Clinical and methodological confounders in assessing the cerebellar cognitive affective syndrome in adult patients with posterior fossa tumours

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The Cerebellar Cognitive Affective Syndrome (CCAS) was first described by Schmahmann and Sherman as a constellation of symptoms including dysexecutive syndrome, spatial cognitive deficit, linguistic deficits and behavioural abnormalities in patients with a lesion in the cerebellum with otherwise normal brain. Neurosurgical patients with cerebellar tumours constitute one of the cohorts in which the CCAS has been described. In this paper, we present a critical review of the literature of this syndrome in neurosurgical patients. Thereafter, we present a prospective clinical study of 10 patients who underwent posterior fossa tumour resection and had a detailed postoperative neuropsychological, neuropsychiatric and neuroradiological assessment. Because our findings revealed a large number of perioperative neuroradiological confounding variables, we reviewed the neuroimaging of a further 20 patients to determine their prevalence. Our literature review revealed that study design, methodological quality and sometimes both diagnostic criteria and findings were inconsistent. The neuroimaging study (pre-operative, n = 10; post-operative, n =10) showed very frequent neuroradiological confounding complications (e.g. hydrocephalus; brainstem compression; supratentorial lesions and post-operative subdural hygroma); the impact of such features had largely been ignored in the literature. Findings from our clinical study showed various degree of deficits in neuropsychological testing (n =1, memory; n = 3, verbal fluency; n = 3, attention; n = 2, spatial cognition deficits; and n =1, behavioural changes), but no patient had full-blown features of CCAS. Our study, although limited, finds no robust evidence of the CCAS following surgery. This and our literature review highlight a need for guidelines regarding study design and methodology when attempting to evaluate neurosurgical cases with regard to the potential CCAS.

Neuroimaging study

For this part of the study, the neuroimaging findings (CT/MRI) of patients (10 pre-operative cases and 10 post-operative cases) diagnosed with cerebellar tumours, in the Division of Clinical Neurosciences, Western General Hospital of Edinburgh, were randomly selected and evaluated by two independent reviewers Cases of vestibular schwannoma and tumours extending into or from the brain stem were excluded. In addition to the cerebellar/posterior fossa lesion itself, we were interested in the incidence and nature of associated neuropathological abnormalities.

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In particular, the variables analysed were as follows: neuropathology of the lesion, whether intrinsic (gliomas and metastatic) or extrinsic (meningiomas and subarachnoid cyst), any coexistent hydrocephalus and/or signs of raised intra-cranial pressure; degree of associated tissue oedema caused by the lesion; distortion of brainstem; along with lesion size and location, whether right, left, or vermian; other abnormalities such as concurrent lesions in the cerebellar and/or cerebral hemispheres were also described. In the post-operative imaging, we were interested in the incidence and nature of additional abnormalities such as post-operative haematoma; subdural hygroma of the infratentorial and supratentorial compartments; hydrocephalus; completeness of resection of the lesion; and the presence and size of any In our clinical study, patients showed variable and at times abnormal results which could not be explained by mood or motor disturbance and which were at odds with estimated premorbid IQ and results in other domains of cognition. It is perhaps adds weight to the notion that persons with cerebellar dysfunction may present with some degree of cognitive disturbance, but any conclusions are hampered by the lack of a control group and the small numbers in our study. The use of a control group in any prospective surgical study could be addressed by assessing patients who have neurosurgery for adult Chiari Malformation syndromes, since these patients have a posterior fossa craniectomy without any direct interference with the integrity of cerebellar tissue.

We argue that it is worth pursuing a larger study of cognitive function in persons with cerebellar tumours for the following reasons; our clinical study shows that it is possible to administer a comprehensive test battery in patients approached within 2 weeks of surgery. Such a battery applied post-operatively can be supplemented with pre-operative qualitative reports from next of kin/close family. The tumour group has the advantage that they are relatively young and therefore the concern about confounding vascular or Alzheimer ' s related cognitive impairment is minimized. Neither is there concern about supratentorial cognitive impairment associated with other neurodegenerative groups. Another compelling reason for further

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study of CCAS features in neurosurgical patients is that this cohort can provide insights into the pathophysiological basis of cerebellar cognitive dysfunction. Although our radiological studies are simplistic, and rather qualitative, modern neuroimaging techniques such as DTI, fMRI and volumetric assessment can enable much more precise anatomicofunctional correlates. As demonstrated, however, it is unlikely that such studies will be free of various confounders, particularly hydrocephalus, and thus, integrating these into anatomicofunctional correlates will be challenging.

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