pediatric stroke

Isolated Lenticulostriate Aneurysm Causing Intracerebral Hemorrhage in a Pediatric Patient

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Abstract:
Nontraumatic intraparenchymal hemorrhage (IPH) is an uncommon clinical entity in pediatric patients. Aneurysmal IPH is an uncommon underlying cause, and lenticulostriate aneurysms (LSA) are even less common. We present a case of a 9-year-old male with no known risk factors who presented with encephalopathy and right sided weakness, who was found to have a nontraumatic IPH. Angiographic studies demonstrated a left medial lenticulostriate aneurysm. The patient underwent endovascular repair of this aneurysm. In this case report we review the pathogenesis and treatment of these aneurysms.

Introduction:
Nontraumatic intraparenchymal hemorrhage (IPH) is an uncommon entity in the pediatric population and when present, often has an idiopathic etiology. The overall incidence of stroke in children ranges between 2 and 13 per 100,000[1-4], half of which are hemorrhagic [4].Vascular malformations are the underlying etiology for 40-90% of pediatric intracranial hemorrhage,[5, 6] a minority of which are aneurysmal. While most pediatric aneurysms involve the anterior circulation,[7] involvement of the distal lenticulostriate arteries is exceptionally rare, with less than 50 reported cases in adults and only 6 in children[8, 9]. We present the case of a 9-year-old male who presented with intraparenchymal hemorrhage of the left basal ganglia secondary to a lenticulostriate aneurysm.

Methods:
A retrospective chart review was performed. Verbal consent was obtained from the family, and our institutional review board does not review individual case reports.

Clinical Case:
A 9-year-old male with no significant past medical history presented with confusion, disorientation, unintelligible speech and associated weakness of the right lower face, right upper extremity, and right lower extremity. Prior to presentation, he was playing basketball with friends, but with no witnessed trauma. Blood pressure on presentation to tertiary center was 130/84 mmHg. Initial work-up including urine drug screen and coagulation studies were unremarkable. Non-contrasted head computed tomography (CT) demonstrated a 6 mL IPH centered around the anterior limb of the left internal capsule, with a small associated intraventricular hemorrhage (**Figure 1**). CT angiogram (CTA) of the head and neck demonstrated intact vasculature with no feeding vessel or concern for vascular malformation. In addition, there was no findings on CTA consistent with fibromuscular dysplasia, tortuosity of the vessels, nor dissection. Initial Pediatric NIH stroke scale (PedNIHSS) was 14 for level of consciousness, level of consciousness questions, facial palsy, not holding any extremity antigravity, dysarthria, and minimal aphasia. There was no excessive joint laxity on exam. Parents reported that patient had recurrent epistaxis but denied melena, hematochezia, dyspnea on exertion, telangiectasias, or other neurocutaneous stigmata. The patient remained normotensive and did not require anti-hypertensive therapy from initial presentation to our institution until discharge. On hospital day 3, digital subtraction angiography (DSA) was performed, demonstrating an aneurysm versus pseudoaneurysm of a left medial lenticulostriate vessel measuring 2.6 x 3.0 x 1.8 mm (AP x CC x TV). Microcatheter angiography confirmed that the medial

lenticulostriate vessel supplied the aneurysm and minimal branching vasculature supplied nearby parenchyma. Accordingly, n-butyl cyanoacrylate (n-BCA) embolization of the aneurysm/pseudoaneurysm was performed. Upon removing the microcatheter and repeating angiography from the left internal carotid artery, a small amount of active extravasation from the medial lenticulostriate artery near the previous microcatheter location was appreciated. The blood pressure was reduced to less than 100 mmHg systolic and the proximal medial lenticulostriate artery was re-catheterized. Microcatheter angiography demonstrated that the extravasation had resolved spontaneously. Postoperative dual-energy CT demonstrated stability of the hemorrhage without any adverse change. Initially the patient had weakness in his right upper and lower extremity postoperatively. This largely resolved over the course of two days and the patient was discharged with 4+/5 strength in the right upper and lower extremities. Given the atypical aneurysm location, patient underwent genetic testing (Invitae® Connective Tissue Panel, 92 genes) which did not reveal any pathologic variants.

Discussion:
Intracranial aneurysms are rare in children, although constitute a significant proportion of intracranial hemorrhage. The most common location of aneurysms in children is the internal carotid artery.[10] Both in adults and children, of which there were three documented cases of pediatric presentations, a majority of lenticulostriate aneurysms are considered idiopathic, although lenticulostriate aneurysms have been associated with arterial dissections, moyamoya disease, rheumatic diseases, arteriovenous malformations, and intraventricular tumor.[8, 9] The mechanism of aneurysm formation in children is relatively unknown; however, there have been correlations between genetic connective tissue disorders and aneurysm [11] lending credence of tissue and cellular abnormalities that lead to fragile vessel wall and increase risk of aneurysm formation. Atypical aneurysm location, such as the distal lenticulostriate artery, should raise suspicion for underlying vascular disorders (e.g., connective tissue disorders, neurofibromatosis type 1, autosomal dominant kidney disease). Angiography in our patient did not demonstrate additional vascular pathology beyond the distal lenticulostriate aneurysm. If initial DSA in non-traumatic IPH is non elucidating, then could consider repeat DSA in the future, much like is done in the setting of non-traumatic subarachnoid hemorrhage. Genetic testing for hereditary connective tissue disorders was negative in our patient, specifically including COL3A1, COL4A1 and PKD2 genes, but should be considered in all individuals with atypical aneurysms, especially if personal or family history may suggest an underlying syndromic cause.

Treatment of aneurysms usually involve surgical or endovascular management. Typical surgical treatment includes aneurysm clipping or ligation of the parent vessel, although this intervention may be limited by deep location of aneurysm. Thus, as endovascular techniques have evolved, coiling and embolization have become favored interventions. There is a paucity of literature which describes the treatment of such vascular lesions. Management with open surgical techniques, endovascular embolization with an embolic agent or coils, and observation have all been described in the literature.[12] Damage to the parent vessel is an inherent risk of endovascular treatment of small caliber, fragile vessels, such as the lenticulostriate arteries, though endovascular treatment is likely to result in much lower perioperative morbidity compared to open surgical approaches in these deep locations.

Conclusion:
Lenticulostriate aneurysm is an uncommon finding and cause for intraparenchymal hemorrhage in pediatric patients. This case highlights the continued rarity of this pathology and describes a straightforward treatment strategy.

Figure 1: CTH 2.9 x 2.2 x 3.4 cm (AP x TR x CC), unchanged hemorrhage with adjacent parenchymal edema, and small amounts of ventricular hemorrhage in the left frontal horn, right foramen of Monroe, the nondilated third ventricle, the left ventricular atrium and temporal horn and left side of the fourth ventricle.

Figure 2: (A)DSA demonstrating a left medial lenticulostriate vessel aneurysm or pseudoaneurysm measuring 2.6 x 3.0 x 1.8 mm (AP x CC x TV), (B) Successful n-BCA embolization of an aneurysm or pseudoaneurysm arising from a left medial lenticulostriate vessel measuring 2.6 x 3.0 x 1.8 mm (AP x CC x TV)

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