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Case report

Postencephalitic parkinsonism and selective involvement of substantia nigra in childhood

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Abstract

Parkinsonism is a rare complication of encephalitis in childhood. Association to an isolated involvement of substantia nigra is exceptional. Mechanisms of nigral cells neurotropism remain hypothetic. We report on three children presenting with postencephalitic parkinsonism and selective involvement of substantia nigra, with literature review and we discuss pathogenic mechanisms.

Keywords: Parkinsonism; Encephalitis; Childhood; MRI; Substantia nigra

1. Introduction

Secondary parkinsonism is rare in childhood. Etiologies are often related to neuroleptic intake or traumatic injuries [1]. Cerebral infection in childhood is an uncommon etiology of acquired parkinsonism [2–5]. We report on three children with postencephalitic parkinsonism and isolated involvement of substantia nigra with literature review and we discuss the pathogenic mechanisms.

2. Case 1

An 11-year-old Malian boy presented to our department with hypokinetic movement disorders. He had no family or personal medical history. Two months earlier, the child had a cerebral infection with fever (39 °C), altered vigilance, agitation and tonic–clonic seizures. He was admitted in a Malian hospital. Cerebral computerized tomography (CT) scan was normal. Electroencephalography and nerve conduction velocities were normal. Routine biological tests showed abnormalities: hemoglobin 10.7 g/dL (11–15 g/dL), platelets 25.000 e/mm^3 (150.000–450.000 e/mm^3), calcemia 2.08 mmol/L (2.2–2.6 mmol/L) and elevated liver enzymes: alanine amino transferase 410 IU/L (5–35 IU/L) and aspartate amino transferase 235 U/L (5–40 IU/L). Blood smear showed *Plasmodium falciparum* trophozoite. Cerebral malaria was diagnosed so the patient received quinine and intravenous steroids. Two days after the onset, the patient developed extrapyramidal syndrome with hypokinetic movements, anarthria, dysphagia and drooling. Cerebrospinal fluid (CSF) analysis was normal. The fever disappeared and improvement of consciousness was noticed after treatment, but extrapyramidal signs persisted. The patient was admitted in our department (2 months after the onset). Neurological examination showed parkinsonian features: akinetic

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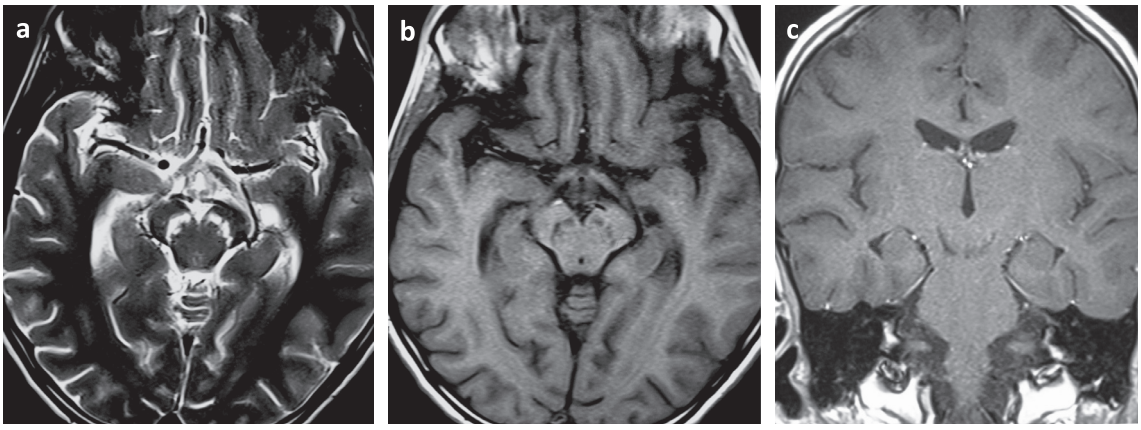

Fig. 1. Axial brain MRI (a and b) showing bilateral hypersignal of the substantia nigra on T2-weighted image (a) with decreased signal on T1-weighted image (b). No contrast enhancement was noticed on coronal T1 weighted image with gadolinium (c).

mutism, sialorrhea, generalized akinesia, intermittent resting tremor of upper limbs, dystonia in lower limbs and cogwheel rigidity prevailingly in upper limbs. He had also brisk tendon reflexes and plantor flexor. Brain MRI revealed increased signal of substantia nigra on T2 weighted images with decreased signal on T1 (Fig. 1). The patient received progressively 400 mg daily of levodopa with dramatic improvement within 10 days. Three months after continuous dopa therapy, fluctuations and biphasic dyskinesias were noticed. No improvement was obtained even after increasing the frequency of levodopa doses and association to amantadine. Control brain MRI showed persistent lesions of substantia nigra with gliosis.

3. Case 2

A 2-year-old female, presented with an acute onset of tremor and slow movements of the left upper limb. She had no family or personal medical history. The patient had encephalitic signs with fever (39 °C), headache, drowsiness, and vomiting. She developed motor symptoms 24 h after the onset of encephalitis. Neurological examination showed unilateral parkinsonism with intermittent rest tremor, bradykinesia and cogwheel rigidity in upper and lower left limbs. The patient was able to walk without aid but with mild difficulties. Blood cell count revealed leukocytosis 20.000 e/mm^3 (4000–10.000 e/mm^3) with lymphocytosis 15.000 e/mm^3 (1500–5000 e/mm^3). The other routine biological tests, chest X-rays and cerebral CT scan were normal. Urine tests for toxic substances or neuroleptic intake were negative. Brain MRI revealed isolated increased signal of the substantia nigra on T2 weighted images prevailingly on the right side (Fig. 2). Copper tests were normal. CSF analysis showed no abnormalities. Viral serologies (measles, Epstein–Barr virus, herpes simplex virus and human immunodeficiency virus) were negative on blood and CSF samples. The patient received acyclovir 500 mg/m^2 and 50 mg of levodopa daily. Parkinsonian features improved 7 days after receiving dopa therapy. No dopa-induced complications were noticed. Control brain MRI, 3 months after onset, showed partial resolution of the substantia nigra lesions. The patient had full

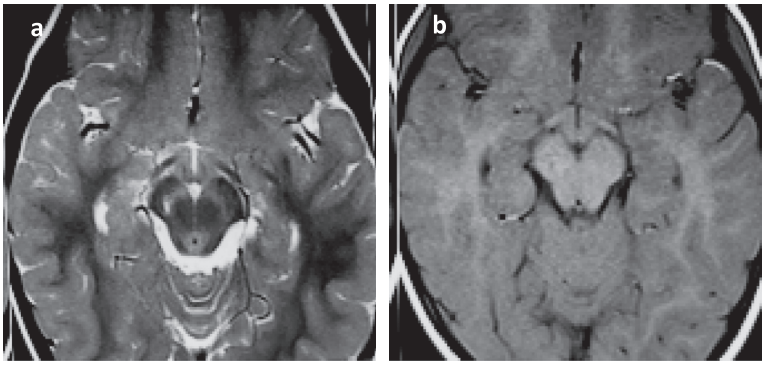

Fig. 2. Axial brain MRI (a and b) showing increased signal of the right substantia nigra on T2 weighted images (a) with normal signal without contrast enhancement on T1 weighted images with gadolinium (b).