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Original Article

Brainstem Strokes in Children: An 11-Year Series From a Tertiary Pediatric Center

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ABSTRACT

METHODS: Potential clinical barriers to making a timely diagnosis of pediatric brainstem stroke and pitfalls of noninvasive vascular imaging are presented. **METHODS:** An institutional review board–approved institutional database query from 2001-2012 yielded 15 patients with brainstem strokes. Medical records were reviewed for symptoms, stroke severity using the Pediatric National Institutes of Health Stroke Scale, and outcomes using the Pediatric Stroke Outcome Measure. Magnetic resonance angiography was compared with digital subtraction angiography. **RESULTS:** There were 10 boys and five girls; 9 months to 17 years of age (mean 7.83 years). Symptoms were headaches (eight); visual problems (eight), seizure-like activity (seven), motor deficits (six), and decreased level of consciousness in four. Time since last seen well was 12 hours to 5 days. Pediatric National Institutes of Health Stroke Scale was 1-34; <10 in eight; 3 in 1, 10-20 in two, and >20 in four. Strokes were pontine in 13/15 and involved >50% of the pons in six and <50% in seven; 2/15 had medullary strokes. Magnetic resonance angiography showed basilar artery occlusion in 8/13 patients and vertebral artery dissection in two. Digital subtraction angiography done within 9-36 hours of magnetic resonance angiography in 10/15 patients confirmed the basilar artery occlusion seen by magnetic resonance angiography and showed vertebral artery dissection in four patients. Patients were systemically anticoagulated without hemorrhagic complications. One patient died. Pediatric Stroke Outcome Measures at 2-36 months is 0-5.0/10 (mean 1.25). **CONCLUSIONS:** Vague symptoms contributed to delays in diagnosis. Magnetic resonance angiography was equivalent to digital subtraction angiography for basilar artery occlusion but not for vertebral artery dissection. Even with basilar artery occlusion and high stroke scales, outcome was good when systemic anticoagulation was started promptly.

Keywords: pediatric, brainstem stroke, magnetic resonance angiography (MRA), outcome, arterial dissection, digital subtraction angiography

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Introduction

Compared with adults, pediatric ischemic strokes are uncommon, with a frequency of 1.8-3.3/100,000.[1-4] Among pediatric strokes, strokes involving the brainstem are rare, accounting for <8% of childhood strokes.[1-3] There are no widely accepted guidelines for vascular imaging in acute pediatric strokes although magnetic resonance angiography (MRA) is widely used.[4,5] The optimal medical management for pediatric stroke resulting from large artery disease is not clear.[1,2,6-8] We reviewed a decade-long experience with pediatric brainstem strokes examining issues impacting the timely diagnosis, practice patterns with respect to imaging and accuracy of noninvasive imaging at detecting vertebrobasilar pathology in children, and outcomes in a population treated with systemic anticoagulation.

Materials and Methods

This was retrospective review of patients with brainstem strokes seen at a single tertiary referral hospital over 11 years ending March 2013

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identified by an institutional review board–approved analysis of an institutional database. Hemorrhagic strokes and brainstem strokes associated with anterior circulation infarcts were excluded.

A board-certified pediatric neurologist (M.M.D.) with special expertise in pediatric stroke had either evaluated the patients at presentation or reviewed medical records for patient demographics, clinical presentation, time since last seen well, risk factors, and Pediatric National Institutes of Health Stroke Scale (PedNIHSS).[9] The PedNIHSS retains the examination items and scoring ranges of the NIHSS and can be retrospectively scored from medical records with a high degree of reliability and validity.[1,9] Laboratory investigations were per our institutional pediatric stroke guidelines. Neurological status at follow-up was characterized using the Pediatric Stroke Outcome measures (PSOM),[10] which provides objective assessment of right sensorimotor, left sensorimotor, language expression, language reception, and cognitive/behavioral in children taking into account expected age-related abilities. The PSOM total score (0-10) is the sum of these five subscale scores. A PSOM total score <0.5 indicates normal or insignificant deficit, 1-1.5 indicates a moderate deficit, and ≥ 2 indicates a severe deficit in at least one subscale.[10] PSOM score of 2 within a domain is equivalent to an adult modified Rankin score of 2 (e.g., unable to perform all previous activities).

Imaging

Magnetic resonance imaging was done at 1.5T and included three-dimensional time-of-flight MRA through the head without contrast; 11 patients also had MRA of the neck. Digital subtraction angiography (DSA) was done 9-36 hours after MRA and included selective bilateral vertebral artery injections with frontal and lateral projections from the origin of the vertebral artery to the vertebral-basilar confluence.

Experienced neuroradiologists reviewed magnetic resonance imaging and MRA for location(s) of the strokes and arterial abnormalities and DSA and reached consensus blinded to the clinical status of the patients. MRAs at presentation were graded as being of diagnostic quality or inadequate with respect to visualization of the vertebrobasilar system and analyzed for patency and absence of structural abnormalities of the vertebral-basilar system. Findings on MRA were compared with those seen by DSA.

Results

Clinical

Sixteen children were identified during this interval. One patient was excluded: a 5 year old with clival osteomyelitis and septic cavernous sinus thrombophlebitis complicated by a pontine infarct. The remaining 15 patients were 9 months to 17 years of age (mean 7.83 years); 10 boys and five girls (Table 1). Risk factors included recent trauma in seven; three were football related. One patient had neck cellulitis, one was on oral contraceptives, and one patient had known congestive cardiomyopathy and was poorly compliant with anticoagulation. The other six patients had no known risk factors although one was subsequently found to have a cardiomyopathy and one a patent foramen ovale. No patient had a cervical spine fracture, connective tissue disorder, or had undergone chiropractic manipulation; risk factors are indicated in Table 1.

The most common presenting symptom was headache in eight patients with onset 12-48 hours before diagnosis. Visual problems including gaze preference, nystagmus, and diplopia were seen in eight patients; five also had unilateral hemiparesis and patient was densely quadriparetic. Seven patients had intermittent rhythmic movements at presentation initially attributed to seizures. Depressed level of consciousness seen in four patients ranged from somnolent but arousable to comatose and on ventilatory support in one. Time elapsed from last seen well to diagnosis was 12 hours to 5 days.

Maximal PedNIHSS was 1-34 (mean 17.38) at presentation and had worsened from 8 to 15 in one patient and 22 to 34 in another. All patients were loaded with 75-80 units heparin per kilogram of body weight at diagnosis of brainstem stroke and maintained on heparin until conversion to low-molecular-weight heparin and/or low-dose aspirin without hemorrhagic complications. No patient received intravenous tissue plasminogen activator.

Imaging

Magnetic resonance imaging showed the brainstem stroke involved the pons in 13 patients and the medulla in two. Pontine infarcts affected $<50\%$ of the brainstem in six and $\geq 50\%$ in seven. The medullary strokes were $<50\%$ of the width of the medulla in both patients and involved the inferior cerebellar peduncle and cerebellar flocculus in one. Of the 13 pontine strokes, four were limited to the pons, six were associated with other posterior circulation strokes, and three involved the pons and pontomesencephalic junction. Of the six patients with multifocal posterior circulation strokes, four had infarcts of different ages.

The pontine strokes were associated with basilar artery occlusion by MRA in 8/13 patients (Table 2) and a normal basilar artery in five. Pontine strokes associated with basilar artery occlusion were $\geq 50\%$ of the transverse diameter of the pons in six patients and $<50\%$ in two. MRA of the neck was acquired in 11 of 15 patients; there was adequate visualization of the extra dural segments of the vertebral arteries in seven and inadequate visualization in four (Figs 1 and 2). The V3 segment was most problematic by MRA, as were hypoplastic vertebral arteries (Figs 2 and 3). MRA missed two of four vertebral artery dissections seen by DSA. Medullary strokes were associated with a hypoplastic vertebral artery in one and thrombus within the ipsilateral distal vertebral artery in one.

DSA was not done in five patients because of withdrawal of support in one, cardiomyopathy in two, and unequivocally normal MRA in two. DSA confirmed basilar artery occlusion seen by MRA in seven of eight patients and was not done in one of eight. The vertebral arteries were normal by DSA in three patients with basilar artery occlusion. DSA showed vertebral artery dissection in four patients with basilar artery occlusion. Two patients each had posterior circulation emboli and congenital unilateral vertebral artery hypoplasia by DSA. One patient, a 17-year-old boy, underwent endovascular intervention about 16 hours after the first onset of symptoms after rapid neurological deterioration to “locked-in” state over several hours despite adequate systemic anticoagulation. Clot extraction from the basilar artery was done with the 0.032-inch Penumbra thromboaspiration system (Penumbra Inc., Alameda, California) and a total of 4 mg of intra-arterial tissue plasminogen activator. There were no procedural or hemorrhagic complications.

Follow-up magnetic resonance imaging ranging from 2 to 22 months is available in nine patients and showed encephalomalacia and gliosis corresponding to regions of restricted diffusion in the brainstem (Fig 3). Seven patients