

Vascular anomaly at the craniocervical junction presenting with sub arachnoid hemorrhage: Dilemma in Imaging Diagnosis, Endovascular Management and Complications

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ABSTRACT

We present a case of a ruptured vertebral artery dissecting aneurysm that mimicked a presumed vascular anomaly by CTA (Computerized Tomographic Angiography). A parenchymal arteriovenous malformation (AVM) or a dural arteriovenous fistula (DAVF) at the craniocervical junction can present with a subarachnoid hemorrhage and cannot be differentiated from a vertebral artery dissection by non invasive imaging. Catheter based cerebral angiography revealed a dissecting pseudoaneurysm of a diminutive right vertebral artery terminating in the posterior inferior cerebellar artery (PICA) that to our knowledge has not been previously reported. NBCA (N-Butyl Cyanoacrylate) embolization of the pseudoaneurysm lumen and sacrifice of the parent vessel resulted in cerebellar infarction, requiring an emergent decompressive craniectomy. The patient recovered to a functional neurologic status.

CASE REPORT

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A 36 year old female presented to the emergency department (ED) with acute onset headache. After receiving a diagnosis of migraine, she was sent home. Her symptoms did not improve. Her headache became profound the following day, and she also experienced right sided paresthesia and shortness of breath. She returned to the ED and a Computerized Tomographic (CT) study of the head and CT-Angiography (CTA) was performed. The head CT revealed acute subarachnoid hemorrhage, intra-ventricular hemorrhage in the fourth ventricle, diffuse cerebral edema and a focal hyperdensity at the craniocervical junction of indeterminate etiology (Fig. 1). The CTA identified a cluster of tortuous vascularity with an aneurysm corresponding to the focal hyperdensity seen on the non contrast head CT study. (Fig. 2a and b).

A preliminary diagnosis of a ruptured aneurysm within an AVM nidus was made and patient was taken to the operating room. Prior to induction of anesthesia, she underwent cardio-respiratory arrest. She was resuscitated and the surgical attempt to devascularize the presumed AVM was cancelled. She remained intubated. A catheter based angiogram was then performed. This identified a dissecting aneurysm of a diminutive right vertebral artery terminating in PICA (Fig. 3).

Under general anesthesia, NBCA (N-butyl cyanoacrylate) (Cordis, Miami, FL) microcatheter embolization of the dissecting aneurysm was performed with sacrifice of the parent vessel. Right common femoral artery access was initially secured with a 7 French sheath. A 5 French Envoy guide catheter (Cordis, Miami FL) was positioned within the right subclavian artery. A Prowler 14 microcatheter (Boston Scientific, Fremont, CA) in tandem with a Transend 14 platinum tip microguide wire (Boston Scientific, Fremont CA) was advanced into the diminutive right vertebral artery through

the guide catheter. Microcatheter angiography confirmed a dissecting aneurysm of the vertebral artery terminating in the posterior inferior cerebella artery. NBCA (Cordis, Miami Florida) (2cc) was injected with fluoroscopically observed obliteration of the pseudo aneurysm (Fig. 4 a and b). The patient awoke to base line neurologic status with reversal of the general anesthesia. Twelve hours after the procedure, the patient was noted to have marked decline in her mental status. A non contrast head CT revealed infarction with mass effect within the right PICA vascular territory (Fig. 5). A decompressive craniotomy was subsequently performed (Fig. 6). The patient recovered to an independent functional status after a period of rehabilitation.

DISCUSSION

Dissecting aneurysms of the vertebro-basilar system account for 3-7% of non traumatic subarachnoid hemorrhage (SAH) (1). A study by Kurata et al demonstrated non-dominant vertebral artery involvement in approximately 27% of cases (2). A dissecting aneurysm of the non-dominant vertebral artery terminating in PICA has not been previously reported.

Dissections of the intradural vertebral artery present most commonly with subarachnoid hemorrhage (SAH) from associated pseudoaneurysms. This as a result of a thinner tunica media and adventitia with fewer elastic fibers compared to the extradural segment (1). Due to a high rate of rupture, 10% at presentation and 58% within 24 hrs after the first attack (2), and a significant improvement of mortality (20% if treated, 50% if untreated), treatment is of significant benefit (1).

CT angiography identifies more intimal flaps, pseudoaneurysms, and high-grade stenoses than MR imaging and MR angiography in the evaluation of vertebral artery dissections (3). The study by Chen et al showed greater CT angiography sensitivity (100%) than specificity (98%) in diagnosing vertebral artery dissection relative to catheter angiography. In their patient cohort of 17 in each category, a single misdiagnosis of a vertebral artery aneurysmal type dissection was made for severe atheromatous disease (4). As the management and natural history of these of these two entities are markedly different, catheter angiography is undoubtedly necessary if any intervention is to be considered.

Catheter-based angiography allows for determination of flow characteristics within a brain vascular anomaly. Shunt vascularity from artery to vein as in a pial AVM or dural arteriovenous shunt can be differentiated from other abnormalities. The absence of such in this case directed the diagnosis to a vertebral artery dissection. The most common finding for vertebral artery dissection by Rabinov et al was a pseudoaneurysm with irregular vessel lumen narrowing proximally (1), as was identified in this case.

The diminutive parent vessel caliber and marked anatomic tortuosity of the dissected segment precluded any other

endovascular method of treatment, such as stent vessel reconstruction or non stent assisted independent coil occlusion. Stent delivery within a vessel requires an adequate caliber proximal and distal to the lesion as well a non tortuous intervening segment. Stent flexibility is additionally required to traverse a lesion with any vessel angularity. There are no intracranial stents available that would conform to the size required by the caliber of the vascularity feeding into and exiting this pseudoaneurysm. Additionally, none are flexible enough to be navigated through the cork screw configuration of the identified dissected segment vascularity. A liquid embolic agent such as NBCA is not limited by anatomic vessel caliber and tortuosity in occluding a pseudoaneurysm, as its delivery is primarily blood flow dependent. A diminutive ruptured pseudoaneurysm with fragile walls is more likely to be at risk for procedural perforation with introduction of coils within the lumen. A liquid embolic agent allows for occlusion of a pseudoaneurysm without placement of any device within the aneurysm lumen proper. The disadvantage of a liquid embolic agent is that there is no control of non target embolization within vessels distal to the lesion.

Better long term clinical outcomes are associated with aneurysm involving one artery where complete thrombosis can be achieved by parent vessel occlusion (5). This however is limited in the context of an isolated vertebral artery terminating in PICA, due to the consequent risk of cerebellar infarction.

TEACHING POINT

Imaging diagnosis of vascular pathologies of the brain may be potentially misleading if CT Angiography is the only method used. If a patient is clinically stable, catheter-based angiography remains the gold standard for definitive characterization of intracranial vascular pathologies and should be considered prior to any intervention.

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FIGURES

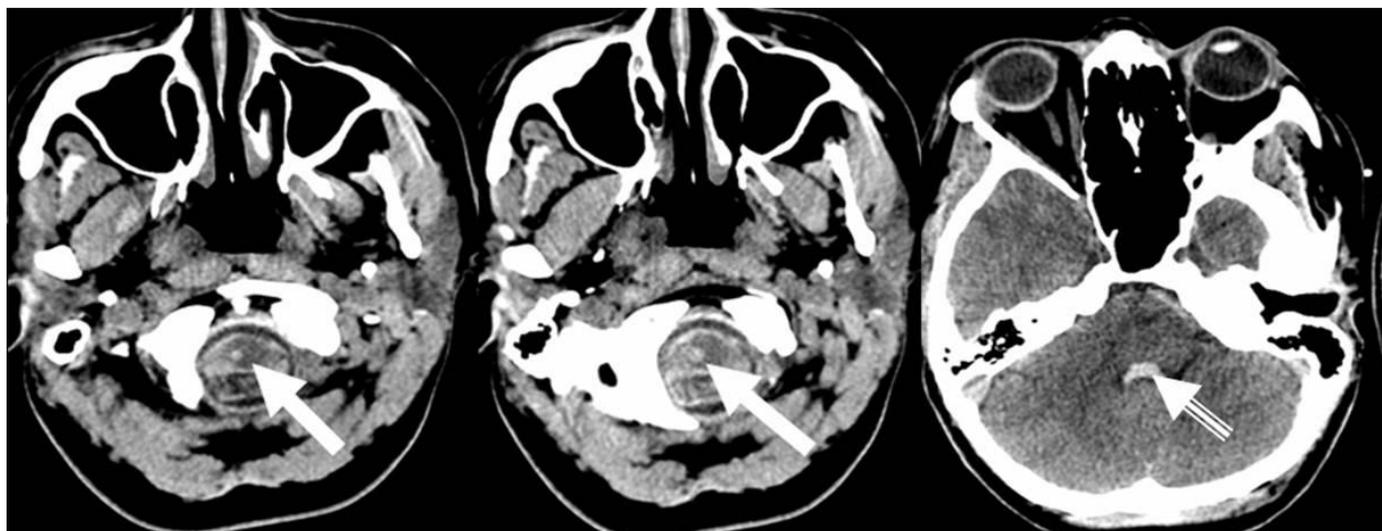


Figure 1: 36 year old female with dissecting pseudoaneurysm of the right vertebral artery terminating in the posterior inferior cerebellar artery (PICA) indistinguishable from a craniocervical vascular anomaly by non invasive imaging. Non contrast head CT selected axial slices through the posterior fossa demonstrate acute subarachnoid hemorrhage with a focal hyperdensity at the craniocervical junction (white arrows). Intra-ventricular hemorrhage is identified within the fourth ventricle (striped white arrow).

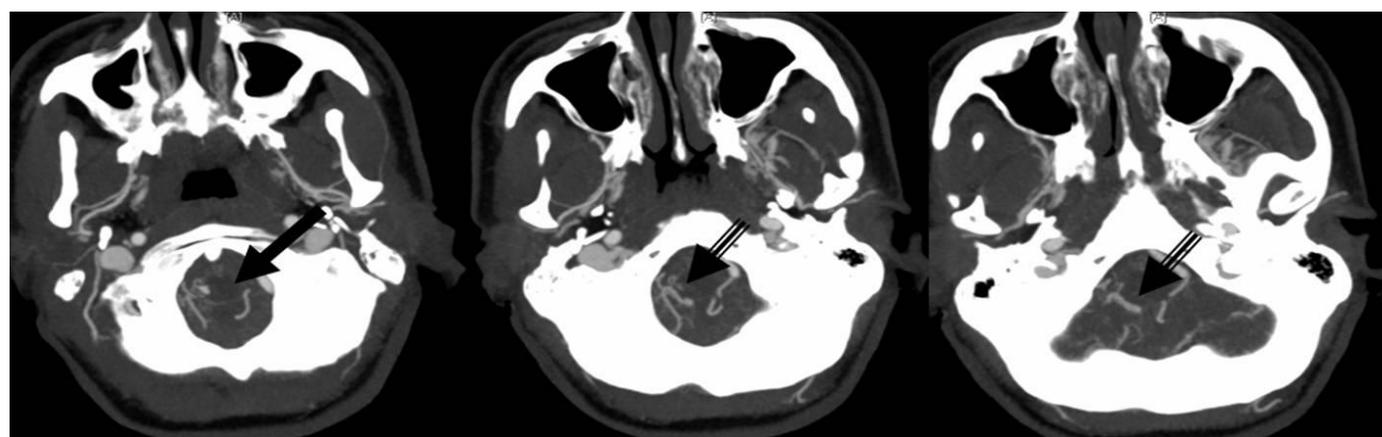


Figure 2a: 36 year old female with dissecting pseudoaneurysm of the right vertebral artery terminating in the posterior inferior cerebellar artery (PICA) indistinguishable from a craniocervical vascular anomaly by non invasive imaging. Selected axial maximum intensity projection contrast enhanced CT angiogram (CTA) of the head images in the arterial phase through the posterior fossa demonstrates a cluster of abnormal vascularity at the craniocervical junction (striped arrows) with a preliminary diagnosis of an intra-nidal aneurysm (solid black arrow).

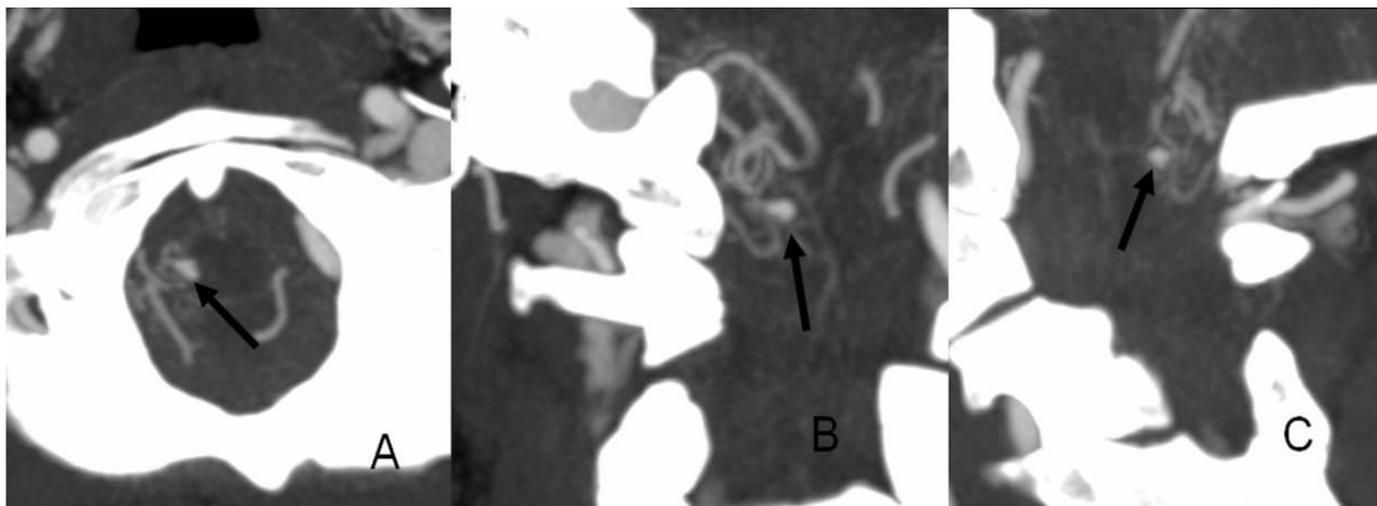


Figure 2b: 36 year old female with dissecting pseudoaneurysm of the right vertebral artery terminating in the posterior inferior cerebellar artery (PICA) indistinguishable from a craniocervical vascular anomaly by non invasive imaging. Selected axial (A), right (B) and left (C) oblique coronal maximum intensity projection contrast enhanced CT angiogram (CTA) of the head imaged in the arterial phase through the craniocervical junction demonstrates what was initially presumed to be an intra-nidal arteriovenous malformation aneurysm (solid arrows).



Figure 3: 36 year old female with dissecting pseudoaneurysm of the right vertebral artery terminating in the posterior inferior cerebellar artery (PICA) indistinguishable from a craniocervical vascular anomaly by non invasive imaging. Right vertebral artery injection diagnostic catheter angiography frontal view during the arterial phase demonstrates an abrupt change in the caliber of distal right vertebral artery with segmental narrowing of the intra-cranial vertebral artery in conjunction with a solitary broad neck dissecting pseudoaneurysm (solid arrow in A). A posterior inferior cerebellar artery is identified (double arrows) without early arteriovenous shunting. Early capillary phase (ring in B) revealing a vertebral artery terminating in the PICA (posterior inferior cerebellar artery) without identification of an early draining vein that is characteristic of an arteriovenous malformation (AVM). Late capillary, early venous phase angiography (C) identifies expected venous drainage from the right PICA cerebellar vascular territory. Concordant venous drainage with the remaining brain venous outflow during the venous phase (D).

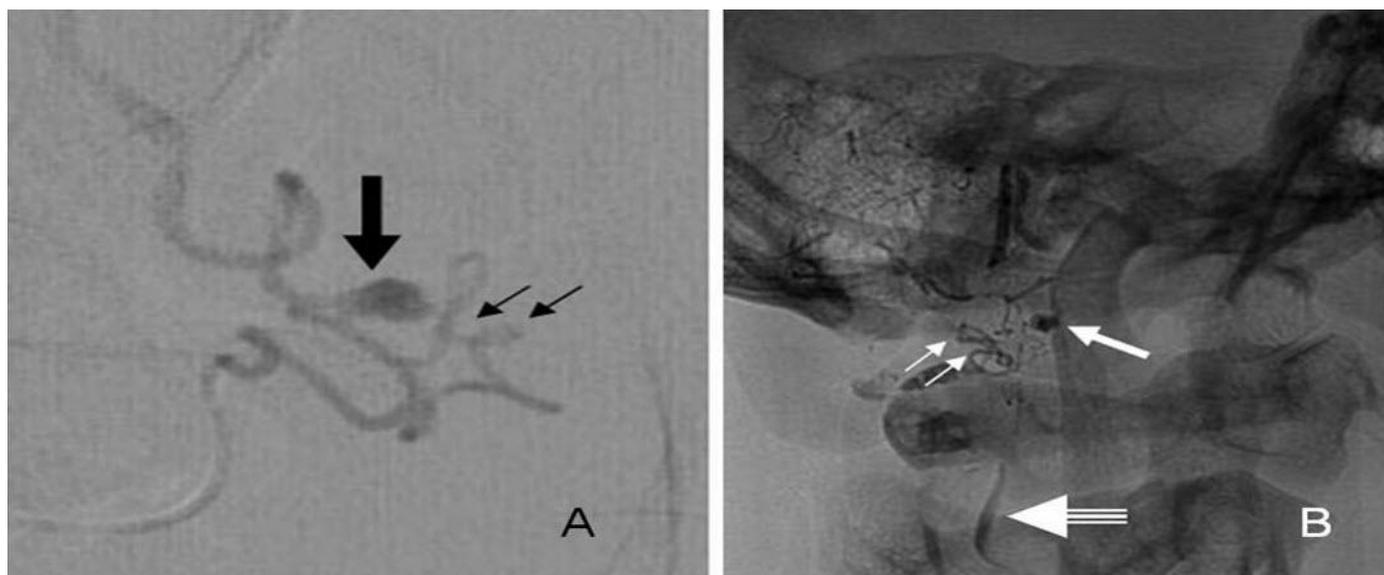


Figure 4: 36 year old female with dissecting pseudoaneurysm of the right vertebral artery terminating in the posterior inferior cerebellar artery (PICA) indistinguishable from a craniocervical vascular by non invasive imaging. Coronal oblique view microcatheter injection confirming a diminutive tortuous intracranial right vertebral artery (double black arrows) terminating in the posterior inferior cerebellar artery (PICA) and a dissecting pseudoaneurysm (solid black arrow) without an AVN nidus or early draining vein (Fig A). Unsubtracted fluoroscopic frontal oblique view (Fig B) demonstrating a NBCA (N-butyl cyanoacrylate) glue cast within the pseudo aneurysm (single white arrow) and distal intracranial vertebral artery (double white arrow). A stagnant column of contrast is seen in the cervical vertebral artery (striped white arrow).

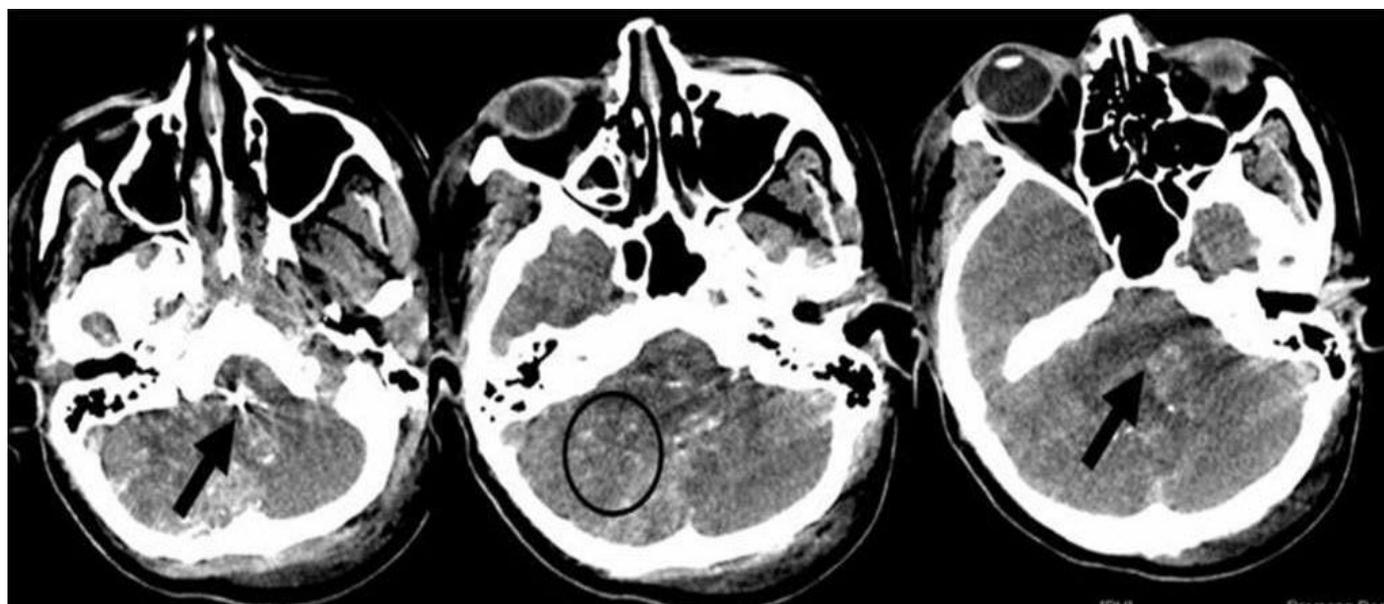


Figure 5: 36 year old female with dissecting pseudoaneurysm of the right vertebral artery terminating in the posterior inferior cerebellar artery (PICA) indistinguishable from a craniocervical vascular anomaly by non invasive imaging. Post-embolization non contrast axial head CT images through the posterior fossa show artifact from the NBCA (N-butyl cyanoacrylate) embolysate (black arrow). Low attenuation from PICA territory infarction (ring) and effacement of the fourth ventricle (black arrow last image).

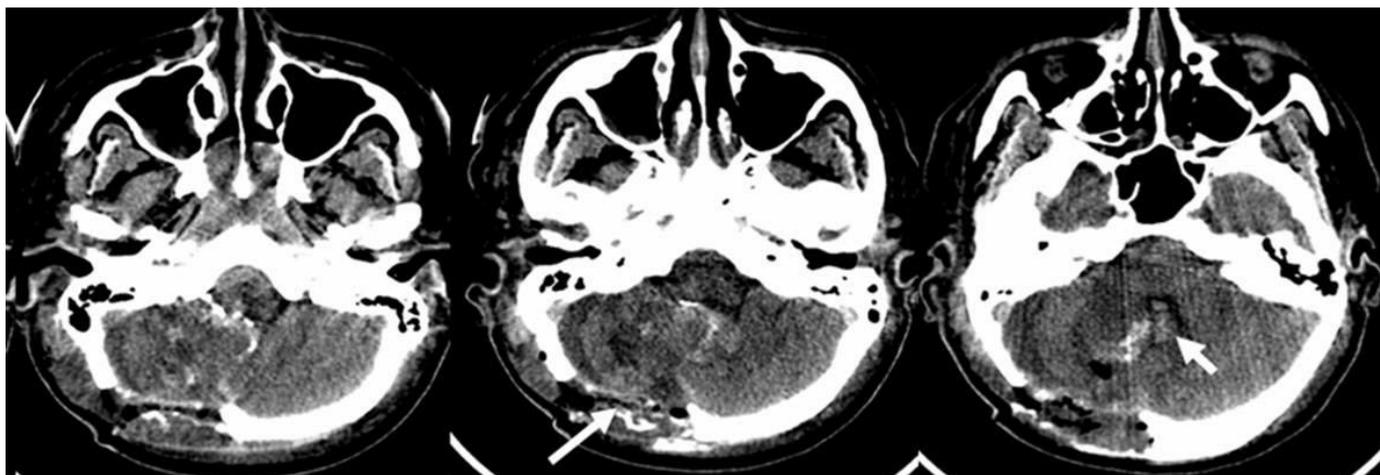


Figure 6: 36 year old female with dissecting pseudoaneurysm of the right vertebral artery terminating in the posterior inferior cerebellar artery (PICA) indistinguishable from a craniocervical vascular anomaly by non invasive imaging. Selected axial non contrast CT head images after decompressive craniectomy (long arrow) and resolved mass effect on the fourth ventricle (short arrow).

ABBREVIATIONS

AVM = Arteriovenous malformation
CTA = Computerized Tomographic Angiography
PICA = Posterior inferior cerebellar artery
NBCA = N-butyl cyanoacrylate

KEYWORDS

Subarachnoid hemorrhage, Vertebral artery dissection, Arteriovenous Malformation, Cyanoacrylate embolization

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