Cerebrovascular Amyloidosis with Cerebral Hemorrhage*

K. Jellinger

Division of Neuropathology, Neurological Institute of the University of Vienna, Schwarzspanierstr. 17, A-1090 Wien, and Dept. of Neurology, Lainz-Hospital Vienna, Wolkersbergenstr. I, A-1130 Wien, Austria

Summary. More than 1400 necropsies performed on patients with either a non-traumatic cerebral hemorrhage (400 cases) or with dementia over the age of 55 (1010 cases), or both, have been reviewed. There were 15 cases in which a cerebral hemorrhage had occurred together with cerebral amyloid angiopathy all of whom had been demented. Eight of the 15 patients were hypertensive. The 7 non-hypertensives showing only the amyloid change included two cases of "atypical" Alzheimer's disease with acute neurological features, and 5 cases of senile dementia (aged 72 to 78 years) coupled with focal neurological disorders. In the hypertensive patients, aged 67 to 86 years, with a progressive dementing syndrome and acute neurological signs, multiple ball-like hemorrhages (7 cases) and/or cerebral hematomas (3 cases) were associated with a combination of amyloid and hyalinar (hypertensive) angiopathy, often affecting segments of the same pial and cortical vessels. From these data and recent reports on lethal cerebral hemorrhage occurring spontaneously or after neurosurgical procedures in demented old people, cerebral amyloid angiopathy, which is not necessarily associated with systemic amyloidosis or severe (pre)senile cerebral degeneration, may be considered a rare but important cause of cerebral hemorrhage in the aged. The "vascular" type of presenile dementia, occasionally complicated by focal cerebrovascular lesions or bleeds, is considered a variant of Alzheimer's disease. The mechanism leading to formation of cerebral amyloid is unknown.

Key words: Cerebrovascular amyloidosis – Congophilic angiopathy – Cerebral hemorrhage – Alzheimer's disease – Senile dementia.

Zusammenfassung. Über 1400 Autopsien an Patienten mit nicht-traumatischen Hirnblutungen (400 Fälle) oder mit organischer Demenz jenseits des 55. Lebensjahres (1010 Fälle) bzw. deren Kombination wurden untersucht. Es fanden sich 15 Fälle, bei denen Hirnblutungen bei cerebraler Amyloidangiopathie aufgetreten waren. Bei allen Patienten bestand eine Demenz. 8 der 15

* Dedicated to Prf. St. Környey on the occasion of his 75th anniversary

Introduction

Cerebral amyloid angiopathy [8] or congophilic angiopathy [27], also referred to as plaque-like ["drusige"] angiopathy [31] or dyshoric angiopathy [24, 36], in which amyloid deposits within and around the walls of the small pial and intracerebral vessels have been demonstrated by both light [4, 8, 22, 31, 32, 41] and electron-microscopy [23, 29, 30, 39, 44], is a common but inconstant finding in Alzheimer’s disease and senile dementia [6, 10, 22, 27, 32, 36, 43], and appears as well in nondemented old people [4, 9, 22, 27, 43], and in aged dogs [4, 29, 37, 44]. Cerebrovascular amyloidosis (CVA) in younger patients is rare [12], but has been observed in a demented boxer [3] and in elderly mongols [28, 32].

Recently, the importance of CVA as a cause of non-hypertensive hemorrhage has been stressed [9, 15, 16, 39, 42], and hereditary cerebral hemorrhage associated with amyloid deposits in the brain arteries has been described [12]. CVA, which is often combined with senile parenchymal changes, may give rise to multiple small hemorrhages and cortical microinfarcts or even to large hematomas which are the common CNS complications in hypertensive disease [1, 26, 41]. While hypertensive angiopathy, usually seen in malignant hypertension [1], is rare in old demented people with or without hypertensive cardiovascular disease [9], a simultaneous affection of the pia-cortical vessels by hyalinosis and amyloid angiopathy has been observed [10, 14, 22, 27, 40].

The purpose of this report is to study the incidence and clinical significance of CVA complicated by hemorrhage. The material consists of 15 cases with congophilic angiopathy, eight of which were associated with massive cerebral hemorrhage, while small old and/or recent bleeds were present in 11 of these brains.