Paramedian diencephalic syndrome

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The clinical characteristics of paramedian diencephalic syndrome (PDS) are described on the basis of two recently observed cases: one with paramedian thalamic infarct, the other with paramedian thalamopeduncular infarct. Analysis of the clinical symptoms and the results of CBF study show that PDS is a complex neurological syndrome characterized by symptoms due to both anatomical lesions and functional disconnections.

Key Words: paramedian thalamic infarct — thalamopeduncular infarct — dementia — SPECT.

Introduction

Paramedian diencephalic syndrome (PDS) is an infrequent clinical condition observed in cases of paramedian thalamic and thalamopeduncular infarct [8]. It results from the occlusion of a thalamostubthalamic perforating artery (the paramedian thalamic artery), which supplies the paramedian part of the upper midbrain and thalamus, including the intralaminar nuclear group and most of the dorsomedial nucleus [20]. On the basis of two recently observed cases, one with paramedian thalamic infarct and another with paramedian thalamopeduncular infarct, we here analyse the clinical characteristics of this syndrome and make some physiopathological considerations.

Case reports

Patient No. 1: Paramedian thalamic infarct

A 73 year old right-handed woman was found unarousable in bed. She had been well the previous evening, and her past health record was good apart from moderate arterial hypertension which had been treated with ace-inhibitors since she was 70 years old. On admission to the hospital of her town, she was stuporous and showed slight paresis and showed slight paresis and hypertonia on the right side. Laboratory investigations including T3, T4, FT3, FT4 and TSH were normal. Chest X-ray, ECG, echocardiography and cerebrospinal fluid were also normal, as were a Brain CT scan performed on the day of admission, a supraaortic vessel duplex scan and transcranial doppler. Carotid echotomography revealed a small calcific atheromatous plaque at the righ bifurcation. EEG showed some 3 Hz slow waves on both sides. Two days after admission, although still stuporous, the patient was able to speak when vigorously stimulated. She appeared to be disoriented and had hallucinations and confabulations. Over the following days, there was a noticeable improvement in alertness but nevertheless, she was still slightly drowsy and disoriented 20 days after the admission. She was admitted to our hospital thirty days after the onset of the disease. On admission, she was drowsy, confused and disoriented in terms of both time and place. Neurological examination showed down-gaze paralysis, partially limited upgaze, ocular convergence failure, ataxic gait with retropulsion and symmetrical brisk deep tendon reflexes. Fifteen days later, the patient was still confused although less drowsy. The results of neurological examinations remained unchanged.

A neuropsychological evaluation was made using non-standardised tests for attention, language, visuo-spatial functions and memory. The patient tended to fall asleep when external stimulation ceased; when awake, her attention would wander to distracting stimuli. She was disoriented to both time and place and not cooperative. Spontaneous speech was reduced and literal paraphasias were present. Comprehension of spoken speech was normal. Body image was unimpaired, as was right-left orientation. Simple oral calculations were...
correctly executed. Writing showed some motor dysgraphias and a great deal of motor perseverations. Reading could not be assessed. Her marked difficulty in executing transitive and intransitive motor commands suggested ideomotor apraxia. Copies of various drawings revealed constructional apraxia. There was no hemineglect syndrome. Memory for both anterograde and retrograde events was severely impaired and accompanied by confabulations.

Proton density and T1 weighted brain MRI images recorded 30 days from the onset of the disease showed two hyperintense areas within the medial region of both thalamic nuclei, compatible with ischemic lesions (Fig. 1a-2a), and a small hyperintense area in the caudate nucleus. A SPECT-CBF study was performed 33 days after the onset of the disease, using 15 mCi of Tc-99m HM-PAO and a dedicated brain camera (Mediomatic 564). After computerised image reconstruction, a set of six 10 mm thick transverse slices was obtained (9 mm spatial resolution in the transverse plane) from the cerebellum up to the cortical mantle, revealing reduced perfusion of both frontal lobes (prevailing on the left side) and the left temporal lobe; the thalami appeared to be normally perfused (Fig. 3a).

Patient No. 2: Paramedian thalamopeduncular infarct

We admitted a 68 year old right-handed man, who had experienced sudden dizziness followed by loss of consciousness after a few minutes. His past medical history included lung TBC when he was 35 years old, lung embolism when he was 55 and thyroid carcinoma surgically removed when he was 65 followed by hypoparathyroidism treated with calcium, calciferol and thyroxine.

On admission, the patient’s blood pressure was 110/70, and he had an arrhythmic pulse of 105. ECG revealed atrial fibrillation. He was comatose. Neurological examination showed that the right pupil was mydriatic and unresponsive to light, there was a lack of motor responses in the left and purposive responses to noxious stimuli in the right arm, and bilateral Babinski sign. The laboratory analyses were normal except for a blood calcium level of 7.4 mg/dl (normal values 8.7-10.2 mg/dl) and a phosphorus level of 5.8 mg/dl (normal values 2.5-4.9 mg/dl). An unenhanced brain CT scan performed on admission was normal. The results of a supra aortic vessel duplex scan was normal, but carotid echotomography showed two calcific atheromatous patches at the bifurcation of the right internal carotid artery. Transcranial doppler and echocardiography were normal. Two days later, the patient was stuporous but capable of following a few verbal commands when stimulated. He showed mild left facio-brachial paresis, hypertonia on both sides, right-sided non-responsive mydriasis, right eyelid ptosis, the loss of adduction of the right eye, vertical gaze paralysis and bilateral Babinski sign. There was no relevant change in his clinical condition during the following 45 days. After this period, his consciousness began to improve and he finally became fully aware. He answered easy questions, but showed memory dysfunction (see mental status examination). He could feed himself and walk with some help. Up- and downgaze were limited, as was adduction of the right eye. Mild weakness, hyper-reflexia and Babinski sign were still present on the left side. Four months after discharge, he is able to walk and his upgaze has improved; memory dysfunction remains unchanged.

Mental status examination. The patient was poorly cooperative and tended to fall asleep as soon as external stimuli ceased. He was easily distracted and showed long latency in both verbal and motor responses. Emotional responses were generally poor and flat. He was severely disoriented in terms of both time and place, and showed ideational and motor perseverations. Speech was non-fluent, akinetic and simplified in syntactic structure, suggesting motor transcortical aphasia. Understanding was intact but not testable with complex material because of attention disturbance. Word repetition was intact. Writing and reading could not be assessed. Simple drawings and mimic gestures suggested ideational and constructional apraxia. Memory and learning were globally and severely impaired, as well as being associated with confabulations.

An enhanced brain CT scan performed three days after the onset of disease showed a hypodensity within the right ponto-mesencephalic matter extending upwards to the median mesencephalic region (Fig. 1b), and two bilateral hypodense areas within the thalamic nuclei (Fig. 2b) compatible with ischemic lesions. Brain MRI performed after 20 days revealed the presence of thalamic and periaqueductal hyperintense areas on T2 weighted images.

A SPECT-CBF study using the method previously described was performed 60 days after the onset of disease. It revealed reduced perfusion in both prefrontal regions and the left temporal region. As in case No. 1 the thalami appeared to be normally perfused (Fig. 3b).

Discussion

Given the anatomical variability of the vascular system supplying the paramedian thalamic region, there are three main topographical arrangements in paramedian thalamic infarcts: unilateral and bi-