pediatric stroke

Case Report

Corkscrew Basilar Artery: Incidental Anomaly in a Pediatric Patient with a Prolactinoma

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Abstract

Background: A corkscrew basilar artery anatomical variation is incredibly rare. We report the incidental finding of a corkscrew tortuous basilar artery anatomical variation in a pediatric patient with a prolactinoma. *Lessons*: We discuss a possible origination hypothesis for this vascular abnormality and management of the patient's basilar artery anomaly.

Abbreviations: Magnetic Resonance Imaging (MRI), Magnetic Resonance Angiograph (MRA) and Computed Tomography angiography (CTA), Digital Subtraction Angiography (DSA)

Introduction: We report the incidental finding of a corkscrew tortuous basilar artery anatomical variation in a pediatric patient with a prolactinoma.

Illustrative Case

We report the case of an incidental corkscrew basilar artery in a pediatric male patient with a sellar mass lesion. The patient initially presented at our tertiary center at 9 years of age with a 3-month history of left frontal headaches extending into the left eye. He was neurologically intact on physical examination with normal visual acuity and visual fields on ophthalmological evaluation. He underwent magnetic resonance imaging (MRI) of the brain that demonstrated a sellar lesion (Fig. 1A) as well as a 5-mm diameter basilar artery lesion concerning for a fusiform

aneurysm or paired basilar arteries (Fig 1B). On endocrinology evaluation, his prolactin was significantly elevated at 261 ng/ml (normal value: <25ng/ml) and he had normal thyroid stimulating hormone, T4, growth hormone, insulin growth factor-1, and cortisol levels. This was diagnostic for prolactinoma. Surgical intervention was not performed as he demonstrated no neurological dysfunction and had normal visual fields and acuity. The patient was started on cabergoline for medical management of his prolactinoma.

Due to the incidental finding of a fusiform basilar aneurysm or paired basilar arteries on MRI, further vascular imaging involving magnetic resonance angiograph (MRA) and CT angiography (CTA) of the brain were obtained to better characterize the anatomical morphology of the lesion. MRA and CTA demonstrated a proximal basilar artery that was dilated and tortuous in its course at the mid-basilar region with an expanded diameter of the conglomerate structure of 5 mm. There were no signs of thrombus or intimal dissection on 3D reconstruction of the CTA. The diagnosis of corkscrew basilar artery was made and a fusiform aneurysm was ruled out. The patient was discharged and followed serially by MRI brain at 3 months and 1 year. His prolactinoma responded well to cabergoline with almost total resolution at 1 year. Additionally, there was no change in characteristics of his vascular abnormality at 1 year follow-

up. At 2 years follow up, he underwent CTA of the head that was concerning for basilar artery dissection and prompted evaluation via digital subtraction angiography (DSA) of the brain. DSA demonstrated again the tortuous corkscrew course of the basilar artery without dissection or fusiform aneurysm. This provided a definitive diagnosis of a tortuous basilar artery.

Discussion

Observations

A tortuous or corkscrew basilar artery is a rare anatomical variant that has only been reported twice in the neurosurgical literature. [1,2] The first case was reported by Moser et al. in 2007 of a pediatric patient with an incidental finding of tortuous basilar artery after a trauma. [2] Lim and Chung in 2012 reported the second case of a corkscrew basilar artery in an adult patient with a ruptured basilar artery aneurysm. [1] Here we report a tortuous basilar artery in a pediatric patient with a prolactinoma.

Three main differential diagnoses for the presence of a tortuous or corkscrew intracranial artery are fibromuscular dysplasia, remote arterial dissection leading to alterations in connective tissue that results in tortuous morphology, and anatomical variant. Fibromuscular

dysplasia is well described in the adult population with vascular imaging demonstrating a typical string of beads arterial architecture resulting in a corkscrew pattern.[3] However in pediatric patients vascular imaging does not demonstrate the typical string of beads pattern or corkscrew arterial architecture of fibromuscular dysplasia.[3]

Another hypothesis is that of a previous remote dissection leads to changes in the connective tissue of the blood vessel that leads to spiraling of the dissected lumen around the true lumen and results in the tortuous kinking morphology.[4] Wei et al identified a higher incidence of intracranial vascular tortuosity in pediatric patients after developing a dissection as compared to other etiologies of ischemic stroke.[4] Furthermore, Saba et al identified a higher likelihood of kinking or coiling with carotid artery dissection.[5] However, the patient had no history of remote ischemic strokes or acute ischemic infarct on his presentation which makes remote cerebral vascular dissection an unlikely etiology.

This patient's presenting phenomenon is most likely a variant of the embryological formation of the basilar artery. The embryological formation of the posterior cerebral circulation is induced by the occipital lobe and brain stem growth resulting in hypoxic/ischemic factors that promote

angiogenesis. [6] The posterior fossa is initially vascularized by two parallel neural arteries supplied by the carotid-vertebrobasilar anastomoses of the trigeminal artery, otic artery, hypoglossal artery, and proatlantal artery. [4] The basilar artery is formed by consolidation of the neural arteries and provides the majority of the blood flow to the posterior circulation.[6] It is during these key embryonic stages that anatomical variants may form. [6] However, there is no reported evidence that this is the cause of the corkscrew basilar artery.

Although the embryological origin of the corkscrew basilar artery is an important factor in posterior cerebral blood circulation, of higher importance is the risk for developing vascular disease, namely aneurysmal formation and ischemia from these anatomical variants. There is sparse pediatric literature on the natural history of aneurysms. The mechanisms of aneurysmal formation have been attributed to genetics, inflammation, dysregulation of vascular homeostasis, and wall shear stress. [7] This has been demonstrated in adults with an upregulation in inflammatory markers leading to dysregulation of vascular homeostasis induced by risk factors such as smoking, hypertension, and atherosclerosis. In pediatric patients who do not have these risk factors, we postulate that genetics and wall shear stress result in dysregulation of vascular homeostasis, leading to aneurysmal formation. In their study of

unruptured pediatric aneurysms, Bisson et al. demonstrate that 43.1% of pediatric aneurysm patients are syndromic, thereby, associating a genetic correlation to aneurysm formation. [7] This is not the case with this patient; therefore, we look to wall shear stress as a predictor for future aneurysmal formation in this patient. Wall shear stress is best defined as the frictional force of blood per unit area of endothelium and thereby is the force exerted by blood on the blood vessel. [7,9] Due to the tortuosity of the corkscrew basilar artery, there is a concern that this might lead to increased wall shear stress from blood flow resulting in an increased risk of aneurysm formation. [7,9] Additionally, the tortuosity of the basilar artery also raises a concern for asymmetric length, caliber, and size of the basilar perforators to the brainstem and cerebellum. [2,10] This may lead to an increased risk for asymmetric perfusion and ischemic events. [2,10]

Lim and Chung reported a case of a tortuous basilar artery associated with a ruptured basilar artery aneurysm in an adult patient, while Moser et al. reported a tortuous basilar artery in a pediatric patient with no congenital anomalies. [1,2] These authors suggest that there is a higher likelihood for development of aneurysms in patients with long tortuous basilar arteries.

Serial surveillance with vascular imaging is needed to evaluate for any future aneurysmal formation.

Imaging modalities for vascular anomalies include MRA, CTA, and DSA. [11,12] Each has varying sensitivities and specificities with the gold standard being DSA. [11,12] This patient was evaluated initially with MRI and MRA of the brain due to symptoms of headaches on presentation concerning for tumor. His MRI demonstrated a prolactinoma and MRA revealed the corkscrew basilar artery that was initially thought to be a fusiform aneurysm. Further characterization of the basilar artery with CTA and DSA demonstrated that the basilar artery dilatation seen on MRA was actually the corkscrew tortuous architecture of the variant basilar artery. If symptoms of ischemic stroke present, imaging should be pursued as there is an increased risk of dissection with increased vessel tortuosity in pediatric patients. [13] With the concern for future formation of aneurysms based on the arterial architecture of the corkscrew basilar artery and the novelty of a corkscrew basilar artery variant, we propose a monitoring mechanism involving DSA of this anatomical variant. We recommend monitoring the basilar artery with MRA and confirmation of morphologic change with DSA to prevent need for frequent procedures and radiation exposure. In addition, if basilar artery pathology is suspected

based on MRA or CTA in pediatric patients, confirmation of the suspected abnormality with DSA is recommended as it may result in need for treatment for hemorrhage prevention (if indeed basilar artery aneurysm).

Lessons

The tortuous corkscrew basilar artery, although asymptomatic, can cause future complications. This rare abnormality may lead to an increased risk of aneurysms and ischemic events, which requires serial surveillance.

Disclosures: None

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Figure Legends

FIG1. **A**) Initial MRI demonstrating prolactinoma. **B**) Initial MRI of the corkscrew basilar artery.

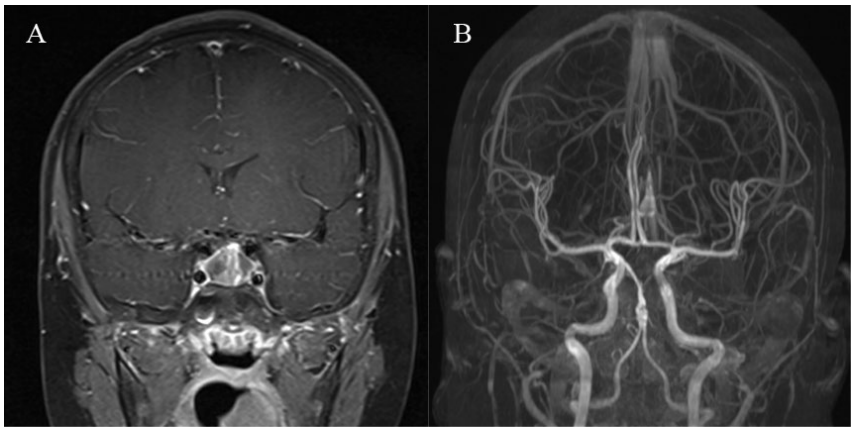

FIG2. **A**) Corkscrew basilar artery without dissecting/fusiform aneurysm seen on digital subtraction angiography at 2-year follow up. **B**) Corkscrew basilar artery seen on 3D reconstruction of CT Angiography.