A 13-Year-Old Male with Ptosis

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A 13-year-old, previously healthy African-American male presented with a 12-hour history of double vision and inability to open his left eye. This was associated with 8 days of moderate to severe left temporal headache that was made worse by sunlight, television, and noise. He had also experienced intermittent nausea, vomiting, and fever over the prior 10 days. During the initial course of his illness, he took ibuprofen because he was diagnosed with possible migraine at an outside hospital.

On clinical presentation, he was febrile with a temperature of 101.4°F. On cardiac examination, there was a grade 2/6 systolic murmur at the left lower sternal border. Ophthalmologic examination showed left eye ptosis (see Figure 1), mydriasis, and absent consensual/direct light reflex. Extraocular movements were limited in lateral, medial, and downward directions. Visual acuity was 20/30 in the right eye and 20/50 in the left eye. There were no dental caries and no evidence of odontogenic infection. The rest of the physical examination was unremarkable.

Laboratory tests revealed a white blood cell count of 24.3 x 10⁹/L with a neutrophilia of 80.6%. C-reactive protein level was elevated at 18.09 mg/L. Sodium was 127 mEq/L and chloride was 88 mEq/L. Liver function, urine analysis, and HIV tests were normal. Lumbar puncture showed clear cerebrospinal fluid with a high white blood cell count 70 cells/mcL with neutrophils 79%, glucose 65 mg/dL, and protein 51 mg/dL. Antinuclear antibody, antineutrophil cytoplasmic antibody, complement component 3, complement component 4, and immunoglobulin panel, which were performed to rule out a possible vasculitis or immunodeficiency were normal. Echocardiogram showed no evidence of any intracardiac vegetations or valvular abnormalities.

Magnetic resonance imaging (MRI) showed a contrast-enhancing soft tissue lesion involving the left cavernous sinus. Minimal mucoperiosteal thickening involving bilateral ethmoidal and left sphenoid sinus was also seen. MR angiography (MRA) revealed the diagnosis, and subsequent blood culture revealed the causative organism.

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Editor’s note: Each month, this department features a discussion of an unusual diagnosis in genetics, radiology, or dermatology. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at Pediatrics@Healio.com.
Case Challenge

Diagnosis:
Left cavernous sinus thrombophlebitis with mycotic aneurysm of intracavernous internal carotid artery

An MRA scan revealed a 13.1 mm x 5 mm x 9.5 mm lobulated aneurysm involving the left cavernous internal carotid artery (see Figure 2). Blood culture grew *Streptococcus milleri/constellatus*. A diagnosis of septic left cavernous sinus thrombophlebitis associated with intracavernous internal carotid artery mycotic aneurysm was made.

DISCUSSION
Cavernous sinus thrombophlebitis and internal carotid artery aneurysms are infrequently seen in children. When they occur, there is a high risk of morbidity and mortality if not promptly and adequately treated. Here, we discuss a case of an adolescent patient presenting with a cavernous sinus thrombophlebitis that was complicated by a left internal carotid artery mycotic aneurysm associated with *Streptococcus milleri* infection.

Septic cavernous sinus thrombophlebitis is a rare complication of several conditions. The primary source of infection is usually in the head and neck, being caused by such things as otitis media, sinusitis, periodontal problems, cutaneous facial, orbital infections, and dental procedures. In some cases, an etiology is never found; it may be a distant focus with septicemia preceding the thrombosis. Septic cavernous sinus thrombosis can lead to ophthalmoplegia, blindness, cranial nerve palsies, meningitis, and intracavernous internal carotid artery aneurysm, as well as its complications (ie, rupture and carotid cavernous fistula).

The reported incidence of cerebral aneurysms in the pediatric population varies from 0.5% to 4.6% of all cerebral aneurysms. Traumatic and infectious aneurysms are more frequently seen in pediatric age group, and up to 10% are of infectious origin. The word “mycotic” is a misnomer, as the most common pathogens of mycotic aneurysms are gram-positive cocci. Mycotic aneurysms are caused by arterial wall damage from septic emboli. The most common clinical setting reported for mycotic aneurysm is bacterial endocarditis. The cavernous carotid artery is at risk of infection from septic cavernous sinus phlebitis, leading to arteritis due to its location. In this case, the aneurysm most likely resulted from the septic cavernous sinus thrombosis.

Intracavernous mycotic aneurysms are rare. In a review of 36 cases, out of which 13 were pediatric patients, the most commonly found organism was *Staphylococcus aureus*, whereas less commonly encountered pathogens include *Streptococcus* species and gram-negative and fungal pathogens.

Carotid intracavernous mycotic aneurysm usually presents with fever, double vision, headache, retroorbital pain, and ophthalmoplegia. With septic cavernous sinus thrombosis and clinical features of ophthalmoplegia, arteriography is necessary for diagnosing any vascular abnormality. MRA is preferred because it has a good spatial resolution, decreased interference from turbulence, and can be performed in the same time span as a regular MRI. Digital subtraction angiography and computed tomography (CT) angiography can also be used in diagnosing this condition, each with its advantages and disadvantages.

The recommended treatment for unruptured mycotic aneurysm is long-term antibiotic treatment and serial angiography to monitor aneurysm size. Intracranial infectious aneurysms are thin walled, friable, and exhibit a high risk of rupture that may lead to consideration of surgical or endovascular treatment. Repeated imaging after endovascular...
therapy at various intervals is necessary to determine the degree of aneurysm obliteration and assess any change.

The treatment options for the internal carotid artery aneurysm include observation, craniotomy with clipping, and endovascular techniques. In this patient, due to the initial size of the mycotic aneurysm and the lack of change during the course of antimicrobial treatment, it was decided to treat the aneurysm by endovascular technique, as this may prevent complication of aneurysm rupture and carotid cavernous fistula.

Long-term outcome of carotid cavernous aneurysm varies depending upon treatment modality. In recent years, surgical and endovascular treatment-related complications have been reduced significantly due to better pretreatment screening with provocative balloon occlusion test, angiography, better equipment, and techniques for endovascular treatment.9

This patient, with a diagnosis of left cavernous sinus thrombophlebitis and mycotic aneurysm of internal carotid artery, was initially started on broad-spectrum antimicrobials. One of four blood cultures drawn over a period of 24 hours grew *Streptococcus milleri/constellatus* sensitive to the beta-lactam antibiotics. Subsequently, antibiotic treatment was narrowed to ceftriaxone. Cerebrospinal fluid culture remained negative. Clinically, the patient’s headache and fever improved, but eye findings remained the same. A subsequent MRA showed no change in the size of the lesion. Due to the size of the aneurysm and significant risk of rupture, he was transferred to another hospital for endovascular treatment.

CONCLUSION

Septic cavernous sinus thrombophlebitis is uncommon in the pediatric population. This interesting case emphasizes the importance of early diagnosis of cavernous sinus thrombophlebitis and subsequent complications of carotid cavernous aneurysm. Early diagnosis and treatment is critical for an optimal clinical outcome.

REFERENCES