their activated derivatives. Human plasma-derived FVII concentrate has also been used. None of these products is acceptable to Jehovah's Witnesses, as all are derived from human blood. However, non-blood and recombinant alternatives are acceptable.[11] The patient reported here illustrates the usefulness of rFVIIa in this difficult group of patients, the anticoagulated Jehovah's Witness with life-threatening bleeding.

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Cerebellar diaschisis and contralateral thalamus hyperperfusion in a stroke patient with complex regional pain syndrome

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Abstract

We present a right-hemispheric stroke patient with complex regional pain syndrome (CRPS). The regional cerebral blow flow (rCBF) as determined using single photon emission computed tomography (SPECT) showed contralateral increase of tracer uptake in the left thalamus accompanied by crossed cerebellar diaschisis (CCD) in the left cerebellum. After rehabilitation, the CRPS in the right upper extremity recovered, although hemiplegia persisted on the left limbs. The rCBF determined a second time using SPECT showed that uptake was normal in the bilateral thalami, basal ganglia and bilateral cerebella. At a 6-month follow-up, the CRPS had not recurred. Our findings show that analysis of rCBF by SPECT is useful for the clinical evaluation and follow-up of CRPS. To the best of our knowledge, this is the first reported case with this particular pattern of symptoms amd symptom resolution.

Keywords: Complex regional pain syndrome; Thalamus; Crossed cerebellar diaschisis

1. Introduction

Complex regional pain syndrome (CRPS), a neuropathic pain disorder, is characterized by over reaction to

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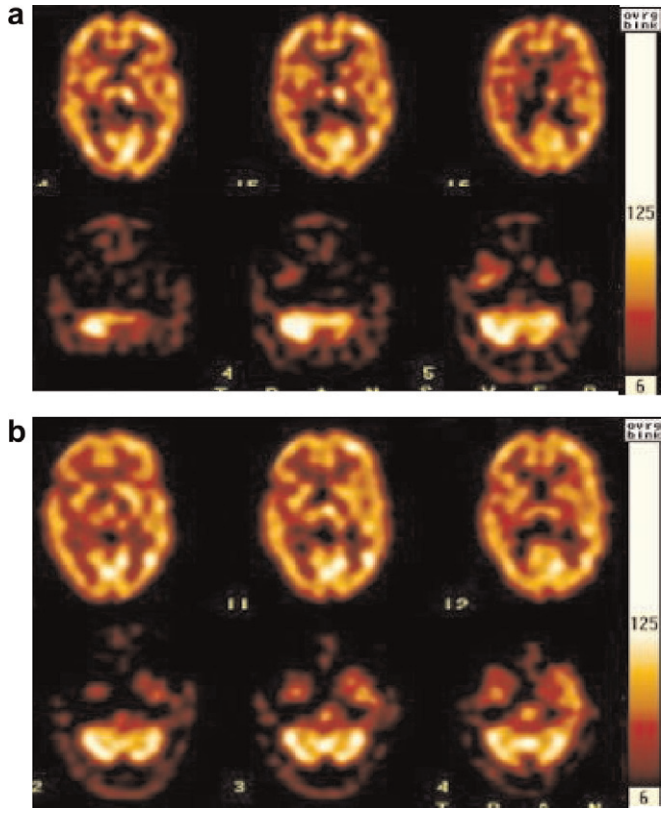

Fig. 1. SPECT brain perfusion images obtained before rehabilitation treatment (a) and after 14 days of rehabilitation (b). Note the relatively increased tracer uptake in the left thalamus before treatment and decreased blood perfusion in the left cerebellar hemisphere in the left cerebellum after 14 days of rehabilitation.

trauma or nerve injury of the extremities, or may also be the aftermath of stroke or myocardial infarction.[1] A diagnosis of CRPS is based on clinical signs and symptoms.[2] The thalamus plays an important role in the early development of CRPS.[3–6] Most articles in the literature confirm a reduction in perfusion in the contralateral thalamus related to CRPS or chronic pain[4] or an increase related to post-stroke central pain.[7] Only a few cases of hyperperfusion in the contralateral thalamus have been reported for patients with CRPS.[4,5] We present the case of a 64-year-old man with acute stroke and subsequent CRPS. The regional cerebral blow flow (rCBF) as determined using single photon emission computed tomography (SPECT) differed greatly before and after resolution of the CRPS.

2. Case report

A 64-year-old man was admitted to our rehabilitation unit after suffering a stroke and developing CRPS. He had a 10-year history of type 2 diabetes mellitus and hypertension controlled by regular medication, and had suffered a right stroke that caused left hemiparesis 5 years previously. Two months before being transferred to the rehabilitation unit, he presented with sudden-onset right-sided weakness due to acute stroke. Brain MRI revealed an acute infarction area of about 1 cm^2 in the periventricular corona radiata of the left parietal lobe, together with an old infarction area in the periventricular white matter of the right parietal lobe and right dorsal thalamus. He was discharged in a stable condition after 3 weeks. He then presented again with pain in his right forearm and right hand, and the grip strength of the right finger gradually decreased. His right forearm and right hand were moderately swollen, and a mild flexion contracture of the right fingers was noted. Allodynia and hyperpathia were also noted. His right upper distal extremity was warmer and redder than the left. The hand disorder was diagnosed as CRPS following the criteria of the International Association for the Study of Pain.[2]

Serum profiles were within normal ranges. Brain SPECT revealed a contralateral increase of tracer uptake in the left thalamus and crossed cerebellar diaschisis (CCD) in the left cerebellum (Fig. 1a). During the hospital stay, the patient's CRPS symptoms improved progressively after 2 weeks of rehabilitation treatment. The CRPS in the right upper limb remitted, although hemiplegia persisted on the left side. The second SPECT scan showed normal uptake in the bilateral thalami, basal ganglia and bilateral cerebella (Fig. 1b). At a 6-month follow-up, CRPS had not recurred.

3. Discussion

CRPS has been demonstrated to be closely associated with central lesions or peripheral damage caused by fracture, direct trauma or tissue injury.[1] In most patients with CRPS, a reduction in thalamic perfusion has been found, but Fukumoto et al. performed SPECT for 10 patients with CRPS and found contralateral thalamic hyperperfusion in some patients with acute CRPS and hypoperfusion in other patients with chronic CRPS.[4] Researchers have postulated that contralateral thalamic perfusion increases in the acute stage as a reaction to pain, and gradually decreases as an adaptive response or possible feedback mechanism to minimize nociception in the chronic stage.[4–6] Cesaro et al. found thalamic neuronal hyperactivity associated with hyperpathic syndromes in two patients with central post-stroke pain; in these patients they used brain SPECT to identify increased rCBF in the thalamus contralateral to the painful limbs.[7] The reaction of the thalamus might vary in response to external stimulation or just change naturally over time, which would explain why we observed the changes in the rCBF in the contralateral thalamus after rehabilitation in the present case.

CCD is a phenomenon of hypoperfusion and hypometabolism in the cerebellum after a contralateral supratentorial lesion.[7] Our case had CRPS with hyperperfusion of the contralateral thalamus, which coincided with CCD at the left, pre-existing surpatentorial lesion. There is a strong relationship between CCD and hyperperfusion of the contralateral thalamus. The patient did not need medication because the CRPS responded extremely well to rehabilitation. The second SPECT scan revealed a reversal of the contralateral thalamic hyperperfusion and restoration of the contralateral cerebellar blood flow. Gold and Lauritzen

pointed out that a functional neocortical lesion results in an abrupt and severe decrease in contralateral Purkinje cell spiking activity and blood flow, which is rapidly restored when the excitatory input to the cerebellum returns.[8] In our present case, 14 days of rehabilitation produced an improvement in CRPS, accompanied by a reversal of the hyperperfusion of the contralateral thalamus and resolution of the CCD. These results indicate that the cerebellar thalamocortical axis plays an important central role in stroke with CRPS, and may reorganize connections between the spinal cord, cerebellum and sensory thalamocortical axis. The SPECT results in this case indicate that analysis of rCBF with SPECT is useful for the clinical evaluation and follow-up of CRPS.

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Secondary intracranial hypertension with acute intracranial pressure crisis in superficial siderosis

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Abstract

Superficial siderosis of the central nervous system is a very rare disease related to hemosiderin deposits in the brain, brainstem, cerebellum and spinal cord due to chronic subarachnoid hemorrhage. Chronic increased intracranial pressure develops in about one-third of affected cases. We report a patient with superficial siderosis and sudden intracranial pressure crisis. A 29-year-old man experienced a subacute episode of headache, tinnitus and blurred vision. Magnetic resonance imaging of the brain revealed hemosiderin deposits characteristic of superficial siderosis. Extensive diagnostic work-up excluded causative pathologies of bleeding. Lumbar puncture and continuous intra-ventricular cerebrospinal fluid (CSF) pressure monitoring revealed continuous CSF pressure increase. Implantation of a ventriculo-peritoneal shunt led to complete clinical recovery. Our case emphasizes that patients with superficial siderosis may present with sudden elevation of intracranial pressure due to chronic intracranial hypertension. In this situation permanent CSF drainage provides a useful therapeutic option.

Keywords: Superficial siderosis; Central nervous system; Intracranial hypertension; Intracranial pressure; Chronic subarachnoid hemorrhage

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