Case Report

Extracranially located PICA aneurysm presenting with supratentorial IVH: A rare event with diagnostic pitfalls

Amey Savardekar, Manoj Kumar Tewari, Rajeev Garg, Vivek Gupta[1], Chirag Ahuja[1]

Department of Neurosurgery, [1]Department of Radiology, PGIMER, Chandigarh, India

ABSTRACT

Extracranially located posterior inferior cerebellar artery (PICA) aneurysms are rare with only 21 cases reported till date. They may arise either from the proximal segment of an extracranially originating PICA or from the tip of its caudal loop when it dips below the foramen magnum. A 16-year-old female presenting with sudden onset severe headache and intraventricular hemorrhage (IVH) in the occipital horns of the lateral ventricle and the fourth ventricles, was diagnosed to have an extracranial proximal segment PICA aneurysm on a four vessel digital subtraction angiography (DSA), after initially missing it on the brain magnetic resonance imaging (MRI) with angiogram (MRA) because of its extracranial location. During surgery, the aneurysm was clipped following a far lateral suboccipital craniectomy with C1-C2 hemilaminectomy. The patient showed good recovery. Thus, we emphasize the need for a dedicated four vessel angiography to diagnose such lesions.

Key words: Extracranial aneurysm, intraventricular hemorrhage, PICA aneurysm

Introduction

Posterior inferior cerebellar artery (PICA) aneurysms comprise 1% of all intracranial aneurysms, which are usually located at the vertebral artery (VA)-PICA junction or just distal to it.[1] The PICA is well known for its varied anatomical origin and course, originating below the foramen magnum in 18% normal arteriograms.[2,3] Extracranially located PICA aneurysms are rare; only 21 cases have been reported till date. We report an unusual case of extracranially originating saccular PICA aneurysm, presenting with hemorrhage into the fourth and lateral ventricles. The patient underwent microsurgical clipping of the aneurysm and showed good recovery. In this report, we highlight the diagnostic pitfalls that may befall clinicians managing such patients.

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Case Report

A 16-year-old female presented with sudden onset severe headache followed by transient loss of consciousness but no neurological deficits. She developed vasospasm on post-ictus day 9 with paraparesis, which improved gradually. Computed Tomography (CT), Magnetic Resonance Imaging (MRI), and Magnetic Resonance Angiography (MRA) of the brain performed at a regional center revealed intraventicular hemorrhage (IVH) in the lateral and fourth ventricles but did not reveal any aneurysm or arteriovenous malformation [Figure 1]. The patient was referred to us after one month with severe headache and improving paraparesis. Repeat CT showed non-communicating hydrocephalus, and a four-vessel digital subtraction angiography (DSA) showed extracranially originating right PICA harbouring a 9.5 × 3.6 × 5.2 mm saccular aneurysm directed posteriorly with a neck measuring 2 mm at the level of C1, which was missed on the initial brain MRI; as the aneurysm was extracranial [Figure 2]. In view of the raised intracranial pressure because of the hydrocephalus, the patient underwent a right ventriculo-peritoneal shunt followed by right far lateral sub-occipital craniectomy with C1-C2 right hemi-laminectomy for clipping the extracranially

Address for correspondence:
Prof. Manoj Kumar Tewari, Department of Neurosurgery, PGIMER, Chandigarh, India. E-mail: manojktewari@gmail.com

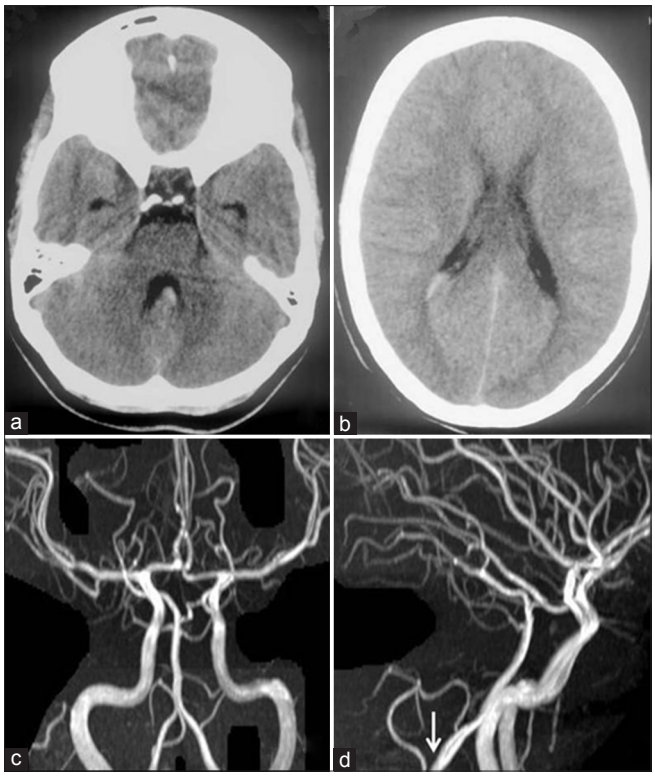

Figure 1: (a and b) Plain axial CT showing presence of supratentorial IVH alongwith fourth ventricle IVH. (c and d) 3D-Time of Flight MR angiography images do not show any obvious vascular pathology. Note the non inclusion of the origin of the right PICA in the MRA slab (arrow) which was noted retrospectively, subsequent to the DSA result

placed PICA aneurysm. The patient recovered without any neurological deficits.

Discussion

Variations in the origin and course of PICA are well documented. PICAs usually arise from the VA, approximately 14-17 mm proximal to the vertebrabasilar junction, but they may be absent (10-15% cases), situated below the foramen magnum (18-35% cases), and may not uncommonly have an extracranial extradural origin from the VA.[4] PICA aneurysms account for 1% of all intracranial aneurysms. Extracranial PICA aneurysms are rare; only 21 cases have been reported till date.[1,5] They arise either from the proximal part (medullary) of the extracranially originating PICA or from the caudal loop (the loop of the tonsillomedullary segment passing near the lower part of the tonsil) when it dips below the foramen magnum.

The etiology of extracranial aneurysms arising from the PICA may involve the tortuous course of the PICA and VA, resulting in hemodynamic stress in the proximal segment (where the PICA branches from the VA) and at the top of the caudal loop. An additional factor that

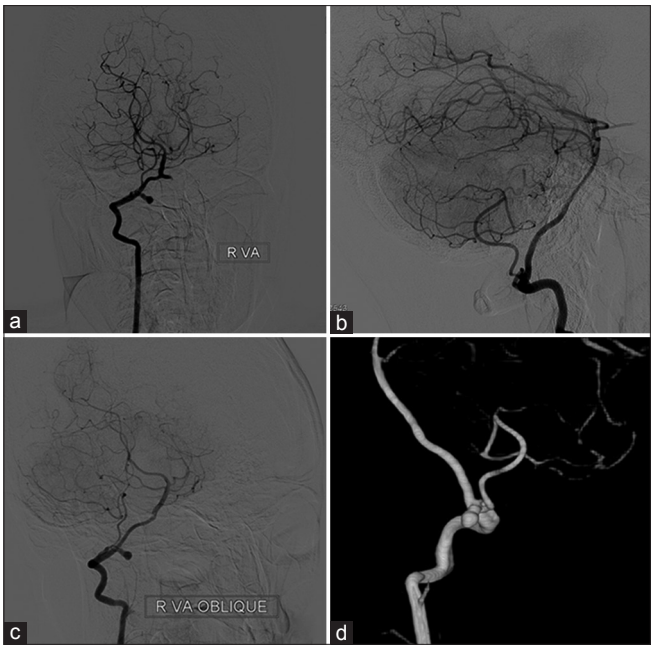

Figure 2: (a, b, c) DSA shows the lobulated tubular aneurysm arising from the right PICA origin directed posteriorly and inferiomedially. (d) Volume rendered reconstruction of the 3D rotational angiography better depicts the aneurysm location and morphology

may play a role is the chronic flexion-extension or rotator stress at the craniovertebral junction, causing an interaction between the C1 lamina and the PICA, which results in vessel wall damage.[5]

Literature reflects a female preponderance (2.2:1) for extracranial PICA aneurysms, with a high occurrence in younger patients.[1] The clinical presentation of ruptured extracranial PICA aneurysms is that of a typical subarachnoid hemorrhage (SAH) with severe headache and nuchal rigidity. Occasionally, it may present with cranial nerve palsies or hemiparesis. The radiological picture usually reveals IVH in the fourth ventricle alongwith SAH in the posterior fossa. It is rare to have IVH in the lateral ventricles; only one case has been reported till date.[6]

In our case, a brain MRI with MRA failed to detect the aneurysm because of its extracranial location[Figure 1] as the vertebral artery was not imaged below the foramen magnum. Also, the blood in the lateral ventricles acted as a decoy and prevented further investigations at the regional center once the MRA was negative. Our patient had an extracranial PICA aneurysm directed posteriorly and medially at the level of the first cervical vertebra, which must have ruptured into the cistern magna and the spurt was hence carried from the fourth ventricles into the supratentorial ventricular system. The initial CT performed on post-ictus day 6 shows blood only in the fourth ventricle and the occipital horns of the lateral

ventricles where the clot would have settled after 5 days of rupture. Thus, hemorrhage was not observed in the cistern magna because of CSF circulation, which washed off the blood. Extracranial PICA aneurysms presenting with isolated IVH have been reported and this should be borne in mind while investigating such patients.

As pointed out by Dammers *et al*., extracranial PICA aneursyms should definitively be ruled out in case of ictus with IVH (especially in the fourth ventricle) with a four vessel angiography (CT or DSA).[7] Such aneurysms have been missed even on three vessel angiograms, thus further reiterating our stand that a four vessel angiography is a must.[7,8] If MR or CT angiography is used primarily to evaluate these patients, it is essential to image them upwards from the aortic arch to avoid missing an extracranial PICA aneurysm. This maneuver will also aid in the planning neuro-vascular interventional procedures.

For proximal segment PICA aneurysms, an inferiorly extended standard paramedian suboccipital approach with drilling of the superomedial part of the condyle has been suggested.[9] We performed a far lateral suboccipital craniectomy along with hemilaminectomy at the C1 and C2, levels to delineate the course of the extracranial VA for proximal control for the aneurysm, if required.

Conclusions

Although, till date, only 22 cases (including our case) of extracranial PICA aneurysms have been reported, it should be noted that in 25% of these cases (including our case), clinicians missed such an extracranial aneurysm during the initial diagnosis, either because a four vessel DSA was not undertaken or the upper cervical region was not included when CT or MR angiography was performed. This emphasizes the need for clinicians to keep this rare possibility in mindto avoid any diagnostic pitfalllike the one encountered in our caseand ask for a dedicated four vessel angiography when dealing with patients presenting with an ictus and fourth intra-ventricular bleeding, even when supratentorial IVH exists.

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