Angiopathy (CAA) is an uncommon cause of spontaneous intracerebral hemorrhage (ICH). It is defined as deposition of amyloid in the wall of brain vessels. Clinically, it differs from hypertension related intracerebral hemorrhage by the location of bleeding. The hemorrhage is often located in the brain cortex, unlike hypertension related intracerebral hemorrhage which is in the basal ganglia or thalamus. Besides, recurrent ICH is more seen in cerebral amyloid angiopathy rather than hypertension. However, cerebral amyloid angiopathy is a pathologic diagnosis, it is often misdiagnosed due to lack of tissue proof. Here, we introduce a case of intracerebral hemorrhage caused by cerebral amyloid angiopathy.

Case Presentation:

A 66-year-old female with a history of end stage renal disease (ESRD) under hemodialysis, coronary artery disease status post coronary angiography and stenting, and hypertension was presented to the emergency department (ED) because of acute onset left limbs weakness.

She took aspirin and doxazosin for her hypertension control regularly. She suffered from left side weakness when she woke up and was accompanied with severe headache, nausea, and vomiting in the morning. Since she arrived in the ED, initial GCS score was 15. Her vital sign at triage showed blood pressure 245/93 mmHg, regular pulse rate 97 per minutes, smooth respiratory pattern, and body temperature 36.2 degrees Celsius. Blood pressure was controlled after medicine use, but conscious unclearness was noticed. Her GCS score dropped from 15 to 6 (E1M4V1) after 1 and a half hours arrived emergency department and the GCS score dropped to 3 after 2 f hours arrived emergency department. Brain computed tomography (Figure 1) showed an acute hematoma with active bleeding in the right temporal-parietal lobe which caused mass effect in the left side, uncal herniation, and trans-tentorial herniation.

Due to intracerebral hemorrhage, the patient arranged craniectomy, removed right side hematoma and partial temporal lobectomy. Further pathology report showed a picture of amyloid angiography with hemorrhage characterized by acellular, eosinophilic material deposited in the vascular wall

(Figure 2) which are highlighted by beta-amyloid stain (Figure 3) and Congo red stain (Figure 4). After the operation, she was admitted to the intensive care unit for further care and transferred to the respiratory care center for weaning ventilator 12 days after operation.

Discussion:

In this case we described a case of spontaneous ICH caused by cerebral amyloid angiopathy. CAA is defined by histopathology—deposition of β-amyloid in the cerebrovascular—and through the 1980s the disorder was only diagnosed in patients with available brain tissue from hematoma evacuation, biopsy, or most commonly postmortem examination. Now, there are two diagnostic criteria for cerebral amyloid angiopathy, one is modified Boston criteria, and another is Edinburgh criteria. Modified Boston criteria is based on pathology, brain image and clinical history. Edinburgh criteria is based on brain CT and APOE ε4 allele. Edinburgh criteria does not need the pathology but need the lab data of APOE ε4 allele, thus Edinburgh criteria is more suitable for patient who had family history. Modified Boston criteria divided cerebral amyloid angiography into four tiers and this case is defined as probable CAA with supporting pathologic evidence. Even though definite CAA need full post-mortem examination, we can still diagnose a patient as probable or possible CAA by brain image and clinical history without pathology.

The common symptoms of CAA are intracerebral hemorrhage and cognitive impairment. Cognitive impairment is much more common than ICH, this is usually clinically diagnosed as Alzheimer disease. Besides, due to the risk of recurrence hemorrhage is high, the use of antiplatelet agent or anticoagulant agent should be used carefully. During intracerebral hemorrhage occurred, these agents should be held. However, when intracerebral hemorrhage is stable, added back these agents should be concerned. The safety use of antiplatelet therapy in patients with CAA is considered controversial. Some studies showed antiplatelet therapy had a higher frequency of microhemorrhage. Other studies demonstrated that the risk of antiplatelet agents on ICH recurrence and severity was smaller than that for anticoagulant agents. We should evaluate the pros and cons of antiplatelet or anticoagulant agents carefully when ICH is stable. In this case, due to the history of coronary artery disease and end stage renal disease, aspirin is necessary and should be added back.

Conclusion:

Cerebral amyloid angiography is an uncommon cause of spontaneous ICH and often found in elderly. Without tissue proof, we could diagnose probable/possible CAA by brain image and clinical history. Due to high risk of recurrence ICH, the use of antiplatelet or anticoagulant agents should be carefully prescribed.

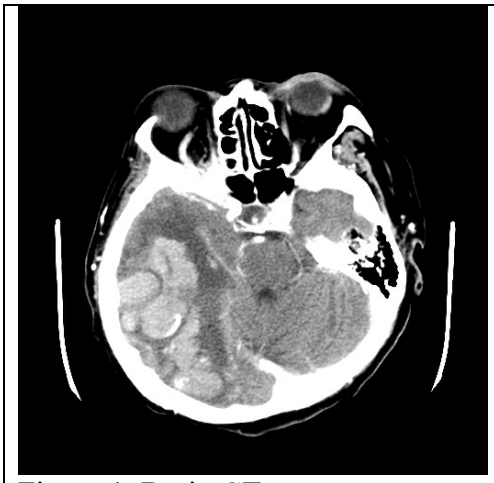

Figure 1. Brain CT

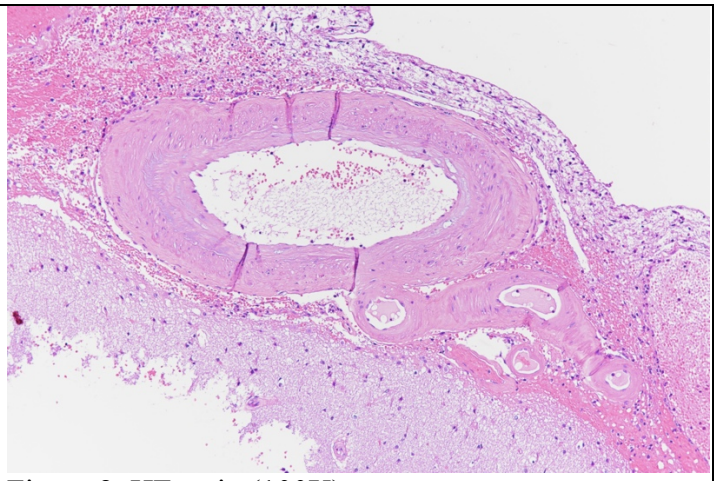

Figure 2. HE stain (100X)

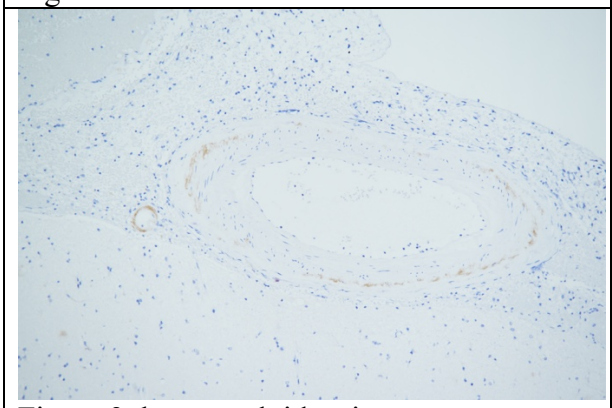

Figure 3. beta amyloid stain

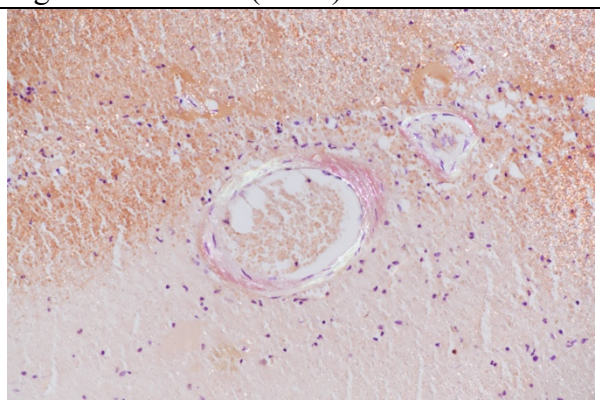

Figure 4. congo-red stain