

Bálint-like syndrome as an unusual representation of non-convulsive status epilepticus

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ABSTRACT – The clinical signs of posterior cortex dysfunction are, due to their paucity and subtlety, very often ignored as non-specific during clinical evaluation of non-convulsive status epilepticus. Therefore, focal non-convulsive status epilepticus emerging from the posterior cortex, and especially the parietal lobes, can be fairly under-recognised. We report a 66-year-old patient with focal non-convulsive status epilepticus presenting as isolated Bálint-like syndrome, successfully treated to full clinical and electrophysiological recovery. The diagnostic and pathophysiological features are discussed. Focal non-convulsive status epilepticus can be associated with negative phenomena such as neuropsychological deficits mimicking those detected more often in degenerative and vascular brain diseases. *[Published with video sequences]*

Key words: Bálint-like syndrome, chronic epilepsy, nonconvulsive status epilepticus, parietal lobe, FDG-PET



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Non-convulsive status epilepticus (NCSE) presents in protean ways, accounting in part for its under-recognition and underdiagnosis (Drislane, 2000). Focal status epilepticus restricted to non-motor brain regions may well be overlooked if neuropsychological and behavioural changes are not observed

cautiously. Ictal cognitive deficits can be very variable in focal NCSE (Geier, 1978). Neuropsychological testing during NCSE may reveal findings as subtle as discrete reduction of visuo-constructive skills, disturbances of verbal memory tasks, and use of neologisms, perseveration, and reduced ability

to produce speech automatisms (counting and naming weekdays) (Profitlich *et al.*, 2008).

Bálint's syndrome is comprised of psychic paralysis of gaze (oculomotor apraxia), misreaching, in the absence of elementary visuo-motor impairments (optic ataxia), and spatial disorder of attention (simultanagnosia) (Bálint, 1909). This rare syndrome usually affects bilateral posterior parietal lobes and has been described with various conditions (degenerative dementia, vascular, demyelinating, prion or infectious diseases) (Moreaud, 2003).

We report a patient with an unusual presentation of focal NCSE consisting of the unilateral clinical features observed in Bálint's syndrome.

Case study

A 66-year-old, right-handed woman with early normal development, and absence of epilepsy risk-factors (febrile seizures, brain trauma or infectious diseases), suffered from medically intractable seizures from the age of 49. Since that age, she presented with brief automotor seizures (staring, intensive breathing, and repetitive clenching of both fists). No preceding aura was reported. A seizure frequency of 4-6/week in the last three years could not be altered by various medications. Reportedly, she had intermittent episodes of waxing and waning confusion lasting for hours or days with various problems in everyday activities over the same period (slowness in mentation, erroneous naming of persons, or inappropriate use of accessories), but a history of status epilepticus was not obtained.

The patient was referred to our video-EEG monitoring unit for evaluation of her epilepsy and mental capacity. At the time of admission, she was being treated with valproic acid (1,000 mg per day) and carbamazepine (1,200 mg per day). Her physical and neurological examinations were normal. Mini-Mental State Examination (MMSE) revealed moderate cognitive impairment (score 20/30) at the moment of examination. No clinical seizures were observed.

Video-EEG monitoring (surface electrodes placed according to the 10-20 international system with additional anterior temporal electrodes, ECG and respiration) was commenced, and antiepileptic therapy was gradually discontinued over five subsequent days. Three interictal EEG epileptiform populations were recorded in the: right anterior temporal (maximum T2>F8 electrode; 80% of interictal discharges), right frontal (maximum F8 >Fp2 electrode; 10% of interictal discharges) and right posterior temporal region (maximum T6>O2-P4; 10% of interictal discharges) (*figure 1A-C*). Within five days of video-EEG monitoring, six automotor seizures matching those described by witnesses and more than 50 non-clinical electrographic seizures were recorded. Ictal EEG showed right temporal onset (T2>T4 electrodes). Lamotrigine monotherapy was gradually initiated at the end of video-EEG monitoring. On the second day following video-EEG monitoring, the patient complained of visual disturbances (*"I can't control my eyes"*). Neurological examination showed mild disturbance of attentiveness (more complex tasks such as serial seven subtraction were disturbed by impaired attention, slow responses, and perseveration), appropriate orientation, normal motor performance, intact visual fields, and no signs of aphasia. The main finding

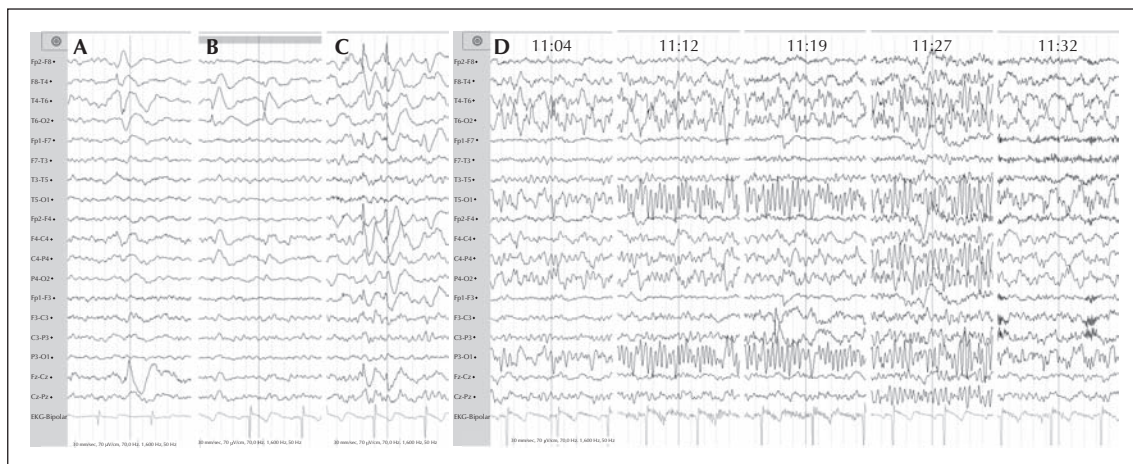


Figure 1. Interictal and ictal EEG pattern during NCSE.

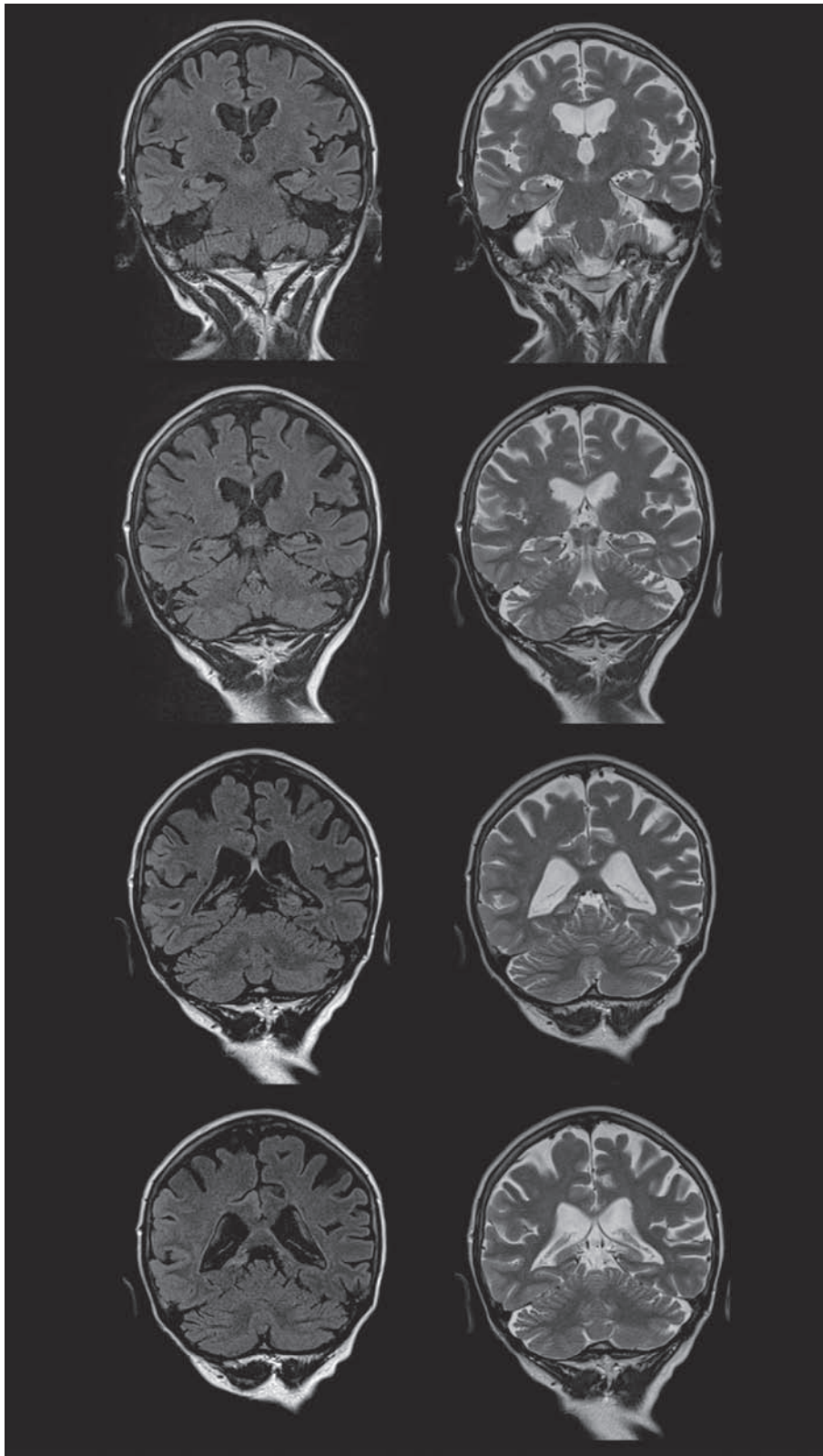


Figure 2. Brain MRI shows increased cortical thickness and FLAIR/T2 hyperintense signal in the posterior aspect of the middle and inferior temporal gyrus, posterior aspects of the inferior parietal lobule, and parieto-occipital junction on the right hemisphere, in addition to global mild-to-moderate cortical atrophy.

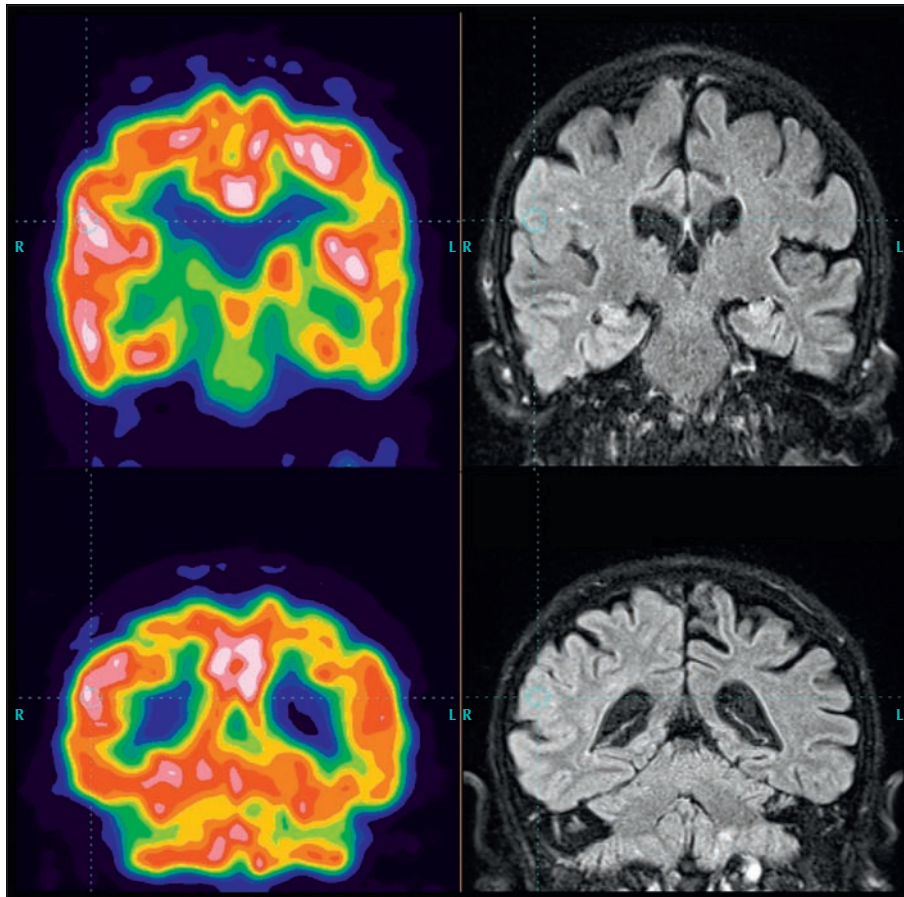


Figure 3. FDG-PET hypermetabolism matches the brain MRI lesion on corregistered images.

was Bálint's syndrome, markedly pronounced on the left side (oculomotor apraxia, optic ataxia, and simultanagnosia), in addition to left hemineglect (*video sequence 1*). Simultaneous EEG recorded a prolonged ictal EEG pattern in the right parietal, occipital, and posterior temporal region (*figure 1D*). Intravenous diazepam (20 mg/2 hours) led to temporary somnolence, mild improvement of some of the clinical features (including better performance of visual tracking and visually guided movements) and moderate improvement of the EEG pattern.

Brain MRI (performed on the second day following NCSE) showed increased cortical thickness and FLAIR/T2 hyperintense signal in the posterior aspect of the middle and inferior temporal gyri, posterior aspects of the inferior parietal lobule, and parieto-occipital junction in the right hemisphere, in addition to global mild-to-moderate cortical atrophy (*figure 2*). DWI was unremarkable. FDG-PET performed on the fourth day following NCSE with simultaneous EEG (showing intermittent and repeti-

tive spikes in the right parietal and posterior temporal region) indicated symmetric bitemporal and biparietal parasagittal hypometabolism and hypermetabolism in the right parietal, posterior temporal and occipital region that corresponded to the MRI lesion (*figure 3*).

Gradually increasing doses of topiramate and lamotrigine were given and full resolution of symptoms (*video sequence 2*) and normalisation of EEG were documented on the eighth day of the focal NCSE onset. The patient was discharged on lamotrigine (550 mg) and topiramate (400 mg). Cognitive function improved and MMSE score was 26/30 at the day of discharge.

Discussion

To the best of our knowledge, features resembling those of Bálint's syndrome are not reported as a clinical presentation of NCSE. Focal NCSE emerging from

extratemporal lobes was described previously and NCSE confined to frontal lobes was estimated to be most frequent (Williamson *et al.*, 1985). Furthermore, NCSE of frontal origin is clinically and electrophysiologically well documented (Thomas *et al.*, 1999). Surprisingly, although this involves a significant portion of the surface of the human brain, reports on the posterior cortex origin of NCSE are scarce in the literature (Thomas *et al.*, 1999; Yilmaz *et al.*, 2004). The easily overlooked, subtle and non-motor signs arising from this brain region may be one of the reasons.

Negative ictal phenomena other than motor inhibition, blindness or aphasia are rare. We describe a patient who presented with unilateral (left) features matching those of Bálint's syndrome, in addition to left hemineglect, as a consequence of continuous epileptic discharges over the contralateral (right) parietal, occipital and posterior temporal region. Brain-damaged patients with lesion or dysfunction involving the parietal cortex may show a variety of neuropsychological impairments involving spatial cognition. The more frequent and disabling deficit is the hemineglect syndrome that consists of a bias of spatial representation and attention, usually contralateral to the hemispheric lesion. The deficit is more frequent and severe after damage to the right hemisphere (Çiçek *et al.*, 2007). Another less frequent deficit that has relevant localising value is optic ataxia, typically brought about by damage to the superior parietal lobule (Auerbach and Alexander, 1981). Optic ataxia (in association with oculomotor apraxia and simultanagnosia) constitutes Bálint's syndrome, which is *sensu stricto* a consequence of bilateral posterior cortex damage. Hemineglect syndrome was the prominent feature in reports by Bálint. Critical lesions in Bálint's syndrome are bilateral and occupy the parieto-occipital junction, whereas lesions in the hemineglect syndrome occupy the temporo-parietal junction. Yet, these anatomical distinctions are not clear-cut, and the behavioural overlap between these two syndromes is obvious (Rizzo and Vecera, 2002).

Additionally, our case further supports that FDG-PET can be a useful tool to better understand the interplay between discrete brain locations, the clinical manifestation of NCSE, and the location of ictal discharges. In even rarer cases with the absence of obvious ictal epileptiform EEG changes during a period of EEG recording, enhanced FDG-PET imaging may suggest the epileptic nature of subtle clinical manifestation (Stayman and Abou-Khalil, 2011; Van Paesschen *et al.*, 2007). □

Disclosures.

The authors have no financial disclosures to report. The authors have declared that no conflict of interest exists.

Legends for video sequences

Video sequence 1

Neuropsychological testing during video-EEG monitoring on the first day of NCSE.

Video sequence 2

Neuropsychological testing reveals full resolution of symptoms on the day of discharge.

Key words for video research on

www.epilepticdisorders.com

Syndrome: focal non-idiopathic parietal

Etiology: dysplasia (architectural)

Phenomenology: status epilepticus (non-convulsive); automotor seizures

Localization: posterior cortex parietal

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