Cerebral amyloid angiopathy (CAA) is a neurovascular disease characterized by b-amyloid fibrils deposited in the walls of cerebral blood vessels (Biffi & Greenberg 2012). A contemporary systematic overview paper which collectively studied the results of four population-based pathological studies concerning the link between the presence of CAA (of any severity) and dementia found that 55-59% of patients suffering from dementia suffered from CAA as well, while a CAA prevalence of only 28-38% was determined in patients without dementia (Knouse et al., 2001). CAA microbleeds are detected on MRI to occur in greater than 30% of adults over the age of 70, and are potentially even more prevalent given the amount of symptomatic individuals with micro bleeds (Vinters et al., 2006). CAA is diagnosed in various settings including stroke units, memory clinics and geriatric psychiatry. CAA is observed in community dwelling populations as well (Vernooij et al., 2008). Risk factors in the development of CAA include age (Vinters 1987) and a genetic factor, apolipoprotein E alleles (Verghese et al. 2011). Clinical presentations including neuropsychiatric presentations were described in the last two decades. These include symptomatic intracerebral haemorrhage, cognitive impairment and dementia, rapidly progressive cognitive and neurological decline and transient neurological symptoms (Chard et al., 2012). Various neuropsychological manifestations have been described which include impairment in perceptual speed, episodic memory, semantic memory, attention and executive functions, and global cognitive impairment. Neuropsychological manifestations included a new manifestation of high impulsivity, in addition to organic personality change, and depression. This study focuses on neuropsychological impairments and psychiatric manifestations observed in CAA patients and discusses the possibility of a neuropsychological profile for CAA.

Background

Cerebral amyloid angiopathy (CAA) is a neurovascular disease characterized by b-amyloid fibrils deposited in the walls of cerebral blood vessels. A contemporary systematic overview paper which collectively studied the results of four population-based pathological studies concerning the link between the presence of CAA (of any severity) and dementia found that 55-59% of patients suffering from dementia suffered from CAA as well, while a CAA prevalence of only 28-38% was determined in patients without dementia. CAA microbleeds are detected on MRI to occur in greater than 30% of adults over the age of 70, and are potentially even more prevalent given the amount of symptomatic individuals with micro bleeds. CAA is diagnosed in various settings including stroke units, memory clinics and geriatric psychiatry. CAA is observed in community dwelling populations as well. Risk factors in the development of CAA include age and a genetic factor, apolipoprotein E alleles. Clinical presentations including neuropsychiatric presentations were described in the last two decades. These include symptomatic intracerebral haemorrhage, cognitive impairment and dementia, rapidly progressive cognitive and neurological decline and transient neurological symptoms. Various neuropsychological manifestations have been described which include impairment in perceptual speed, episodic memory, semantic memory, attention and executive functions, and global cognitive impairment. This study focuses on neuropsychological impairments and psychiatric manifestations observed in CAA patients and discusses the possibility of a neuropsychological profile for CAA.

Discussion

In our case series we could not demonstrate histopathological confirmation of CAA. However these cases all meet the modified Boston radiological criteria for probable CAA (Figure 1). MRI findings in these cases (see figure 2) all include impaired memory and visual hallucinations compounded by anxiety and apathy. This culminated in an attempt to jump out of a moving car. Following admission to a psychiatric unit she was diagnosed with delirium secondary to a urinary tract infection. A brain CT showed extensive presumptive pathological calcification within the basal ganglia and cerebellum suggestive of microbleeds and small vessel white matter ischaemic change. The patient died six months later from a cardiac arrest at her home. The pattern of frontal cognitive dysfunction was already highlighted in the CAA population by Kong et al. 2120. In clinical settings modified the Boston criteria allow for probable or possible CAA diagnosis or exclusion of diagnosis of CAA (Figure 1). The Modified Boston criteria were proposed in 1999 in order to incorporate superficial siderosis into the radiological diagnosis of probable CAA (Linn et al.). They comprise of combined clinical, imaging and pathological parameters, and are based upon the original Boston criteria (Klawans et al., 1991). The Modified Boston criteria for diagnosis of probable CAA was pathologically validated in 2001, and compared to the Boston criteria had an increase in sensitivity (95%, 95% confidence interval (CI) 76% to 99%) with only a modest decrease in specificity (86%, 95 CI 62% to 93%).

CAA is mainly a neurological illness, possibly may present with psychiatric symptoms. Currently there are no studies that address the psychiatric manifestations of CAA in a systematic way. We report a case series describing three cases with neuropsychiatric manifestations presented to a geriatric psychiatric unit.

Neuropsychological manifestations (Table 1) of our cases include cognitive impairment, organic personality change, depression, focal temporal lobe seizures and delirium. A direct causality cannot be ascertained in these cases, but it is possible that CAA may have caused or contributed to current presentations. Other neuropsychological issues were demonstrated in these cases including utilization of poor impulse control, impaired concentration, poor cognitive flexibility, processing speed, impaired free recall, executive function and apathy. All these disturbances were described in previous studies (Gurl et al. 2012; Chard et al., 2012; Boyle et al., 2015), though there are no reports on organic personality change in similar cases or accounts of possible psychiatric presentations.

Conclusion

The clinical spectrum of CAA continues to grow. Despite remarkable recent interest, CAA remains under-recognised by neurologists and stroke physicians (Chard et al., 2013). At present an awareness needs to be created among psychiatrists to recognise cerebral amyloid angiopathy as a possible cause of psychiatric presentations. More insight is needed in demonstrating psychopathology and its relation to CAA. Case control studies are needed to delineate possible causes of psychiatric presentations and its relation to CAA.