

J. Ghika
J. Bogousslavsky
J. Henderson
P. Maeder
F. Regli

The “jerky dystonic unsteady hand”: a delayed motor syndrome in posterior thalamic infarctions

Received: 6 July 1993
Accepted: 25 January 1994

J. Ghika (✉) · J. Bogousslavsky
J. Henderson · F. Regli
Service de Neurologie, CHUV,
CH-1011 Lausanne, Switzerland
P. Maeder
Service de Radiologie, CHUV,
CH-1011 Lausanne, Switzerland

Abstract We report the cases of three patients with a thalamic infarct in the territory of the posterior choroidal artery involving the posterior thalamic nuclei. These patients developed delayed complex hyperkinetic motor syndromes, associating ataxia, tremor, dystonia, myoclonus and chorea, which we call “the jerky dystonic unsteady hand”. One patient had a severe myoclonic and ataxic-dystonic choreoathetosis; another showed a so-called rubral tremor (myoclonic ataxia with resting, action, and wing-beating tremor) with dystonia; and the third one had a dystonic and ataxic hand with intermittent mild action myoclonus. All of them had sensory dysfunction; two had also presented with a painful Déjérine-Roussy syndrome. All had

CT or MRI-proven infarcts in the territory of the posterior cerebral artery involving the posterior choroidal territory with an abnormal signal in the posterior area of the thalamus (pulvinar nucleus) but sparing the other thalamic, subthalamic and midbrain structures. These delayed myoclonic complex hyperkinetic syndromes have not been reported before, and we did not observe them in other topographic forms of thalamic infarcts. They may thus represent a new entity of movement disorders due to lesions in the posterior thalamic nuclei, with specificity for posterior choroidal artery infarcts.

Key words Myoclonus · Choreia
Dystonia · Rubral tremor · Thalamus

Introduction

Sensory and ataxic thalamic syndromes are well-recognized entities [1–12]. Motor hyperkinetic thalamic syndromes have been mentioned by Déjérine and Roussy [1] and Foix and Hillemand [2], but very few cases have been reported and analysed [10–14]. We had the opportunity to study three different delayed myoclonic complex hyperkinetic syndromes in CT or MRI-proven thalamic infarcts in the territory of the posterior choroidal artery [15–19] from the posterior cerebral artery territory with involvement of the pulvinar nucleus. None of the 54 patients with thalamic infarcts in another territory that we examined (inferolateral, paramedian or polar territories) showed these abnormal movements, but they were all observed in the acute phase [4]. Infarcts in this deep territory of the posterior

cerebral artery are known to present with sensorimotor syndromes and sectorial or quadrantic visual field defects, coma, cognitive or behavioural syndromes [15–19]. A delayed tremor was recently reported in such an infarct [12], but delayed complex myoclonic hyperkinetic syndromes have not yet been reported to our knowledge. The posterior choroidal artery supplies the geniculate bodies, the colliculi, the fornix, the pineal body, a part of the midbrain and the pulvinar nucleus of the thalamus, but not the lateral or posterior nuclei [14, 18, 19] (Fig. 1).

Case Reports

Patient 1 was a 90-year-old hypertensive woman with atrial fibrillation associated with mitral valve disease. She was known to have an essential familial action and postural tremor which became symptomatic when she was in her sixties. In 1972, she had a verte-

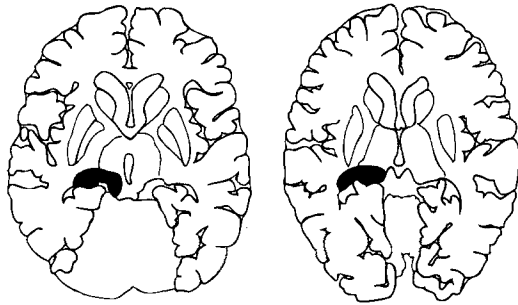


Fig. 1 Territory of the posterior choroidal artery (■) (modified from Bogousslavsky et al. [4])

brobasilar minor ischaemic incident which lasted 24 h with vertigo, left sensorimotor deficit but a persistent increase in amplitude of her action tremor on the left side. A physician thought she had Parkinson's disease and treated her successively with amantadine, trihexyphenidyl and tetrabamate with no effect. In May 1977, she had a new ischaemic event with vertigo, diffuse headache, and increased weakness on the left side with paraesthesia and dysaesthesia of the hand. On examination, there was a left upper homonymous quadrantic visual field defect, a sensory deficit predominating on the left arm for all superficial, deep and complex sensory modalities with sensory extinction but no hemineglect on the left hemibody, and dysaesthesia of the left upper extremity. A mild, slightly spastic, motor hemisindrome was present on her left side with brisk reflexes and a distal paresis of 20%, but no abnormal movements except for the essential tremor could be observed. By August 1977, she began to experience an hyperpathic disorder of the left arm (Déjerine-Roussy syndrome) together with constant, involuntary, high-amplitude involuntary movements and total loss of control of her left upper extremity. On physical examination, there was no change in the sensorimotor deficit, but a high-amplitude, hyperkinetic, very jerky hemichoreoathetotic syndrome was described. She was successively treated with amitriptyline, haloperidol and tetrabenazine without benefit.

In 1991, she was examined in our movement disorder unit. She was wheelchair-bound, totally unable to control or use her left arm for any of her daily activities. The left arm, forearm and hand were constantly restless, with irregular complex distal high-amplitude involuntary choreoathetotic jerky motions of the distal upper extremity, which she attempted to decrease by holding her left forearm with her right hand against her body. The motions were increased on stress and became myoclonic and explosive on action, sometimes ballistic-like. The fingers were in a dystonic posture in extension. She was unable to hold a glass of water, to drink without spilling all the water, and could not eat or write, button or even comb her hair or dress with her left arm. Her left shoulder had a wing-beating type of tremor at rest, and she had a high-amplitude 4 Hz "no-no" type of head titubation, a tremulous voice, and a generalized resting and action tremor of the four extremities. There was a severe left hemiataxia, much more pronounced in the upper extremity, with gross hypermetria, asynergia, and asynchronometria, pathological rebound, and a total inability to perform a rhythm or to make a ballistic coordinated motion. Only a few low-amplitude jerky motions were present on her left leg. The sensorimotor deficits were unchanged on the left hemibody. CT in 1992 showed an infarction in the territory of the posterior cerebral artery involving the territory of the posterior choroidal artery (Figs. 1, 2), and no evidence of subthalamic nucleus involvement; MRI was not performed.

Patient 2 was a 61-year-old male patient with a history of abrupt headache and coma of several days duration in 1969. After regain-



Fig. 2 Patient 1 (CT scan). Small infarct in the right posterior thalamic nuclei corresponding to the territory of the posterior choroidal artery; enlargement of the occipital horn in relation to an old superficial posterior cerebral infarct

ing consciousness, he presented with a memory deficit and a right sensorimotor syndrome with a M4 + distal deficit in strength. An angiogram was performed, but was discarded together with his chart after the legally prescribed period of time, and was therefore not available. Neither CT or MRI were available at this time. A few months later, he developed a painful Déjerine-Roussy syndrome in the right hand and upper extremity up to the shoulder and on the right sole and calf with a constant burning sensation. He had major difficulty in using his right hand owing to unawareness of its position, as well as disproportionate strength and awkwardness, but had no trouble walking. He consulted in March 1992 because of an unrelated L4 sciatic pain on the right side. On physical examination, there was a right upper homonymous quadrantic visual field defect complete loss of sensation for all superficial, deep and complex sensory modalities on the whole right hemibody, a dystonic hand with athetosis of the fingers, a right kinetic hemiataxia with hypermetria, asynergia, adiokokinesia, asynchronometria and rebound, difficulty in rhythmic and ballistic activity of the right upper extremity, which was much less pronounced in the right lower extremity. On performing the finger-to-nose manoeuvre, there were some jerky motions, better seen when he drank a glass of water. The patient not only spilled all of the water from the plastic cup, but completely broke it. MRI in 1992 (Fig. 3) performed with a Magnetom SP 63 1.5 T with T1- and T2-weighted images showed a posterior cerebral artery infarct with involvement of the posterior choroidal territory (Fig. 1), with no abnormal signal in other thalamic or subthalamic nuclei or in the midbrain.

Patient 3 was a 32-year-old man with a familial and personal history of migraine without aura, but with no other cerebrovascular risk factors. In April 1988, he complained of an abrupt, unusual headache, which was followed, half an hour later, by a right sen-

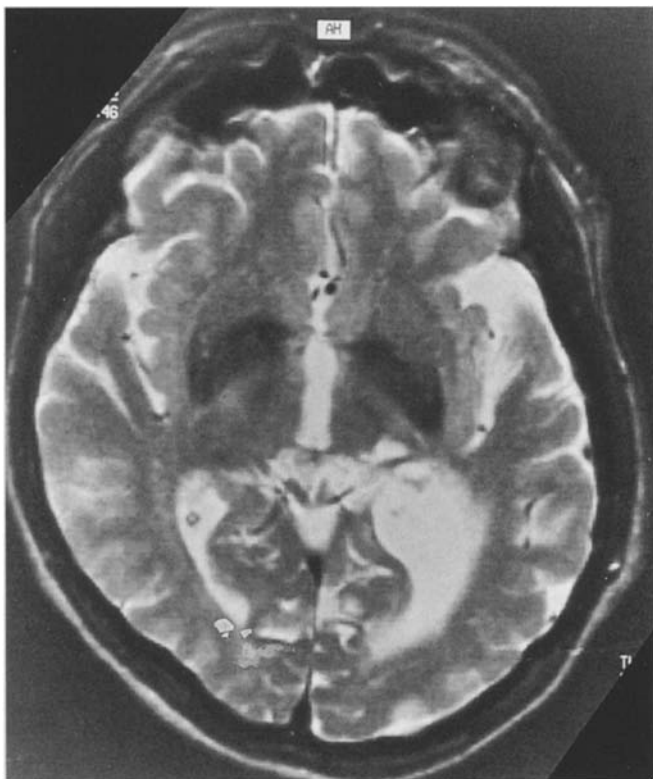


Fig. 3 Patient 2 (MRI; T2-weighted image 2500,90). Left posterior cerebral infarct with involvement of the posterior thalamic nuclei in the territory of the posterior choroidal artery



Fig. 4 Patient 3 (MRI; T1-weighted image 500,40). Left posterior thalamic infarct in the territory of the posterior choroidal artery (superficial posterior cerebral artery not shown here)

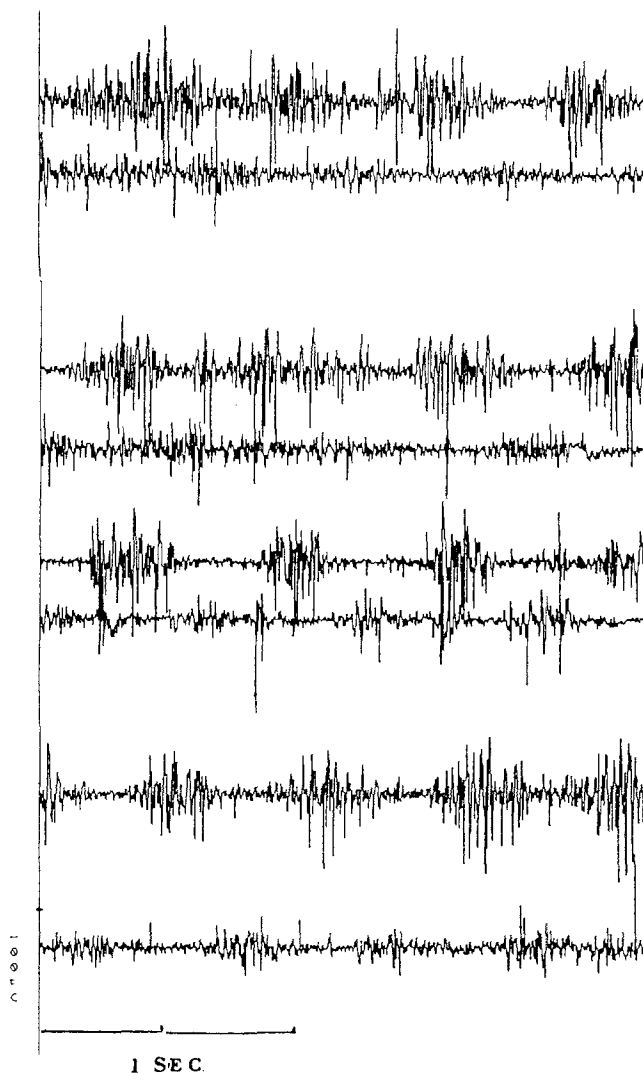


Fig. 5 EMG recording of patient 3, showing a rubral tremor with alternating patterns in antagonist muscles at 3 Hz (*lower part*) and more dystonic-athetotic pattern (*upper part*)

sorimotor deficit. On physical examination, he had a right hemianopia, which resolved in an upper quadrantanopia, a slow pursuit left optokinetic nystagmus, and a complete right hemibody sensory deficit for all epicritic, protopathic and complex sensory modalities. There was also a sensory hemiextinction without neglect, and a predominantly distal normotonic hyperreflexic motor paresis 20–30%, which resolved in 24 h. CT showed a left posterior cerebral infarct with involvement of the pulvinar. A left carotid and vertebral angiogram showed arteriovenous shunts in the territory of the left PCA compatible with a recent stroke; the posterior choroidal artery could not be seen. The CSF examination was unremarkable. The EEG showed bradyrhythmia on the left temporo-parieto-occipital lobes, and the temporo-parietal waves of sensory evoked potentials were absent on the left. By September 1988, the patient started to have “jerks of the right arm on action”. He frequently dropped things, could not write, use buttons, or use his right arm for any precise motion. MRI in September 1988 (Fig. 4), performed with a Diasonics 0.35 T T1- and T2-weighted images, showed a decreased signal in the posterior thalamic nuclei (pulvinar) and the lingual gyrus compatible with a left posterior cerebral

infarct with involvement of the posterior choroidal artery territory (Figs. 2, 3). The subthalamic nucleus and midbrain were intact on MRI. The patient was successively treated with propranolol 120 mg, clonazepam 1.5 mg, primidone 750 mg, trihexyphenidyl 20 mg and flunarizine 40 mg without any benefit.

He consulted our movement disorder unit in January 1992. On examination, the above-mentioned deficits were all present, but in the right upper extremity, there was a resting tremor of the right arm with wing-beating of the shoulder. The tremor increased on action and posture with large irregular amplitude of frequency around 3 Hz. On wrist extension, there was an athetoid, oscillating, low-frequency, high-amplitude, tremor-like movement with suggestion of dystonia of the fingers. On the finger-to-nose manoeuvre, and on drinking a glass of water with the right hand, there were myoclonic high-amplitude jerks. There was a right hemi-ataxia, almost exclusively localized on the upper extremity, with hypermetria, asynergia, adiadokokinesia, asynchronometria, pathological rebound, difficulty in beating a rhythm or producing a ballistic motion with his right hand. Writing showed ataxic tremor and myoclonus. His gait and standing were normal except that they increased the left arm tremor and dystonia. The surface EMG recording (Fig. 5) shows the classical 3 Hz alternating pattern of a rubral tremor (lower part of the recording) but also more dystonic-athetotic patterns on occasion (upper part of the recording).

Discussion

Hyperkinetic syndromes are sometimes encountered in the acute phase of a stroke. Acute hemichorea-hemiballism, asterixis, or hemidystonia, or "dyskinesias" have been reported [1–14, 20–37], and have been described more frequently with striatal or subthalamic rather than thalamic lesions. Delayed hyperkinetic syndromes are less frequently reported after strokes [12, 22–25]. Well described are delayed athetosis and dystonia [22–25], but delayed tremor has only recently been reported [12].

Thalamic motor hyperkinetic syndromes are infrequent. Déjerine and Roussy in 1906 reported choreoathetotic motions and ataxia in the "thalamic syndrome", which were described as "small movements localized to the fingers and hand, sometimes disorganized like chorea, sometimes slow and sinuous like athetosis" [1]. Foix and Hillemand [2] wrote about the "superior syndrome of the nucleus ruber or rubrothalamic syndrome" to describe a coarse intentional tremor accompanying the cerebellar ataxia, and reported choreoathetotic movements in thalamogenicular infarcts and the rubrothalamic syndrome. The choreoathetosis is described as spontaneous or motion-dependent [2]. These authors, and later Garcin [10], also reported a "thalamic dystonic hand". The so-called rubral tremor [28, 29, 34–37] is a controversial entity, characterized by large-amplitude resting and action tremor associated with "wing-beating" tremor of the arm at around 3 Hz, and a kinetic cerebellar ataxia. Lesions along the dentato-rubro-thalamic cerebellar efferent pathway can cause this syndrome, which is reported in patients with multiple sclerosis, meso-diencephalic infarcts, tumours, abscesses or vascular malformations, but also in the postanoxic myoclonic syndrome [30, 32]. Stereotactic

surgery in the ventrolateral nucleus abolished this syndrome in a case of postanoxic intention myoclonus reported by Lhermitte [32]. Myoclonus [33], rhythmic hemiballism [21] or action-induced rhythmic dystonia [21, 26] have been described in lesions in or near the ventrolateral nucleus. Stereotactic surgery in this region has shown improvement in various dyskinesias [38, 39].

We have reported the cases of three patients with a thalamic infarct in the territory of the posterior choroidal artery [15–19] who suffered from a rubral-like tremor syndrome associated with a complex myoclonic hyperkinetic syndrome that could be further analysed on videotaping. One patient had a complex choreoathetotic myoclonic syndrome with total loss of control of the arm. The movements were spontaneous, markedly increasing on action with ataxo-ballistic-myoclonic features. Another patient presented with a more classical "rubral tremor" with ataxia, low-frequency, high-amplitude resting and action "tremor" with wing-beating of the arm, but myoclonic on action and posture, with a slightly dystonic posture of the fingers. The third patient had a dystonic "thalamic hand" syndrome with ataxia and rare action myoclonic jerky motions. All had presented with a mild and almost totally regressive corticospinal syndrome with normal or even increased uncontrollable strength; all had a sensory thalamic syndrome, and two of them presented with a delayed central painful syndrome of Déjerine-Roussy [1]. All had a superior homonymous quadrantanopia, but none had oculomotor abnormalities.

The most striking feature was that the syndrome was delayed, appearing months or years after the stroke. The reason for this delay, which is also found with the Déjerine-Roussy syndrome and posthemiplegic dystonia, is still obscure and may be due to late effects of reorganizing processes in the brain after deafferentation. All patients had an ischaemic lesion in the territory of the posterior cerebral artery with involvement of the posterior choroidal artery territory, and a decreased signal in the posterior thalamus, a region corresponding to the pulvinar nucleus, but no lesion was visualized in the lateral thalamic nuclei, even though the patients presented with a sensory thalamic syndrome (possibly due to involvement of the thalamic sensory radiations in the adjacent internal capsule), and a "rubral syndrome", with probable involvement of the cerebello-rubro-thalamic pathway. We did not encounter this syndrome in the acute phase of 54 patients with a thalamic infarct in the other topographical vascular territories of the thalamus [4].

The pulvinar nucleus is mainly connected to parietal areas and is not a part of the cerebello-efferent pathways to the ventrolateral nucleus of the thalamus, probably responsible for the "rubral syndrome" [27, 34–36]. Acuna et al. [40, 41] clearly showed in monkeys that the unit cell activity in the pulvinar nucleus was essentially correlated with the intention of movement, the projection and hand manipulation, with different thresholds according to the

direction of the movements [41]. This corresponds to the observation in our patients. The pulvinar nucleus, however, cannot be held responsible for either the hemisensory deficit or the painful Déjerine-Roussy syndrome present in our patients, nor for the cerebellar type of ataxia. We suspect the lesion to involve the fascicular cerebellar and sensory pathways within the thalamus before their entry in respectively the ventrolateral and ventro-posterolateral nuclei respectively. This may account for the delay in the appearance of both the sensory and motor syndrome, in presenting as a deafferentation syndrome, but the improvement and partial recovery of the corticospinal motor function through plastic reorganizations or reinnervation could also be necessary for these hyperkinetic behaviours to occur, since it is well established that lesions of the corticospinal tracts alleviate all hyperkinetic movements disorders.

A common characteristic for the three clinical pictures presented by our patients is the jerky myoclonic or ballistic aspect of the dystonic, choreoathetotic or tremulous syndromes, which we coined "jerky unsteady dystonic hand", because we thought this description best fits the polymorphous complexity of the motion. Each of the three patients described here presents a predominance of either myoclonus, dystonia, tremor or choreoathetotic

movements but always in a mixed complex presentation. The most striking feature was a clear-cut worsening of the signs by voluntary activity or intention, which makes this presentation unique to lesions in this region of the brain and may be due to the involvement of the pulvinar nucleus and the strategic crossing of proprioceptive, cerebellar, pyramidal and subthalamic pathways. The potential role of the underlying essential tremor in the first patient remains unclear. In any case, pure proprioceptive deafferentation is unlikely to be responsible for such a complex movement disorder, even though dystonic and ataxic presentations of parietal and thalamic lesions are already well known. The involvement of the territory of the posterior choroidal artery (Fig. 1) [26] seems to be a prerequisite for this syndrome to appear after a posterior cerebral infarct, because we did not find such a presentation in other thalamic infarcts [26]; this could also be a new delayed lacunar syndrome.

In conclusion, we suggest that a delayed complex hyperkinetic syndrome (choreoathetosis, dystonia, myoclonus and action tremor) together with upper quadrantanopia, hemisensory dysfunction, and transient or mild hemiparesis is evidence of an infarct in the territory of the posterior choroidal artery.

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