Giant MCA Aneurysm: A Pediatric Case Report

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Abstract

Pediatric cerebral aneurysms comprise anywhere from 0.5-5% of the prevalence of aneurysms in the general population. Giant aneurysms, defined by an aneursysmal size of over 2.5cm, are even less prevalent. Presentation of these aneurysms can vary from increased intracranial pressure, cranial nerve impairment, nausea and vomiting, or mass effect. In this article, we present a patient with a unique constellation of symptoms that presented to the emergency department with a pediatric giant aneurysm and non-specific mass effect symptoms that had a broad etiology and multiple prior workups based on geographical location and other factors that masked the true etiology.

Introduction

Middle cerebral artery (MCA) aneurysms are a form of cerebral aneurysms and account for around 14-43% of all cerebral aneurysms (Yang). Giant aneurysms are described as any aneurysm over 2.5 cm in diameter and typically deriving from brain trauma, connective tissue disorders or infections (2018). While aneurysms are one of the most common vascular anomalies of the central nervous system, they are far less common in children than in adults, with a reported prevalence ranging from 0.5 to 5 percent of the total prevalence of intracranial aneurysms in the general population. Typically pediatric patients with giant MCA aneurysms commonly present with an acute subarachnoid hemorrhage (Shih). Symptoms include acute onset of severe headache, lethargy, tense anterior fontanelles indicating increased intracranial pressure, nausea, vomiting, papilledema, double vision, speech disturbance, hemiparesis or hemiplegia (2018). Unusual symptom presentations in patients with no family history, no comorbidities, or atypical symptoms that would suggest potential for an aneurysm can be difficult to diagnose without imaging. In this case study, we review an 11 year old previously healthy male who presented to the Emergency Department with left side hemiparesis secondary to a giant MCA aneurym and resultant infarction of the basal ganglia.

Case Presentation

This 11-year-old male presented to the emergency department via ambulance with left leg pain which began two days prior and progressed to inability to move the left arm and left facial droop. Two days prior to presentation to the ED, EMS was called when he awakened with left leg pain and difficulty moving. He was reportedly very anxious and woke from sleep hyperventilating. Paramedics believed his symptoms were due to a panic attack and the decision was made to keep him at home. That same day he was seen in the urgent care setting with slight left facial droop, left arm and left leg tenderness in elbow and wrist. Physical exam reportedly revealed full range of motion and strength. Urgent care providers were suspicious of a tick-borne illness, and he was discharged with doxycycline and oral corticosteroids with blood tests for Rocky Mountain Spotted fever and Lyme disease sent. The day of presentation to the ED, the patient returned to urgent care with persistent symptoms and a new complaint of two brief episodes of double vision, each lasting "a few" minutes. The first episode of diplopia occurred the day prior and the second the same morning as presentation to the ED. The second urgent care exam was significant for findings of significant weakness in the left arm and leg, uncoordinated gait and continued slight left facial droop. The patient was then sent directly to our emergency department by ambulance. Upon presentation to the emergency department, he describes tingling from left elbow to left wrist. There was no history of similar symptoms in the past. Review of systems was negative for fever, headache, visual changes, confusion, neck pain, right sided arm or leg weakness, rash, shortness of breath, abdominal pain, nausea, vomiting, diarrhea or trauma. This patient's past medical history was significant only for attention deficit-hyperactivity disorder, treated with 20mg amphetamine-dextroamphetamine and clonidine daily. Other medications include fexofenadine-pseudoephedrine twice daily for allergies and trazodone 50mg at bedtime. At the time of presentation to our emergency department, vital signs were unremarkable and physical examination revealed left facial droop, right uvular deviation with palate elevation, left arm 1/5 strength, 0/5 left hand grip, 3/5 strength in all muscle groups of the left lower extremity. Speech was clear, and he was alert and oriented with appropriate behavior. An MRI of the head with and without contrast revealed a well-circumscribed extra-axial 3.5X2.7X2.2cm homogeneously enhancing T2 hypointense mass in the right Sylvian fissure. Further mass effect was noted on the right temporal lobe and distal segments of the right MCA. A 2.5X1.9cm area of restricted diffusion was noted in the right basal ganglia with T2 hyperintense layering of fluid in the posterior aspect of the right basal ganglia. A 5mm midline shift was seen to the left. MRI without contrast of the spine was unremarkable at all levels. CBC, CMP and U/A were unremarkable except for low bilirubin (0.2

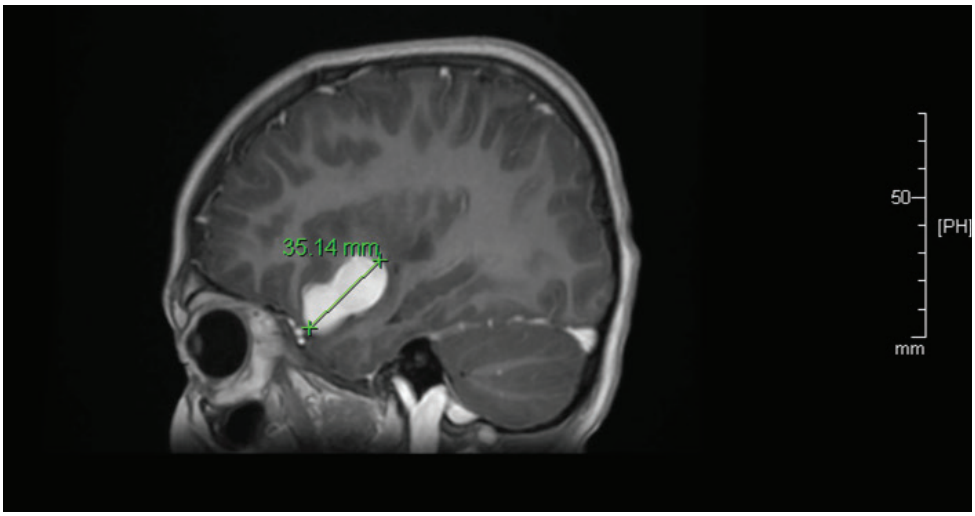

Fig. One: Sagittal view of the brain displaying a 35.14 mm mass.

with reference range 0.3-1.2 mg/dL) and elevated neutrophils (61.4% with reference range 23.0-53.0%). Laboratory specimens for Rocky Mountain Spotted fever and Lyme disease had been collected, but were not resulted during the evaluation in our emergency department. Due to the significant MRI findings it was decided that the patient would benefit from transfer to a tertiary care center with pediatric neurosurgical capabilities. At the tertiary center, the patient was administered aspirin 300mg, clopidogrel 300mg, verapamil 0.5mg/ml intra-arterial, and alteplase 1mg. The giant aneurysm was treated with extensive endovascular coiling.

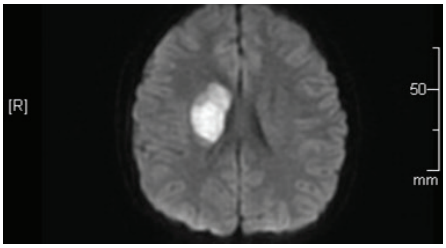

Fig. Two: Coronal view of the brain depicting large mass in the right hemisphere.

Discussion

Particularly remarkable about this patient's presentation was the large, hypointense intracranial mass, initially interpreted as radiologically similar to an extra-axial meningioma, was indeed an giant MCA aneurysm with the ultimate size measured at 3.5X3.3X2.4cm. This originated from the distal M1 segment, at the level of the M2 bifurcation. This was a fusiform aneurysm, with a narrow neck. The mass effect was also resulting in ischemic insult to the perforate arteries supplying the basal ganglia, particularly the lateral lenticulostriate branches, consistent with symptoms described in the patient's initial complaint. "Giant" aneurysms are described as anything over 2.5cm in diameter, and are frequently associated with brain trauma, connective tissue disorders or infections (2018). Recent developments have discovered the pathogenesis of giant intracranial aneurysms; which can be thought of as a vessel wall proliferation induced by inflammatory lipoxygenase activity (Krings). While our patient had no history of trauma or connective tissue disorders, and inflammation measured with ESR was within normal limits; infectious etiologies could be a potential cause in this patient due to the elevated neutrophil count. Currently, there are no studies documenting the incidence of pediatric giant aneurysms; but the current adult estimation is between 0.6-1.4 per 1000c (Paivi). These giant aneurysms are sometimes mistaken for brain tumors, especially when located in a typical tumor location, and mass effect can often be seen as the first clinical sign; as was demonstrated with 18.2% of the cohort with Sharma et al., and 46% of the cohort with Kakarla et al. (Sharma and Kakarla). This patient's aneurysm was located in the typical place for an extra-axial meningioma which is midline, in the area of the Sylvian fissure. This mass effect was indeed compressing the postcentral gyrus both on the cranial surface of the brain and the temporal surface, which could explain the slowly progressive motor dysfunction of the upper and lower extremities of the left side. The aneurysm compressed the perforating arteries feeding the basal ganglia sometime between initial presentation to urgent care and our emergency department, causing the array of symptoms indicating the basal ganglia was infarcted by day two.

This patient's initial complaint of left leg pain and left arm pain with tingling for two days prior to any advanced findings is exceedingly rare. Typical clinical presentation varies, from lethargy, tense anterior fontanelles indicating increased intracranial pressure (in infants), headaches, nausea, vomiting, papilledema, double vision, hemiparesis or hemiplegia all being common findings (Crowley). Seizures and increased intracranial pressure were present in 25% and 90% of giant aneurysms respectively, but all of these presentations should raise the index of suspicion for mass effects of an aneurysm (Levy). For our patient, clinical indication of a child in the midwestern US with tingling pain radiating from elbow to wrist, in addition to painful left leg that awakened him from sleeping and facial nerve paralysis causing facial droop suggested a differential diagnosis with tick-associated infection at the top of the list. When taken in combination of the one day delayed presentation of double vision, the facial droop could have masqueraded the intracranial findings as it is a commonality between diagnoses, but further advancement of ischemic changes to the basal ganglia upon presentation to our emergency department were the first definitive diagnostic symptoms of the intracranial mass effect this aneurysm demonstrated. Furthermore, this patient had the overall symptom progression from pain to full ischemic symptoms over the course of two days; indicating that the initial pain being experienced was not the basal ganglia being infarcted, but was an atypical presentation of central gyri compression. Supporting this is the fact that the patient's motor symptoms progressively worsened over the course of those two days, as compression worsened including temporal lobe compression affecting the upper extremity ipsilaterally.

By the time our patient arrived at the emergency department, the presentation was that of an intracranial infarct or mass with acute neurologic findings. However, when appreciating the patient history and progressive two-day pain and concern of Rocky Mountain Spotted fever or Lyme disease with pending results, it can be much less definitive of a diagnosis to make. With a prevalence estimated between 0.5-5% of all aneurysms in the general population, this giant aneurysm would not be at the top of any practitioner's differential, however after an initial workup it should come to the forefront (Shih). Clinical imaging studies such as MRI with angiography can definitively diagnose this condition with a sensitivity of 89% if the aneurysm is greater than 3mm as occurred in our patient (Okahara). This was accomplished with both an MRI and an MRA of the brain, and transfer of care to a tertiary hospital with pediatric neurosurgical capabilities was initiated. Currently, endovascular coiling from a neurosurgical trained provider is warranted as the most reliable means and the best overall strategy for preventing fatal complications of acute hemorrhagic or ischemic complications, especially with cases of aneurysmal formation, but no comparison has been made between coiling and clipping aneurysms at this time (Thompson).

Learning Points

This patient presentation was a constellation of symptoms that can masquerade as a variety of diagnoses depending on the location geographically, past history, family history and age of the patient. While presenting with symptoms indicative of tick-borne illness, history and timing implicate a mass or aneurysmal etiology. Giant aneurysms are increasingly rare in the pediatric population, and exceedingly rare in the population as a whole. However, aneurysmal complications like mass effect must be included as a diagnosis that must not be missed; saving the life of our patient.

Acknowledgements

The patient's family for allowing us to discuss their treatment and symptom presentation.

Deaconess Healthcare system for allowing us to participate in review of this patient's care.

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