MoyaMoya disease is a rare cerebrovascular arteriopathy that is characterized by progressive narrowing of the supraclinoid internal carotid artery. Collateral blood flow to areas of the brain distal to the narrowed vessels is supplied by dilated vessels, which originate at the base of the brain. On arteriography these dilated collateral vessels produce the characteristic hazy or cloudlike appearance. Common clinical presentations of this disease in children are ischemic stroke, recurrent TIAs, headache, and seizure activity. An uncommon presentation is movement disorder, which has been described to be present in 3%–6% of patients in large series. 

Chorea is characterized as brief, irregular, and involuntary movements that appear to move from 1 muscle group to another. There have been isolated case reports of moyamoya disease patients with chorea as part of their clinical presentation. It has been presumed that this clinical presentation is due to ischemia in the basal ganglia. In some of these reported cases, the chorea resolved after surgical revascularization.

In the present study, our aim was to characterize those patients with moyamoya disease who presented with chorea in terms of clinical course, radiographic findings, and response to surgical revascularization. In our analysis, we had hypothesized that the presence of hypertrophied collateral vessels in the basal ganglia provides the pathological substrate for the chorea. This series is the largest single series of patients with moyamoya disease and chorea as part of their clinical presentation.

Chorea in the clinical presentation of moyamoya disease: results of surgical revascularization and a proposed clinicopathological correlation

Clinical article

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Object. Chorea is a movement disorder characterized by brief, irregular, involuntary contractions that appear to flow from 1 muscle to another. There are a limited number of reports in the literature that have linked moyamoya disease and chorea. The authors describe their experience in treating moyamoya disease in patients in whom chorea developed as part of the clinical presentation.

Methods. The authors conducted a retrospective review of a consecutive series of 316 children who underwent pial synangiosis revascularization for moyamoya disease at the Boston Children’s Hospital.

Results. Of 316 surgically treated patients with moyamoya disease, 10 (3.2%; 6 boys and 4 girls) had chorea as a part of their presentation. The average age at surgical treatment was 9.9 years (range 3.8–17.9 years). All patients had evidence of hypertrophied lenticulostriate collateral vessels through the basal ganglia on preoperative angiography and/or MRI on affected sides. Two patients had cystic lesions in the basal ganglia. Nine patients underwent bilateral craniotomies for pial synangiosis, and 1 patient underwent a single craniotomy for unilateral disease. Follow-up was available in 9 patients (average 50.1 months). The mean duration of chorea was 1.36 years (range 2 days to 4 years), with resolution of symptoms in all patients. One patient developed chorea 3 years after surgical treatment, 4 patients had transient chorea that resolved prior to surgery, and 5 patients experienced resolution of the chorea after surgery (average 13 months).

Conclusions. The authors describe children with moyamoya disease and chorea as part of their clinical presentation. The data suggest that involvement of the basal ganglia by the hypertrophied collateral vessels contributes to the development of chorea, which can wax or wane depending on disease stage or involution of the vessels after revascularization surgery. In most patients, however, the chorea improves or disappears about 1 year after presentation.

Key Words • moyamoya disease • chorea • dyskinesia • movement disorder • pial synangiosis • surgical revascularization • vascular disorders

Abbreviation used in this paper: TIA = transient ischemic attack.

Chorea is characterized as brief, irregular, and involuntary movements that appear to move from 1 muscle group to another. There have been isolated case reports of moyamoya disease patients with chorea as part of their clinical presentation. It has been presumed that this clinical presentation is due to ischemia in the basal ganglia. In some of these reported cases, the chorea resolved after surgical revascularization.

In the present study, our aim was to characterize those patients with moyamoya disease who presented with chorea in terms of clinical course, radiographic findings, and response to surgical revascularization. In our analysis, we had hypothesized that the presence of hypertrophied collateral vessels in the basal ganglia provides the pathological substrate for the chorea. This series is the largest single series of patients with moyamoya disease and chorea as part of their clinical presentation.
Methods

We performed a review of the clinical records of a consecutive series of 316 patients with moyamoya disease in whom surgical revascularization was performed by the senior author (R.M.S.) at the Boston Children’s Hospital between 1985 and 2008. From this series, we identified all patients in whom chorea was part of their clinical presentation. We determined patient age; sex; presenting symptoms; radiographic features, including arteriographic staging and MRI details; clinical course; descriptions of the chorea; surgical details; follow-up course; and follow-up duration.

From the moyamoya disease series of 316 patients, we noted 10 cases (3.2%) in which chorea was part of the patients’ clinical presentations (Table 1). There were 6 boys and 4 girls whose mean age at surgery was 9.9 years (range 3.8–17.9 years).

All 10 patients underwent preoperative MRI. Nine of 10 patients underwent preoperative angiography, and 8 of the studies were available for review at the time of this study. Arteriograms were staged preoperatively and postoperatively using the Suzuki grading system.22 Postoperatively synangiosis collaterals were graded using the Matsushima grading system.10 From 1992 to 2008, 9 patients underwent bilateral craniotomies and 1 patient underwent a unilateral (left-sided) craniotomy for pial synangiosis, a standardized procedure, the details of which have been previously described.1,17,19 Six patients had both operations sequentially on the same day under 1 anesthesia. Follow-up data (mean duration 50.1 months) were available in 9 of 10 patients. Clinical information was obtained from office visitations and communication with referring physicians, patients, and families.

Results

Clinical Presentation

The majority of patients, 8 of 10, initially presented with ischemic symptoms (Table 2). Five patients presented with primarily TIAs. Four suffered completed strokes (neurological deficit lasting more than 24 hours) while 1 presented with both TIAs and stroke. One patient presented with convulsive seizures without ischemic symptoms. Another patient presented without a history of stroke but had rapidly progressive right-sided weakness and development of a hemichorea syndrome. Two patients presented with cognitive decline and developmental delay at 8 and 5 years of age.

All patients exhibited chorea at some point during their clinical course (Table 3). The diagnosis was confirmed by a neurologist in 5 of the 10 cases. Other children were diagnosed by other treating physicians who identified the characteristic involuntary movements. In 9 of 10 patients, the onset of chorea occurred prior to surgical revascularization for the moyamoya disease. One patient experienced new-onset chorea 3 years after surgical revascularization, which lasted for 2 days and resolved on its own. Four of the patients had bilateral manifestation of the chorea, whereas 6 had unilateral manifestation. The mean duration (± SD) of chorea was 1.35 ± 1.46 years (range 2 days to 4 years).

Chorea was the initial presentation of moyamoya disease in 6 of the 10 patients. Three patients were initially diagnosed with Sydenham chorea after a presumed infection with Group A β-hemolytic streptococci and acute rheumatic fever. None of these patients, however, met Jones criteria for rheumatic fever. One patient, a 9-year-old boy, developed uncontrollable flinging of his right arm and twitching of the mouth. He was treated with penicillin with the presumed diagnosis of Sydenham chorea. His symptoms recurred, and his gait and handwriting gradually became impaired. The next patient, an 11-year-old boy, presented with a fever and involuntary movements and coordination problems on the right, which was initially thought to be Sydenham chorea. The third patient, an 8-year-old boy, developed bilateral choreiform movements 11 days after onset of pharyngitis. He was found to also have a fever and heart murmur, which further increased the suspicion of rheumatic fever.

Initial Radiographic Findings

All 10 patients underwent imaging with MRI at the time of presentation and initial diagnosis (Table 4). The common finding in all 10 patients was the presence of hypertrophied collateral vessels within the basal ganglia (Fig. 1 left). Two patients had cystic lesions in the basal ganglia that possibly represented old hemorrhages or strokes (Fig. 1 right). Other MRI findings included previous infarcts in 3 patients and old hemorrhages in 2 patients.

Nine patients underwent preoperative cerebral angiography and 8 studies were available for review at the time of this study. One patient had only preoperative CT scans and MRI/MRA images prior to surgery. The common finding on all 8 angiograms was an extensive network of collateral vessels coursing through the basal

TABLE 1: Summary of patient demographics

<table>
<thead>
<tr>
<th>Factor</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>no. of patients</td>
<td>10</td>
</tr>
<tr>
<td>sex</td>
<td></td>
</tr>
<tr>
<td>male</td>
<td>6</td>
</tr>
<tr>
<td>female</td>
<td>4</td>
</tr>
<tr>
<td>age (yrs)</td>
<td></td>
</tr>
<tr>
<td>mean</td>
<td>9.9</td>
</tr>
<tr>
<td>range</td>
<td>3.8–17.9</td>
</tr>
</tbody>
</table>

TABLE 2: Clinical presentation of 10 patients with moyamoya disease

<table>
<thead>
<tr>
<th>Presentation</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>ischemic symptoms</td>
<td>8</td>
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<tr>
<td>TIA</td>
<td>5</td>
</tr>
<tr>
<td>stroke</td>
<td>4</td>
</tr>
<tr>
<td>seizure</td>
<td>1</td>
</tr>
<tr>
<td>cognitive decline/developmental delay</td>
<td>2</td>
</tr>
<tr>
<td>total</td>
<td>10</td>
</tr>
</tbody>
</table>
Chorea and moyamoya disease

These vessels were typically supplied by both the anterior circulation (from the internal carotid artery apex and posterior communicating artery) and the posterior circulation (predominantly from the basilar apex). Suzuki grading was available for 8 angiograms, or 16 hemispheres. All hemispheres were Suzuki Grade 3 or 4; 10 hemispheres were Grade 3; and 6 were Grade 4. The average Suzuki grade was 3.4, which is comparable to many those reported in major pediatric moyamoya series, including our data from Boston Children’s Hospital.

Surgical Treatment and Perioperative Course

All patients underwent indirect surgical revascularization by craniotomies for pial synangiosis (Table 5); thus, there were a total of 19 operations in the 10 patients (9 bilateral and 1 unilateral). Six patients underwent both operations on the same day under 1 anesthesia, whereas the other 4 underwent staged (or, in the sole case of unilateral disease, a single) unilateral operations. Eighteen of the 19 operations were preceded by standardized admissions to the hospital 1 day prior for preoperative hydration, as has been previously described.15,20 The average hospital length of stay, including the preoperative hydration days, was 4.7 days.

Immediate postoperative ischemic events occurred after 2 of 20 operations—a TIA and a stroke, both in the same patient. The patient was a 17-year-old girl who originally presented with headaches, slurred speech, and involuntary movements on the right side of the body. She underwent a left-sided craniotomy for pial synangiosis, which was followed by transient right-sided weakness considered a TIA, with MRI showing only operative changes. She returned 6 weeks later for pial synangiosis on the right side. After this operation, she developed left facial droop, hemiparesis, and neglect with signs of a temporoparietal stroke on the right side. She recovered from both of these events prior to discharge to home at 6 and 10 days, respectively, after admission.

Another patient, a 5-year-old boy, presented with seizures at initial diagnosis, as mentioned. This child continued to experience seizures during the follow-up period after revascularization.

Outcome After Hospital Discharge

Follow-up was available in 9 of 10 patients during a mean period of 50.1 months (range 13 months to 11.8 years) (Table 6). One patient was lost to follow-up. There were no incidents of late strokes after surgical revascularization. Magnetic resonance images obtained at least 1 year postoperatively were available in these 9 patients and demonstrated no late infarcts. Angiography was typically performed approximately 1 year after surgical revascularization was complete. Eight of the patients had follow-up angiograms that were available for review. In these studies, all 15 hemispheres (7 bilateral, 1 unilateral) displayed development of new surgical collateral vessels. Matsushima grading distribution was as follows: Grade A, 8 hemispheres (53%); Grade B, 6 hemispheres (40%); and Grade C, 1 hemisphere (7%).

<table>
<thead>
<tr>
<th>TABLE 3: Chorea manifestations in 10 patients with moyamoya disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chorea Manifestation</td>
</tr>
<tr>
<td>----------------------</td>
</tr>
<tr>
<td>timing</td>
</tr>
<tr>
<td>before surgery</td>
</tr>
<tr>
<td>after surgery</td>
</tr>
<tr>
<td>sidedness</td>
</tr>
<tr>
<td>unilat</td>
</tr>
<tr>
<td>bilat</td>
</tr>
<tr>
<td>duration</td>
</tr>
<tr>
<td>mean ± SD (yrs)</td>
</tr>
<tr>
<td>range</td>
</tr>
<tr>
<td>other</td>
</tr>
<tr>
<td>chorea as initial sign of moyamoya syndrome</td>
</tr>
<tr>
<td>diagnosed w/ Sydenham chorea</td>
</tr>
<tr>
<td>chorea noted postop</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>TABLE 4: Radiographic findings</th>
</tr>
</thead>
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<td>Imaging Mode/Finding</td>
</tr>
<tr>
<td>----------------------</td>
</tr>
<tr>
<td>MRI*</td>
</tr>
<tr>
<td>collat vessels in basal ganglia</td>
</tr>
<tr>
<td>cystic lesions in basal ganglia</td>
</tr>
<tr>
<td>previous infarcts</td>
</tr>
<tr>
<td>hemorrhage</td>
</tr>
<tr>
<td>angiography</td>
</tr>
<tr>
<td>collat vessels in basal ganglia*</td>
</tr>
<tr>
<td>Suzuki grade†</td>
</tr>
<tr>
<td>3</td>
</tr>
<tr>
<td>4</td>
</tr>
<tr>
<td>5</td>
</tr>
<tr>
<td>mean</td>
</tr>
</tbody>
</table>

* Values reflect the number of patients.
† Values reflect the number (%) of hemispheres.

Fig. 1. Left: Axial T2-weighted MR image demonstrating the common finding of flow voids in the basal ganglia that represent lenticulostriate collateral vessels (black arrows). Right: Axial T2-weighted MR image revealing bilateral cystic lesions in the basal ganglia.
Clinical Course of Chorea and Resolution of Basal Ganglia Collateral Vessels

In all 10 patients resolution of chorea was demonstrated during the study period. These patients can be classified into 3 groups according to the timing of onset and resolution with respect to surgical revascularization (Table 7): 4 patients experienced onset and resolution of the chorea prior to surgical revascularization; 5 experienced chorea resolution after surgical revascularization; and 1 patient, discussed above, suffered chorea after surgical revascularization. This patient was 8 years old when she underwent bilateral craniotomies for pial synangiosis that were uneventful. Postoperative angiography demonstrated development of surgical collateral vessels and persistence of a large thalamoperforating artery that was present on preoperative studies (Fig. 2). Three years after her operations, she developed involuntary movements of her right shoulder, arm, hand, leg, and foot consistent with chorea. The movements resolved after 2 days in the hospital along with an increase in her aspirin dosage.

Five patients had chorea present at the time of surgery. Of these patients, all experienced resolution of the chorea postoperatively, mostly within 1 year’s time (average 13 months, range 11–17.4 months)

Although we are reporting a pediatric case series, we did have 1 adult among our small series of patients with moyamoya disease who presented with chorea. This patient was a 29-year-old woman who developed uncontrolable flicking movements of her arms, hands, and neck 5 months before undergoing bilateral craniotomies for pial synangiosis. By 11 months after her operations, the chorea had resolved. Compared with the preoperative angiogram, angiography performed 13 months after revascularization demonstrated marked reduction in the collateral vessels from the distal internal carotid artery (Fig. 3A and B). The external carotid artery angiogram showed excellent revascularization with collaterals from the superficial temporal artery on both sides. Magnetic resonance imaging performed 4 years after revascularization demonstrated complete resolution of flow voids within the basal ganglia compared with a preoperative study (Fig. 3C and D). The patient’s neurological examination status at this time was normal except for some mild difficulty with rapid alternating movements on the left side.

Discussion

Chorea is one of the less common presenting signs of moyamoya disease. Its incidence ranges from 3% to 6% in large series. Because the movement disorder may be the initial presenting sign or the primary symptom in the clinical course of moyamoya disease, authors have supported the idea that moyamoya disease should be in the differential diagnosis of chorea.
differential diagnosis of persistent or paroxysmal chorea. The pathology is localized to the basal ganglia, as chorea is a known manifestation of ischemia in the basal ganglia. In moyamoya disease, the prominent collateral vessels in the basal ganglia are likely candidates for the pathological substrate in this manifestation of the disease.

Others have reported the association of chorea with moyamoya disease, sometimes as part of the initial manifestation of moyamoya prior to TIAs or strokes (Table 8). In our series, 6 (60%) of the 10 patients displayed chorea as the initial presenting sign of their disease. Notably, 3 of these patients were given the diagnosis of Sydenham chorea with suspicion of rheumatic fever despite not meeting the Jones criteria. One of these patients was treated with penicillin and yet had progressed to worsening neurological function. Gonzalez-Alegre et al. described 2 cases in which young females with unrecognized moyamoya disease developed postpartum strokes years after their initial presentation with chorea. Thus, a timely diagnosis of moyamoya disease despite atypical presenting signs is of paramount importance because of the potential for neurological worsening if treatment is delayed.

Some authors have proposed that ischemia in the basal ganglia–thalamocortical circuits is the pathophysiological mechanism of chorea associated with moyamoya syndrome. Magnetic resonance imaging in our series demonstrated prominent hypertrophied collateral vessels within the basal ganglia in all 10 patients. While the development of basal ganglia collaterals is common to many other patients with moyamoya disease, it is our impression (admittedly subjective and difficult to measure) that the collaterals in this group are substantially more pronounced than in other patients (as evidenced in Fig. 3). In addition, we also have observed evidence of infarcts in the basal ganglia of some of these patients (Fig. 1), further bolstering the claim that this region of the brain contributes to the pathological substrate for the clinical manifestation of chorea. Although the small sample size of this subset of moyamoya disease patients does not allow a statistically supported comparison with the larger group of patients with surgical revascularization, the uniform finding of hypertrophied collateral vessels in the basal ganglia is a notable, albeit not unique, feature of this group. It has been suggested that there is early ischemia in the basal ganglia, which may lead to early presentation with chorea prior to the development of a fixed neurological deficit. This mechanism would apply to the clinical presentation of the patients in our series who presented with chorea as the initial manifestation of their moyamoya disease. Early chorea may result from ischemia in the basal ganglia, and the consistent prominence of basal ganglia collateral vessels can be viewed as an angiogenetic response to the ischemia.

On the other hand, one may also view the presence of these collateral vessels as a physical disruption of normal basal ganglia physiology and, thus, the direct cause of chorea. We cannot determine completely whether these collateral vessels simply serve as a sign of underlying ischemia or whether they cause physical disruption of normal basal ganglia signaling; nevertheless, we found that the presence of these hypertrophied basal ganglia collateral vessels was universal in this series of patients with chorea. The fact that some of the children had prominent collateral vessels bilaterally but only displayed unilateral symptoms suggests that other contributing factors not detectable on imaging may contribute to the manifestation of choreiform movements. Our illustration of the 29-year-old woman who presented with imaging-documented chorea and prominent basal ganglia collateral vessels provides a clinicopathological correlation between these abnormal vessels and her clinical presentation. In her case, the chorea resolved about 1 year after surgical revascularization. With her clinical improvement, radiographic studies revealed marked reduction in the basal ganglia collateral vessels that were previously extensive. With surgical revascularization, there was regression of collateral vessels, presumably from improved perfusion. In this patient’s case, it is possible that the chorea may

<table>
<thead>
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<th>Onset &amp; Resolution</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prior to Revascularization</td>
<td>4 (40)</td>
</tr>
<tr>
<td>Prior to Revascularization &amp; Resolution Afterward</td>
<td>5 (50)</td>
</tr>
<tr>
<td>Resolution After Revascularization</td>
<td>1 (10)</td>
</tr>
</tbody>
</table>

Fig. 3. A–D: Images obtained in a patient who developed right-sided chorea prior to surgical revascularization. A cerebral angiogram with anterior circulation injection demonstrated a prominent network of collateral vessels in the basal ganglia (A). An MR image revealed corresponding flow voids within the basal ganglia (C). The patient underwent bilateral craniotomies for pial synangiosis, and her movements resolved within 11 months. Follow-up cerebral angiography 13 months after surgical revascularization revealed marked reduction in the basal ganglia collateral vessels (B). An MR image 4 years after surgical revascularization showed complete resolution of the signal voids in the basal ganglia (D).
have resolved as a result of restored perfusion to the previously ischemic basal ganglia or as a result of restored physiological basal ganglia function after regression of the large blood vessels within.

This case supports the notion that chorea associated with moyamoya disease results from vascular changes in the basal ganglia and that reduction in basal ganglia collateral vessels may correlate with resolution of symptoms. All patients in this series experienced resolution of the chorea, sometimes even before surgical revascularization, which indicates its self-limiting properties. As the natural progression of moyamoya disease involves eventual resolution of these moyamoya collateral vessels, the timing of resolution may dictate the clinical course of chorea.

Conclusions

Chorea is a rare manifestation of moyamoya disease. In all children with a new presentation of chorea, moyamoya disease should be considered in the differential diagnosis, which warrants MRA and/or angiography. Early recognition of moyamoya disease is of paramount importance due to the risk of stroke and fixed neurological deficit if the disease is left untreated. The characteristic radiographic appearance frequently consists of hypertrophied collateral vessels in the basal ganglia to a greater degree than other moyamoya patients, either with or without infarct in that location. The presence of these collateral vessels in the basal ganglia supports the hypothesis that they play a role in the pathophysiology of chorea in patients with moyamoya disease. Importantly, the movement disorder tends to be self-limiting in this population, and its resolution may correlate with radiographic response after surgical revascularization.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Scott. Acquisition of data: Ahn. Analysis and interpretation of data: Ahn, Robertson. Drafting the article: Ahn. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Smith. Study supervision: Smith, Scott.

References

6. Handa H, Yonekawa Y, Gotoh Y, Hoshimaru M, Komori Y, Minato K: Filing of 1500 cases of the occlusive disease of the circle of Willis: appendix, a follow-up study of 5 years or more on 200 cases, in Handa H (ed): Annual Report of

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TABLE 8: Previously reported cases of chorea in association with moyamoya

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs) at Onset/Sex</th>
<th>Characteristics</th>
<th>Abnormal Signal on Imaging</th>
<th>Outcome</th>
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<tr>
<td>Watanabe et al., 1990</td>
<td>3, F bilat</td>
<td>bilat caudate, periventricular</td>
<td>resolved</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3, M bilat</td>
<td>lt caudate, bilat frontal</td>
<td>resolved</td>
<td></td>
</tr>
<tr>
<td></td>
<td>8, F bilat</td>
<td>bilat frontal &amp; parietooccipital, flow voids in basal ganglia</td>
<td>resolved</td>
<td></td>
</tr>
<tr>
<td>Pavlakis et al., 1991</td>
<td>12, M bilat</td>
<td>rt MCA subcortical, rt basal ganglia</td>
<td>improved</td>
<td></td>
</tr>
<tr>
<td>Takanashi et al., 1993</td>
<td>11, F bilat</td>
<td>flow voids in basal ganglia, thalamus &amp; rt periventricular infarct</td>
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<td></td>
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<tr>
<td>Pelletier et al., 1997</td>
<td>17, F unilat</td>
<td>bilat frontal</td>
<td>resolved</td>
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<tr>
<td></td>
<td>17, F unilat</td>
<td>lt centrum semiomvale</td>
<td>resolved</td>
<td></td>
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<tr>
<td>Han et al., 2000</td>
<td>29, F unilat</td>
<td>bilat frontal, subcortical</td>
<td>improved</td>
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<td>Lyoo et al., 2000</td>
<td>22, F bilat</td>
<td>rt putamen</td>
<td>lost to follow-up</td>
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<td>bilat periventricular, centrum semiomvale</td>
<td>improved</td>
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<tr>
<td></td>
<td>9, F unilat</td>
<td>basal ganglia, corona radiata, centrum semiomvale</td>
<td>not specified</td>
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</tr>
<tr>
<td>Shanahan et al., 2001</td>
<td>18, F unilat</td>
<td>basal ganglia flow voids</td>
<td>improved</td>
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<tr>
<td>Hong et al., 2002</td>
<td>20, F unilat</td>
<td>lt centrum semiomvale; SPECT w/ lt basal ganglia perfusion defect</td>
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<td>Gonzalez-Alegre et al., 2003</td>
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<td>periventricular</td>
<td>resolved</td>
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<td></td>
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<td>head of caudate</td>
<td>resolved</td>
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<td>Zheng et al., 2006</td>
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<td>Pandey et al., 2010</td>
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<td>bilat watershed infarcts</td>
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<tr>
<td></td>
<td>17, F unilat</td>
<td>bilat basal ganglia &amp; frontal infarcts</td>
<td>resolved</td>
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</tbody>
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* MCA = middle cerebral artery.
Chorea and moyamoya disease


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