An unusual presentation of pheochromocytoma

A 16-year-old boy was noted to have an irregular heart rhythm during a screening examination. He had a 6-year history of night sweats. On examination, frequent ectopy was audible, and he had a resting blood pressure of 175/106 mm Hg. He was notably diaphoretic. The heart rate was 140 beats/min, with a normal S1 and physiologically split S2; a II/VI holosystolic murmur was detected at the left lower sternal border radiating to the apex, and a soft early diastolic murmur was detected at the left upper sternal border. An electrocardiogram with rhythm strip (Figure, A) and an echocardiogram were performed; the latter demonstrated mitral and aortic regurgitation and left ventricular hypertrophy. Renal ultrasonography revealed a large mass in the left kidney. Analysis of the urine showed an active sediment with red cells and red cell casts. Urine metanephrine levels were markedly elevated. Abdominal magnetic resonance imaging (Figure, B) confirmed the presence of a large peri-aortic tumor at the renal hilum, compressing the left kidney. The patient was treated with antihypertensive medications for several weeks, followed by surgical resection of the tumor (Figure, C). Examination revealed a massive pheochromocytoma, with renal vein tumor thrombus covered by endothelium. During the postoperative follow-up, he had no recurrence of arrhythmia, ventricular dysfunction, valvar insufficiency, glomerular dysfunction, or tumor. Remarkably, results of the MIBG renal scan were negative despite the elevation of urine catecholamine metabolite excretion and a large, compressive tumor.

Figure. A, Electrocardiographic 3-lead rhythm strip obtained at presentation. The rhythm is sinus, with frequent atrial premature beats occurring in a trigeminy pattern. A 24-hour recording revealed 5000 premature atrial contractions. B, Abdominal magnetic resonance imaging shows a large extrarenal mass, measuring 5.5 x 7.5 cm, displacing the left kidney laterally, with compression of the renal structures and large intestine; the renal artery and vein are splayed anteriorly. C, An intraoperative photograph demonstrates the large peri-aortic pheochromocytoma (P). The tumor is exposed by mobilization of the splenic flexure and medial visceral retraction of the left kidney (K), pancreas, spleen, and stomach. The kidney is rotated anteriorly in order to expose the tumor while preserving the renal vessels during resection of the tumor thrombus.

Charles I. Berul, MD
Lawrence Milner, MD
Kevin P. Moriarty, MD
Michael Linehan, MD
Divisions of Pediatric Cardiology, Pediatric Nephrology, Department of Pediatrics, and Pediatric Surgery, Department of Surgery, Floating Hospital for Children at New England Medical Center, Tufts University School of Medicine, Boston, Massachusetts

(Dr. Berul is currently at Boston Children's Hospital, Harvard Medical School, and Dr. Moriarty is currently with Pediatric Surgical Services, Springfield, MA.)
Carotid Artery Thrombosis and Stroke after Blunt Pharyngeal Injury

Kevin P. Moriarty, MD, Burton H. Harris, MD, and Karim Benitez-Marchand, MD

A 2-year-old child fell on a toothbrush, sustaining a pharyngeal injury. Eighteen hours later, localizing symptoms of a cerebrovascular accident became evident. Magnetic resonance angiography showed occlusion of the internal carotid artery and a cerebral infarction. She was treated with anticoagulation and made a complete recovery. Pharyngeal injuries may be complicated by internal carotid artery thrombosis and embolus. Management includes prompt diagnosis, anticoagulation, and carotid artery exploration in selected cases.

Key Words: Pharyngeal injury, Carotid artery, Stroke. Magnetic resonance angiography.

Pharyngeal impalement is a common childhood injury. A recent experience with a child who sustained serious sequelae of such trauma, and in whom rapid, noninvasive diagnosis was made by magnetic resonance angiography (MRA), prompts this report.

CASE REPORT

A 2-year, 5-month-old right-handed girl was found by her mother crying and holding a blood-stained toothbrush at 2:00 PM. There was no active bleeding or obvious trauma. At 7:00 PM, the child was taken to a pediatrician because of mouth pain, but examination showed only a small abrasion of the left side of the palate for which symptomatic treatment was advised. At 8:00 AM the next day, the patient awoke irritable, drowsy, and reluctant to speak. She was seen again by her pediatrician and found to be aphasic and drooling from the right side of her mouth.

On physical examination at our pediatric trauma center, vital signs were stable. There was a 5-mm abrasion on the pillar of the left tonsil. There was no carotid bruit or cervical mass and both carotid and superficial temporal pulses were palpable. Lateral x-ray films of the neck showed retropharyngeal gas on the left. This finding led to direct fiberoptic laryngoscopy that confirmed the edema and contusion of the left lateral posterior pharyngeal wall. A computed tomographic (CT) scan of the neck showed swelling of the soft tissues of the pharynx in the area of the retropharyngeal gas.

Neurologic examination at the time of admission showed that the child was awake but aphasic, with right-sided hemiparesis and hyperflexion. Cranial CT and magnetic resonance imaging (MRI) scans showed infarction in the distribution of the left middle cerebral artery (Fig. 1), and MRA showed occlusion of the left internal carotid artery with decreased filling of the branches of the left middle cerebral artery (Fig 2).

She was given ampicillin, gentamicin, and clindamycin, no other treatment was necessary for the pharyngeal injury. Because of the elapsed time since injury and the radiologic findings of a completed stroke, the child was treated with intravenous heparin. Four hours after admission, she developed right-sided seizure activity that was easily controlled with phenobarbital: subsequent cranial CT scan showed no...
significant change. Coumadin was started when the neurologic signs stabilized. Rehabilitation was begun, and the patient's weakness slowly improved. Seven weeks after injury, she had full motor recovery except for a mild right upper extremity dystonia, and she was speaking in age-appropriate words. Recovery was complete at 15 weeks.

DISCUSSION

Young, active children often carry objects in their mouths, and if they fall, the object can be driven backward and injure the pharynx. We see two or three such injuries each year, most referred from community hospitals. Injury to the palate is often minimal and even perforating wounds of the palate heal with good results.

Hellman described 131 hospital cases of trauma to the palate and pharynx over 17 years, none with neurologic sequelae. Only 30 cases of cerebral infarction after oral trauma have been described since the first report by Caldwell in 1936. The majority have a latency period up to 48 hours before the onset of neurologic signs.

The mechanism of injury in our patient was most likely an intimal tear from blunt trauma followed by internal carotid artery thrombosis, and then an embolus to the middle cerebral artery. It is one of few such cases in the literature, and reports of similar cases have suggested a variety of thrombogenic injuries, embolism, or dissection, with some demonstrated at postmortem examination. The neurologic component can come from extension of the thrombus or from embolism into the intracranial arteries, explaining the latency period for the development of neurologic signs. Most children are thought to tolerate unilateral carotid artery occlusion because of an intact circle of Willis. The neurologic component can come from extension of the thrombus or from embolism into the intracranial arteries, explaining the latency period for the development of neurologic signs. Most children are thought to tolerate unilateral carotid artery occlusion because of an intact circle of Willis.

The evaluation of pharyngeal impalement injuries by imaging studies is controversial. Diagnostic recommendations have included skull films, carotid Doppler examination, CT scan, and selective carotid angiography. MRA and duplex scanning have been used in diagnosing carotid artery disease. The noninvasive MRA was very helpful in our case, promptly identifying both the internal carotid artery thrombosis and the presumed embolus to the middle cerebral artery.

Reported treatments for blunt carotid artery trauma have included observation, anticoagulation, internal carotid artery exploration, and craniotomy, all with variable outcomes from full recovery to hemiparesis to death. Because none of these patients have a diagnostic study of the cerebral circulation and initiation of therapy before the onset of neurologic symptoms, estimates of the effectiveness of treatment are difficult. Anticoagulation may be useful to prevent propagation of thrombus and embolization.

Based on our experience, we recommend that patients with a pharyngeal impalement injury be evaluated with soft tissue films of the neck and direct pharyngoscopy if there are physical findings or radiographic evidence of penetrating injury. Prompt noninvasive vascular studies, either MRA or duplex scans of the carotid arteries, are useful before the onset of neurologic signs in worrisome cases. If injury to the thrombosis of the carotid artery is suspected, angiography should be performed to determine if the lesion is amenable to direct arteriotomy surgery, and then anticoagulation can be instituted to prevent extension of the thrombus or embolization. Even if neurologic signs are already present, our experience suggests that anticoagulation alone may be successful.

REFERENCES