Moyamoya disease—
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As physician co-leaders of Palmetto Health’s neuroscience service,
we share a vision to provide the most advanced neurology and neurological surgery treatments available to the residents of South Carolina. We are excited to share this edition of our neuroscience journal featuring articles about moyamoya disease and a case presentation of Choroid Plexus Papilloma.
Meet our newest physician

Anil Yallapragada, MD

Stroke Medical Director, Palmetto Health Stroke Center  
Assistant Professor of Clinical Neurology, USC School of Medicine  
Neurologist, Palmetto Health-USC Medical Group Neurology

We wish to welcome Dr. Yallapragada, a specialist in stroke care who is board certified in neurology and vascular neurology.

Dr. Yallapragada received his medical degree from the Medical University of South Carolina and completed medical internships at the University of Illinois College of Medicine, as well as the State University of New York Downstate Medical Center. He completed a neurology residency at the University of Chicago Medical Center and a vascular neurology fellowship at the University of California Los Angeles Ronald Reagan Medical Center.

Dr. Yallapragada has received numerous professional honors and awards and is published in several peer-reviewed journals. He has a strong interest in state-wide stroke care and how we improve stroke incidence across the state.

Joe Niekro Foundation holds ongoing aneurysm support group at Palmetto Health

Joe Niekro was a 22-year veteran of professional baseball who suddenly lost his life from a cerebral brain aneurysm in October 2006. The astonishing lack of public awareness and support of research led to the launch of a crusade to educate and encourage awareness about brain aneurysms and other cerebral disorders.

The Joe Niekro Foundation™ is committed to supporting patients and families, research, treatment and awareness of brain aneurysms, AVMs and hemorrhagic strokes. The Foundation provides education on the risk factors, causes and treatments of these conditions, while funding the advancement of neurological research.

A Joe Niekro Foundation support group now meets at Palmetto Health Richland on the third Thursday of each month, 6-8 p.m., 5 Richland Medical Park Dr., Palmetto Health Richland Auditorium, first floor.

For more information, contact Mary Pat Baldauf at ColumbiaSC@JoeNiekroFoundation.org.
Moyamoya disease is a chronic occlusive cerebrovascular disease presenting as either unilateral or bilateral carotid stenosis or occlusion. Since the carotid occlusion takes place over time, the brain has the ability to form collateral vessels over time called moyamoya vessels. These small vessels produce the radiological image of a hazy “puff of smoke,” giving the disease the Japanese name “moyamoya.” Moyamoya can affect children or adults. In children, the disease usually presents with ischemia. In adults, it can present as either ischemia or hemorrhage. Moyamoya can progress, especially in children.

Moyamoya typically presents with intracranial hemorrhage, transient ischemic attack, brain infarction or sometimes seizures. The hemorrhagic type of moyamoya disease is more characteristic of adult onset. There are some cases where the patient is asymptomatic and the disease is found on imaging. In these patients, the disease can progress and the patient could become symptomatic.

While there are genetic features of the disease, sporadically occurring moyamoya disease is still the most common form. The pathogenesis of moyamoya disease still is not completely clear. There is thickened intima of the major vessels of the circle of Willis. Several factors, such as transforming growth factor beta, have been shown to be elevated in the cerebrospinal fluid (CSF) of patients with moyamoya disease. These factors lead to the formation of moyamoya vessels.

The diagnosis of moyamoya disease is usually made with conventional digital subtraction angiography. On angiography, the stenotic or occlusive segment is usually in the supraclinoid internal carotid artery (ICA). The finding of hypertrophy of perforators and neangiogenesis resulting in angiographic blush or “puff of smoke” is diagnostic. Other causes of stenosis or occlusion, such as arteriosclerosis or autoimmune disease, should be ruled out.

For a patient who presents with suspected ischemia, CT of the brain without contrast is first line with CT angiogram. The CT could quickly show if the presentation is due to intracerebral hemorrhage. Subsequently, MRI of the brain should be performed. The CT angiogram or MR angiogram of the brain could show the occlusion or stenosis of the ICA or other vessels involved. Another study which can correlate to the stage of the disease is a CT perfusion. CT perfusion with Diamox challenge can be performed to determine if there is severe decreased blood flow to the hemisphere. However, the gold standard for documentation of the disease is still conventional cerebral angiogram.

Moyamoya disease can be categorized into six stages, as described by Suzuki and Takaku, 1969. Stage 1 is stenosis of the supraseller ICA, usually bilateral. Stage 2 is development of moyamoya vessels at the base of the brain. Stage 3 is increasing ICA stenosis and prominence of moyamoya vessels (most cases are diagnosed at this stage). Stage 4 is involvement of the entire circle of Willis, posterior cerebral arteries (PCA)s are occluded, extra cranial collaterals start to appear and moyamoya vessels begin to diminish. Stage 5 is further progression of stage 4. Finally, stage 6 is complete absence of moyamoya vessels and major cerebral arteries.
Treatment depends on the aggressiveness of the disease. Most cases are treated surgically using revascularization procedures as surgical treatment has been shown to be more effective than nonsurgical treatment. Medical treatments that have been proposed include vasodilators and antiplatelet agents. However, the efficacy of medical treatment has to be further investigated. Ischemic episodes can be managed using antiplatelets. Patients with ischemic presentation and evidence of ischemia on imaging should be started on ASA.

Surgical treatments are divided into three groups: direct, indirect and combined methods. Direct bypass includes mostly superficial temporal artery (STA) to middle cerebral artery (MCA) bypass. Indirect procedures can involve encephalomyoarteriosynangiosis (EDAS) or a variation of this procedure. With the direct procedure, the STA is used for anastomosis to the MCA. In the indirect method, the STA is harvested and placed directly on the cortex. The STA will promote arborization of new vessels which will help the collaterals in moyamoya. In cases of hemorrhagic presentation, revascularization has also been shown to be beneficial, leading to decreases in future hemorrhagic events.

Case illustration
A 34-year-old woman presents with left arm numbness that lasted for 48 hours. On presentation she did not have any motor or sensory deficits. She subjectively reported the numbness and heaviness of the left arm. A CT of her head without contrast was unremarkable. However, an MRI of the brain demonstrated right hemisphere diffusion restriction signifying areas of ischemia (Figure 1). A CT angiogram of the head demonstrated severe supraclinoid ICA occlusion and right supraclinoid ICA and MCA stenosis (Figure 2). The patient subsequently underwent a cerebral angiogram which demonstrated left supraclinoid ICA occlusion with moyamoya vessels (Figures 3A & 3B). A CT perfusion with Diamox challenge also was performed to assess the degree of perfusion. The CT perfusion showed severe perfusion deficit. Since the patient was symptomatic from the moyamoya, we discussed revascularization with the patient and her family. The patient decided to proceed with surgery for revascularization. We discussed direct STA to MCA bypass versus indirect revascularization with EDAS. The patient and family decided to have EDAS performed.

The patient underwent a left craniotomy for EDAS procedure and six weeks later underwent a right craniotomy for EDAS procedure. Both surgeries were without complications and the patient was discharged home in four days. The patient underwent follow up cerebral angiogram six months from surgery, which demonstrated formation of collateral blood vessels on both hemispheres of the brain from the EDAS procedure (Figure 4). The patient has been continued on ASA and has not experienced ischemic symptoms. The patient will be followed by another cerebral angiogram in one year from the date of EDAS.

FIGURE 1 | MRI of the brain with diffusion weighted sequence demonstrating right basal ganglia acute infarct.

FIGURE 2 | Cerebral angiogram from right internal carotid artery demonstrating early moyamoya with stenosis of proximal middle cerebral artery.

FIGURES 3A-3B | Left internal carotid injection, AP projection (left image) and lateral projection (right image) demonstrating the supraclinoid ICA occlusion and moyamoya vessels and perforators which are typical in this disease.

FIGURE 4 | Six month follow up cerebral angiogram from left external carotid artery demonstrating the arborization of new blood vessels from the left superficial temporal artery that was used for the EDAS procedure.

FIGURE 5 | Right external carotid cerebral angiogram, three months after EDAS procedure demonstrating the start of arborization of new blood vessels from the right superficial temporal artery.
Providers now can call 844-64-BRAIN to transfer urgent and emergent neurosurgical and neurological patients easily and efficiently.

Studies have shown that one of the challenges faced by emergency room providers and referring physicians is fast and efficient access to neurological and neurosurgical physicians in tertiary medical centers. Palmetto Health’s 4-BRAIN line allows emergency room providers and referring physicians to speak directly with a neurosurgeon or neurologist without going through an operator or long waits on the phone. Neurological problems that the 4-BRAIN line may be used for include intracerebral hemorrhages, subarachnoid hemorrhage, aneurysms, vascular malformations and brain tumors. The 4-BRAIN line is answered 24 hours a day, seven days a week.

Call 844-64-BRAIN (27246) for emergent neurosurgical transfers.
Choroid Plexus Papilloma in adults: case presentation and review of literature

by Roham Moftakhar, MD
Chief of Neurosurgery, Palmetto Health Richland
Associate Professor of Surgery, University of South Carolina School of Medicine

Case presentation

A 61-year-old woman presents with progressive imbalance and ataxia. Her examination was unremarkable but the patient reported that she was having trouble with balance. An MRI of the brain with and without contrast demonstrated a contrast enhancing fourth ventricular tumor with compression of the medulla and pons (Figure 1). The differential diagnosis was ependymoma, subependymoma and choroid plexus papilloma or carcinoma. The patient underwent a sub-occipital craniotomy with resection of the tumor through a lower transvermian approach. Intra-operative monitoring of lower cranial nerves V-XII as well as SSEP and motor monitoring was performed. The tumor invaded the floor of the fourth ventricle. Careful dissection of the tumor from the floor of the fourth ventricle was performed. Postoperative MRI demonstrated gross total resection (Figure 2). The patient was discharged from the hospital on day number four without any complications.
Review of literature

Choroid Plexus papillomas are rare tumors of the central nervous system arising from epithelial differentiated tissue. These tumors account for only 0.4-0.8 percent of all brain tumors with the most commonly reported location being the ventricles. Choroid plexus papillomas are WHO grade I tumors versus choroid plexus carcinomas which are WHO grade III with worse prognosis. The possibility of progression from papilloma to carcinoma has been documented (Diengdoh and Shaw, 1993).

The etiology of some choroid plexus tumors has been linked to SV40 infections. The SV40 theory has been linked mostly to infants. In a systemic review, Wolff et al., reported the cerebella-pontine location was associated with older age, gender female and benign pathology. This could relate these tumors to a tumor suppressor gene on chromosome X. The etiology of choroid plexus papilloma in children and adults is most likely different.

In cases of suspected choroid plexus papilloma a biopsy is needed to document the diagnosis. Wolff et al., in their systemic review reported that surgery is most powerful step in treatment of choroid plexus papilloma. The extent of resection is important because it impacts outcomes in terms of survival. In fact, their review supported a second surgery if there was residual and it could be safely resected. Choroid plexus papilloma patients with complete resection had a 10-year survival rate of 85 percent (+/- 10 percent standard deviation). Also, the literature supports adjuvant therapy with radiation after residual tumor has been documented in cases of choroid plexus carcinoma. In fact, in cases of choroid plexus carcinoma, radiation should be used after gross total resection. However, in cases of choroid plexus papilloma, the patient could be followed without radiation after gross total resection. The strategy of radiation therapy does not apply to young children in whom there could be long term sequelae. In these cases, chemotherapy would be recommended.

References:
New Neurointerventional suite opens at Palmetto Health Richland

Palmetto Health Richland has upgraded its neurointerventional suite, an operating room that provides neurosurgeons, neurointerventionalists and their teams with the most advanced technologies in the state for treating numerous neurovascular conditions including acute stroke, ruptured and unruptured aneurysms and other conditions affecting the brain or spine.

The suite is equipped with the Siemens Artis Q biplane system, an interventional X-ray imaging device—one of the most advanced systems used in South Carolina. This system improves
visualization so that surgeons can make more informed decisions as they detect and treat various conditions. It also benefits patients since it is more efficient, enabling improved quality images with less radiation exposure. The system may reduce the time required for diagnosis and treatment. The suite is the latest advancement that continues Palmetto Health’s neuroendovascular surgery team’s ability to perform minimally invasive procedures.

Erwin Mangubat, MD, a neuroendovascular surgeon for Palmetto Health-USC Medical Group Neurosurgery, knows all too well that when dealing with stroke, time is everything. He said, “Traditionally, a patient with a suspected stroke would go to a separate CT scanner and, if that patient was found to be a suitable candidate for intervention, would then be brought to the angiographic suite, which wastes valuable minutes. With the neurointerventional suite, we are able to perform those scans at the same place and then begin treatment immediately.”

Roham Moftakhar, MD, chief of neurosurgery at Palmetto Health Richland and medical director at Palmetto Health-USC Medical Group Neurosurgery, has seen the improved outcomes that technology and focus on minimally invasive surgery have provided. He added, “Years ago we had to open up the brain, and the patient was in the hospital recovering for weeks. Now, except for some extreme cases, we no longer have to open up the skull. After surgery, patients are left with only a small dressing in the groin area. Most can go home the next day, and they can get back to work in just a few days in most cases.”
Contact us for more information or to refer a patient

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