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Primary CNS Lymphoma Presenting as a Choreic Movement Disorder Followed by Segmental Dystonia

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Summary: Clinical presentation of primary CNS lymphoma with an extrapyramidal movement disorder has not been recorded. A 66-year-old woman presented with chorea involving her left arm and subsequently developed right-sided segmental dystonia with prominent hemifacial dystonic spasms, milder torticollis and dystonia of the right arm. Investigations revealed primary CNS lymphoma with extensive involvement of the right-sided basal ganglia as well as lesions confined to the head of the left caudate nucleus and the corpus callosum. Chorea of her left arm subsided with progressing disease while remission of right-sided segmental dystonia was observed following radiotherapy of the brain. This patient's findings and a review of the literature suggest a possible relation between cranio-cervical dystonia and pathology affecting the head of the caudate nucleus. **Key Words:** CNS lymphoma—Segmental dystonia—Caudate nucleus.

Primary CNS lymphoma is a rare condition accounting for only 0.8–1.5% of intracranial tumors (1). Involvement of the basal ganglia is a common computed tomography (CT) and pathological finding in primary CNS lymphoma (2–5), but clinical presentation with signs of extrapyramidal motor dysfunction has not been recorded in large series of this type of intracranial neoplasm (2,4,6–8). We therefore report a case where the initial neurological manifestation consisted of focal choreic involuntary movements followed by contralateral segmental dystonia. The latter was dominated by sustained hemifacial contractions and correlated with a contralateral lesion affecting the head of the caudate nucleus.

CASE REPORT

A 66-year-old woman was well until May 1985 when she suffered from an episode of left-sided uveitis and papillitis, which resolved after a course of ste-

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roids. One year later involuntary flinging movements involving her left hand and arm appeared leading to the patient's admission to another hospital. The involuntary movements subsided with oral treatment with haloperidol (1 mg t.d.s.), but over the following weeks unsteadiness of gait developed together with mental slowing and lethargy. Haloperidol was discontinued 3 weeks prior to admission to this hospital, without reemergence of the left-side involuntary movements.

When first seen by the authors, the patient was drowsy but responsive to verbal commands and there was no dysphasia. Her face was intermittently distorted by frequent and prolonged bouts of dystonic spasms of the right side of the face (see Fig. 1) which occurred both spontaneously and when the patient attempted to speak. These bouts of facial spasms persisted from several minutes to 1 h. In addition, there was slight dystonic posturing of her head with retrocollis and torticollis to the right. Very occasionally minor dystonia of her right hand was also visible, but no other involuntary movements were present.

There was no papilledema, the tendon reflexes were symmetrically hyperactive, and plantar responses were extensor bilaterally. The patient was unable to walk unaided and tended to fall to the left. Cranial CT revealed multiple nodular contrast-enhanced lesions involving the right thalamus, the rostrum of the corpus callosum, and the head of the left caudate nucleus (Fig. 2). Magnetic resonance (MR) showed areas of increased signal intensity in the same brain areas and no abnormalities in the caudal brain stem. Cerebrospinal fluid protein was elevated to 101 mg/100 ml, but there was no pleocytosis or abnormal CSF cytology. Cerebrospinal fluid cultures remained sterile.

FIG. 1. Right-sided dystonic hemifacial spasm in patient.

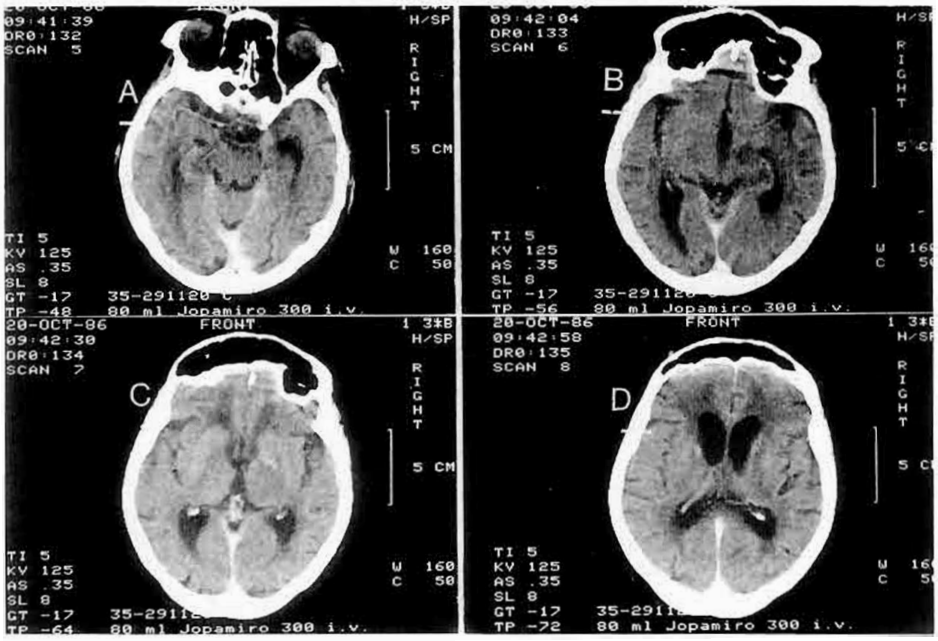

FIG. 3. A–D: Cranial CT scans of the patient following radiotherapy show almost complete remission of the tumor. A small contrast-enhancing lesion is still apparent in the head of the left caudate (D). Note evidence of radiation-induced leukoencephalopathy with ventricular enlargement and periventricular lucency of white matter.

reflected early neoplastic invasion of the contralateral subthalamic nucleus, the structure most consistently involved in symptomatic hemichorea-hemiballism (9,10). When the patient was first examined by the authors, these movements had subsided, and there was extensive infiltration of the right-sided basal ganglia by tumor masses (see Fig. 2).

She had by then developed right-sided segmental dystonia consisting of prominent hemifacial dystonic spasm and less pronounced torticollis to the right plus occasional dystonic hand movements. Although there were multiple subcortical lesions in this patient, her segmental dystonia correlated with structural damage to the head of the left caudate nucleus and this was the only left-sided basal ganglia structure affected by tumor. Similar lesions of the caudate nucleus on CT or at autopsy have been reported in the literature (11–13). Of a total of five reported cases with a lesion confined to the head of the caudate, two had torticollis with deviation of the chin to the contralateral side (11), two contralateral hemidystonia (11,13), and one dystonia affecting the contralateral arm (12). This patient was similar in that there was also torticollis and some minor dystonia of the contralateral hand. However, in none of the cases reported in the literature has contralateral hemifacial dystonia been mentioned as an associated feature of caudate pathology. Furthermore, hemifacial dystonia, spontaneous or action induced, is not mentioned in reports of large series of patients with focal dystonia following

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