pediatric stroke

Case Report

An Unexpected Etiology of Acute Quadriparesis in a Pediatric Patient: An Anatomical Review

Samuel Kamoroff, MD, MHA[a], Douglas R. Nordli III, MD[a], Harry Abram, MD[b]

a. Department of Child and Adolescent Neurology, Mayo Clinic College of Medicine and Health Sciences, Jacksonville, Florida, USA
b. Department of Child and Adolescent Neurology, Nemours Children's Health System, Jacksonville, Florida, USA

Corresponding Author:
Samuel Kamoroff, MD, MHA
Mayo Clinic
4500 San Pablo Rd S.
Jacksonville, Florida 32224
Phone: 904-613-6973
Sam.kamoroff@nemours.org
Kamoroff.samuel@mayo.edu

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Introduction

Acute quadriparesis is a medical emergency. Stroke, especially in the pediatric population, is a rare cause of acute quadriparesis but an increasingly important etiology to consider. While strokes are more common in adults, they still occur in the pediatric population. 22% of children with acute ischemic strokes (AIS) involve the posterior circulation [1]. Further, 50% of strokes that involve the posterior circulation are secondary to vertebral artery dissection [2]. Therefore, we must consider stroke in the differential diagnosis of acute weakness in pediatrics. It is important to understand the causes of stroke in pediatrics as it can widely differ from the adult population. Some etiologies of stroke have no age boundary and can present in a classic fashion, such as a vertebral artery dissection leading to a spinal cord infarction. Accurate localization of stroke requires a thorough neurologic examination and deep knowledge of neurovascular anatomy. The following case reviews an unexpected but important presentation of acute quadriparesis stemming from a posterior circulation lesion in a pediatric patient.

Case Presentation

A 13-year-old girl with no past medical history presented to the emergency department two days after new onset neck pain and difficulty walking. She described that she felt a “pop” on the left side of her neck followed by left upper and lower extremity weakness, difficulty ambulating, and mild sensory changes described as numbness. She reported no history of neck injury, trauma, whiplash, or other accidents. The acute neck pain was associated with headache, nausea, and vomiting. At home, these symptoms progressed to acute quadriparesis. Family history was significant for the patient’s father having a stroke in his 40’s of unknown etiology. There was no other pertinent family or medical history.

Given her deficits, she was evaluated through the hospital code stroke pathway. Her initial NIHSS was five with pertinent findings of left arm and left leg with some effort against gravity (2 each) and right leg with drift (1). Acute intervention was discussed, but as last known normal was over 24 hours prior, she was not a candidate for pharmacologic thrombolysis or mechanical thrombectomy.

Neurologic examination revealed no deficits in cranial nerves 1-12. Motor testing revealed weakness of the left upper extremity. The patient had some effort against gravity in the left upper extremity with distal weakness greater than proximal. Her left lower extremity revealed drift. She had full strength of the right upper extremity and mild drift in the right lower extremity. There were no deficits to light touch, pinprick or proprioception throughout. The patient was hyperreflexic 3+ (Mayo Grading scale) in the right upper and right lower extremity and 2+ in the left upper extremity and left lower extremity. There was no ankle clonus bilaterally. Toes were down-going with plantar stimulation bilaterally. There was normal finger to nose and heel to shin on the right side. The finger to nose and heel to shin examination of the patient’s left side was confounded due to weakness.

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Stroke protocol magnetic resonance imaging (MRI) of the brain without contrast revealed bilateral cerebellar infarcts on diffusion weighted imaging (DWI) (Figure 1a) that corresponded to apparent diffusion coefficient (ADC) (Figure 2) images. Stroke protocol magnetic resonance angiography (MRA) revealed vertebral dissection of the left 3rd and 4th segments (Figure 4). The radiographic findings on MRI and MRA were insufficient to explain the acute presentation of quadriparesis that the patient experienced. Therefore, a lesion elsewhere along the neuro-axis was suspected. Subsequent MRI of the cervical spine revealed significant findings for an acute infarct at levels of C3 and C4 (Figure 3).

Workup for inflammatory or hypercoagulable etiologies was unremarkable and there was no reported family history of hemoglobinopathies or hypercoagulopathies. Transthoracic echocardiogram with bubble study was normal. Therefore, she was started on anti-platelet therapy with aspirin 325 mg for the first 24-48 hours and then transitioned to anticoagulation with enoxaparin 60 mg two times daily for diagnosis of spontaneous vertebral artery dissection with secondary cervical spine infarction. During her admission, her physical examination continued to show improvements in left hemi-body weakness and she was discharged for continued rehabilitation. Neurologic exam at 1 month follow-up visit demonstrated mild to moderate (3+ to 4- out of 5) left-sided weakness with no motor deficits noted on the right side. She had reduced sensation to pinprick in left upper extremity, left lower extremity and right lower extremity. Reflexes were mildly increased in right upper and lower extremities, otherwise no Babinski or clonus was present. Coordination and gait assessment were normal.

Discussion

Pediatric presentations of acute quadriplegia are rare but grave. Such presentations require a detailed history with a swift and thorough physical examination to narrow the broad differential diagnosis (Table 1) and provide therapy as quickly as possible. The purpose of a detailed history and thorough examination is to localize the lesion as accurately as possible. A good pre-test hypothesis ultimately increases the accuracy of diagnostic testing. Cases of posterior circulation deficits may be secondary to pathology involving the vertebral arteries, basilar artery, posterior cerebral arteries, or the smaller branches emanating from these main vessels [7]. Vertebral artery dissection occurs when a tear forms in the innermost layer of the artery (tunica intima) and allows blood to enter the middle layer (tunica media) [3]. This formed space disrupts normal blood flow within the artery and allows for clot formation [4]. Once a clot forms, it can break off and cause downstream infarction.

Dissection can occur spontaneously or as the result of trauma. Traumatic causes of vertebral artery dissection include whiplash injury, direct trauma to the neck, chiropractic manipulation, coughing or routine “cracking” of the neck [3]. Spontaneous causes are likely secondary to underlying conditions that weaken the strength of the arterial wall itself. Causes include Marfan syndrome or Ehlers-Danlos

syndrome [3]. Other genetic mutations have also been implicated, although rare. One must also consider Bow Hunter Syndrome, a syndrome where the vertebral artery is compressed against the bone with neck turning leading to neurologic symptoms. This was less likely as the patient did not experience symptoms with neck movement. Bow Hunter Syndrome is an important differential to consider as cases may remain undiagnosed without dynamic angiography testing. The most common location of vertebral artery dissections occurs between the first and second cervical vertebrae as there is significant movement of the artery given the neck's ability to flex and extend in this area [3]. One must keep in mind the level or segment of the dissection with the vertebral artery as blood supply to the spine can also be affected as in this case. Commonly, dissections can be appreciated with magnetic resonance imaging of the neck. However, cerebral angiography remains the gold standard method of definitive diagnosis. [5]

Treatment is directed towards modifying risk of subsequent stroke events and healing of the existing dissection through anticoagulation or anti platelet agents [3]. Angioplasty with stent placement, surgery and thrombolysis may also be considered in the treatment of dissection. The risk of subsequent events has been estimated to be about 1% annually and remains a concern, especially in the pediatric population [4]. Surgical thrombectomy of infarcts in the spinal cord is not possible due to the narrow nature of the vessels in the spinal column.

Often, it is tempting to become anchored on common differential diagnoses such as transverse myelitis, epidural hematoma or intramedullary neoplasm. Stroke is often not be considered in the pediatric population which can account for delayed times to diagnosis and treatment. However, given the significant morbidity associated with stroke and the possibility of timely disability limiting intervention, it is incredibly important to consider in a patient with acute weakness.

The presentation of spinal cord infarction secondary to a vertebral artery dissection in a pediatric patient may not be expected but can account for around 50% of posterior circulation strokes [2]. Such presentations are important to recognize, and report given the rarity of pediatric strokes. The incidence of vertebral artery dissection in the adult population is 1-1.5 cases per 100,000 people [3]. In the pediatric population, the overall incidence of vertebral artery dissections in cases of stroke is between 10-25% [3]. Genetic factors have only been implicated in 1-5% in cases of heritable connective tissue disorders resulting in spontaneous vertebral artery dissection [4]. The above-described case was fortunately quickly diagnosed but lacked traditional risk factors and clues that would guide the untrained provider to suspect spinal cord infarction.

No triggering event was found in our case, suggesting a spontaneous dissection and subsequent spinal cord and cerebellum infarction. The spontaneous dissection likely gave rise to a clot that flowed downstream from the vertebral artery into the anterior spinal artery subsequently causing infarction.

Most pediatric stroke protocols do not include MRI of the spine during the acute phase. The deficits appreciated on the code stroke sequence MRI brain in our patient did not match with the presenting symptoms, thus careful consideration was given to alternate explanations such as spinal cord stroke. The MRI spine was added to the code stroke protocol MRI brain and ultimately provided an explanation that fit with neurologic exam localization and symptoms upon presentation. Upon literature review, the location of the spinal cord lesion is similar in presentation with the case described by De la Torre et al [8]. This potentially implicates the 4th segment of the vertebral artery being susceptible to dissection.

Typically, in a pediatric patient with a family history of stroke in the young and a presentation of spontaneous vertebral dissection, more advanced diagnostic and genetic studies are warranted. These could include dynamic vertebral angiography to assess for Bow Hunter Syndrome and whole exome sequencing to determine if there are any identifiable and related pathogenic variants. One must also not forget fibrocartilaginous embolism as a possible source [8]. After hospitalization, patient had one follow up visit that showed improvement in overall examination but still had predominantly left side focal deficits. Subsequent CTA of the neck obtained 2 months later demonstrated chronic left vertebral dissection (Figure 5).

Conclusion

Pediatric stroke as a cause of acute quadriparesis secondary to a spinal cord stroke from vertebral artery dissection may contribute to primary symptoms more than most may realize. The etiology is well described in adults and highly associated with trauma and conditions such as Ehlers-Danlos Syndrome, Marfan Syndrome, or more rarely, Bow Hunter's Syndrome. Increased recognition of stroke in pediatric patients is important as acute interventions can be offered. One of the main challenges of pediatric stroke is still recognition in both the community and hospital settings. Importantly, as highlighted in this case, the discrepancy of clinical examination findings and radiographic findings resulted in correct diagnosis in this patient and should always be considered for pediatric stroke patients. Future guidelines may consider the addition of protocoled cervical spinal cord imaging in cases of suspected vertebral artery dissection.

References

Carey S, Wrogemann J, Booth FA, Rafay MF. Epidemiology, Clinical Presentation, and Prognosis of Posterior Circulation Ischemic Stroke in Children. Pediatr Neurol. 2017 Sep;74:41-50

McCrea N, Saunders D, Bagkeris E, Chitre M, Ganesan V. Diagnosis of vertebral artery dissection in childhood posterior circulation arterial ischaemic stroke. Dev Med Child Neurol. 2016 Jan;58(1):63-9.

Park, K.W., et al., Vertebral artery dissection: natural history, clinical features, and therapeutic considerations. J Korean Neurosurg Soc, 2008. 44(3): p. 109-15.

Britt, T.B. and S. Agarwal, Vertebral artery dissection. 2017.

Rafay, M.F., et al., Craniocervical arterial dissection in children: clinical and radiographic presentation and outcome. J Child Neurol, 2006. 21(1): p. 8-16.

Tan, M.A., et al., Late Complications of Vertebral Artery Dissection in Children: Pseudoaneurysm, Thrombosis, and Recurrent Stroke. Journal of Child Neurology, 2009. 24(3): p. 354-360.

Rollins N, et al., Dynamic Arterial Compression in Pediatric Vertebral Arterial Dissection. Stroke. 2017 Apr;48(4):1070-1073.

Susan De la Torre, et al., Spinal Cord Infarct in Pediatric Patient. Pediatric Stroke, 2021; 1:63-75.

Table 1: Etiologies of Acute Quadriplegia in Children

Vascular	• Spinal cord infarction • Spinal cord hemorrhage • Epidural hematoma
Infectious	• Bacterial osteomyelitis • Spinal abscess • Human Immunodeficiency Virus • Acute Flaccid Myelitis
Traumatic	• Traumatic spine injury
Autoimmune	• Transverse Myelitis • Multiple Sclerosis • Systemic Lupus Erythematosus
Metabolic	• Vitamin B12 deficiency • Dural Arterio-Venus fistula
Inflammatory	• Spondylosis
Neoplasms	• Metastatic tumors • Primary tumors
OTHER	• Hereditary spastic paraparesis • Decompression syndrome • Degenerative motor neuron disease

Figure 1.

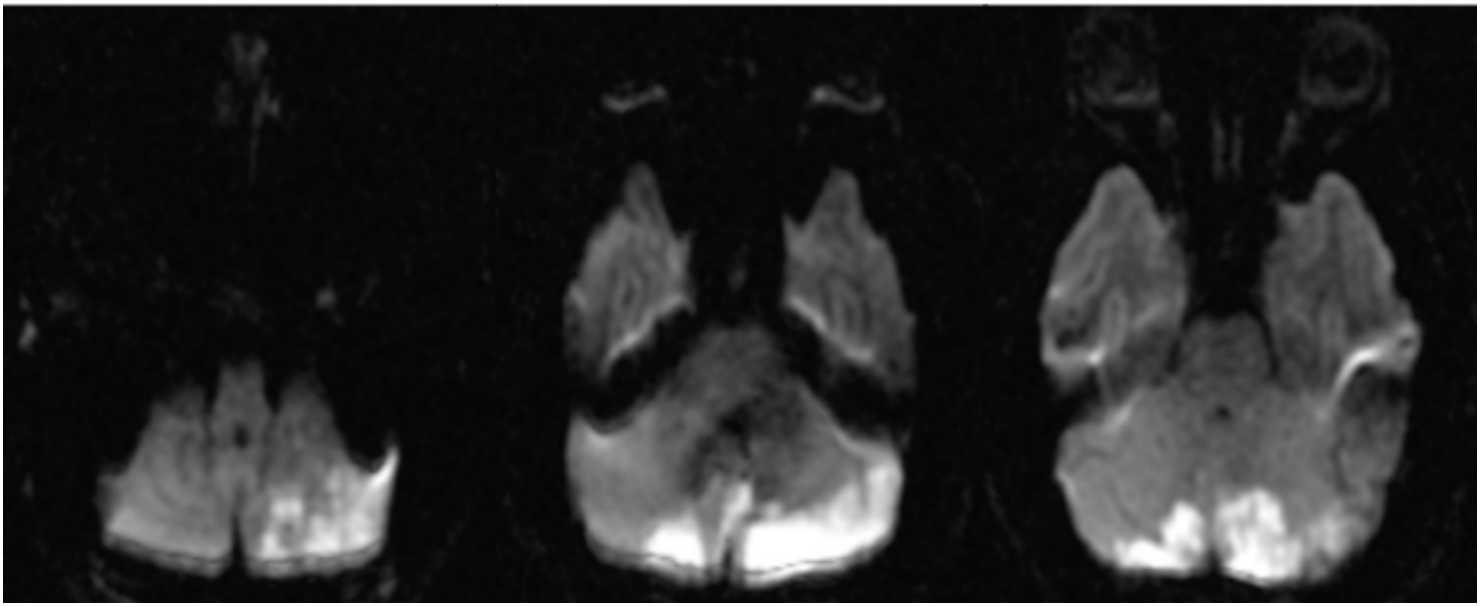
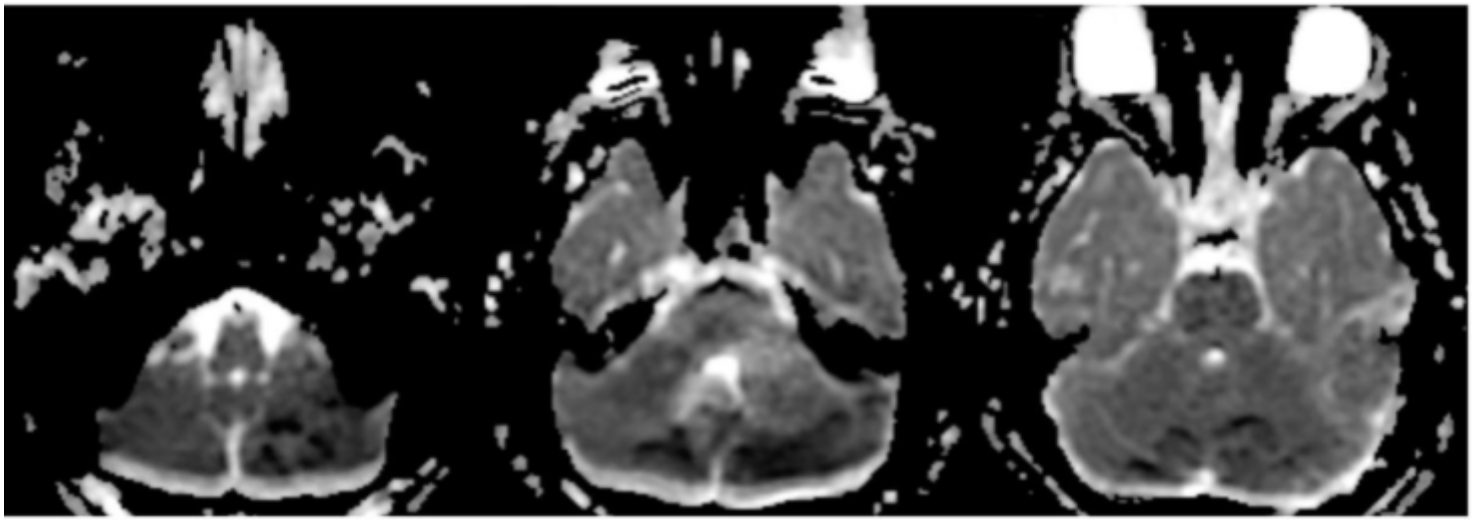


Figure 2.



Figures 1 and 2: *Hyperintense signals seen on diffusion weighted image (DWI)* (Figure 1) with (Figure 2) *Apparent diffusion coefficient (ADC) correlation* noted in left greater than right cerebellar regions indicating acute ischemia.

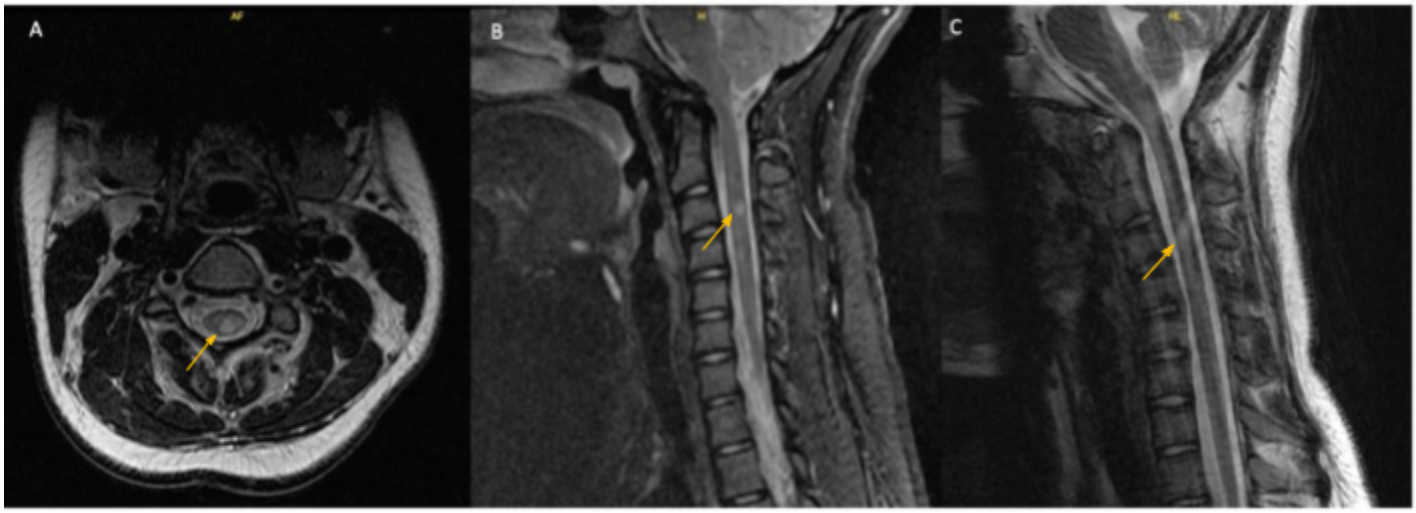


Figure 3*: T2 hyperintense signal with restricted diffusion involving the left side of the cervical spinal cord at the level of C3-C4 on axial (A) and sagittal (B, C) images.*

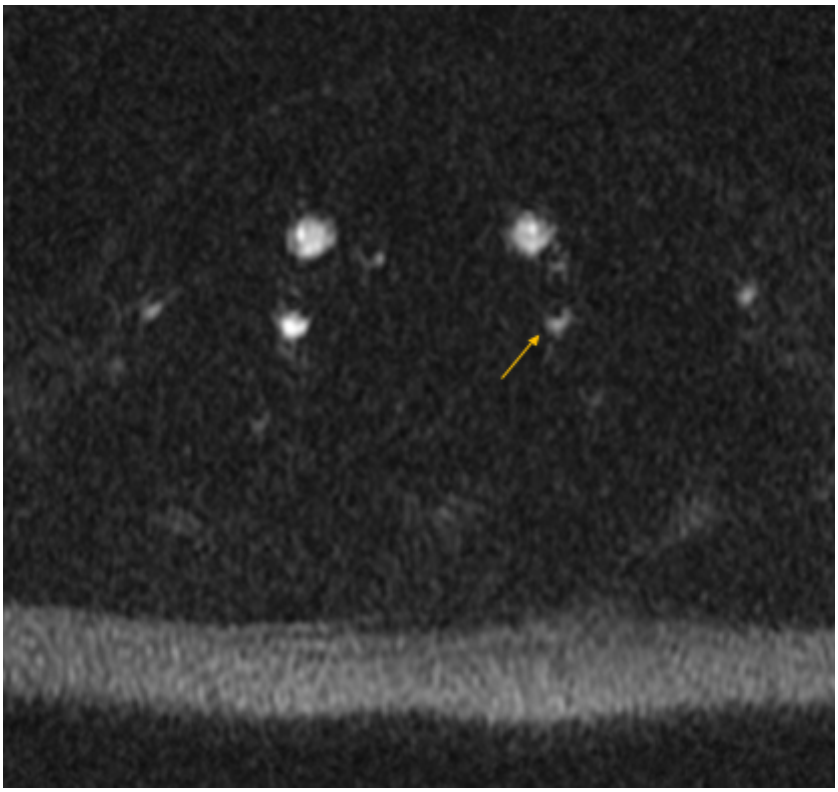


Figure 4: *Initial Magnetic Resonance Angiography (MRA) of the neck with hypointense signal with irregularity of the left vertebral artery*

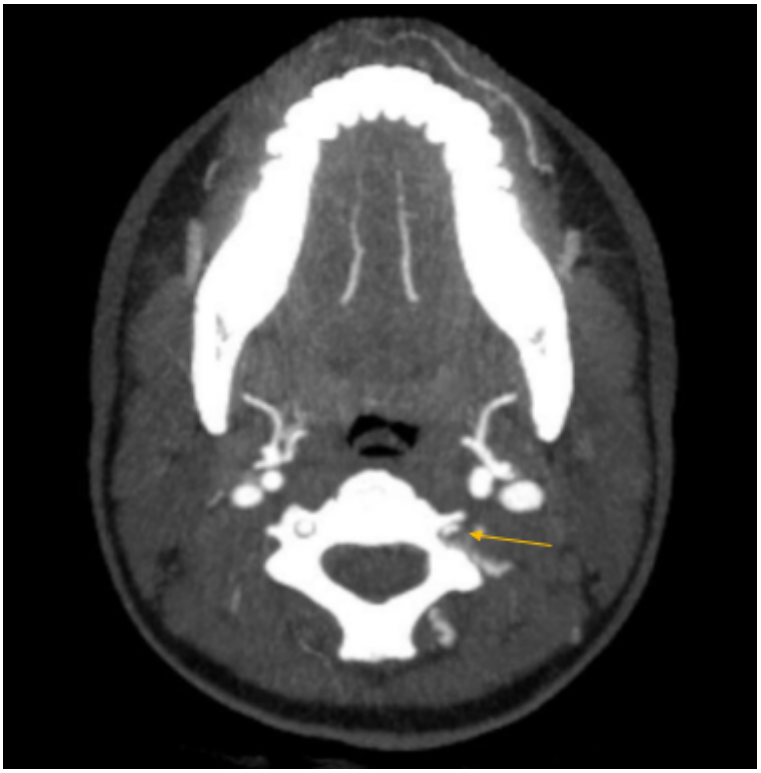


Figure 5: *CTA Neck obtained 2 months later still demonstrating irregular contours at the left V3 segment.*