Case Report

Congenital Agenesis of Both Vertebro-Basilar Artery-A Rare Case Report

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Abstract:
Vertebral and basilar arteries are the major blood suppliers of the brain. They supply nearly one-third of the brain. Congenital absence of these arteries is a rare condition. Usually, this condition presents with stroke like presentation. This is a case presentation of a 40-year-old lady who was diagnosed as congenital agenesis of vertebra-basilar arteries. This lady was presented with intraventricular hemorrhage. Subsequently the diagnosis was made by cerebral angiogram.

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Introduction:

Common carotid, internal carotid, and external carotid arteries supply blood to the head and neck. Other arteries specially the vertebral artery (VA) which is the branch of the subclavian artery that supply about 30% blood to the brain. It is the chief branch of the subclavian artery. The vertebral artery arises from the postero-superior aspect of its first part. The spinal cord (upper part) and its membranes, the brain stem, cerebellum, occipital and temporal lobe of the cerebrum, muscles of the neck and inner ear are mainly supplied by the vertebral artery.[1]

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Normally the VA is 3-5 mm in diameter.[2] Both sided vertebral arteries are fuse and form basilar artery and that is divide and form right and left posterior cerebral arteries.[3] The bilateral vertebral arteries (VA) and an unpaired basilar artery (BA) are unite and comprises vertebrobasilar system. The vertebral artery branches into four segments with its course.[4] Basilar artery formation is unique in our body system that provides significant part in the posterior circulation.[5] The agenesis of bilateral vertebrobasilar junction is an exceptional variation anatomically. The developmental mechanism in volve the anterior radicular artery of Cervical 1 (C1). It is the branch of proatlantal artery. It normally forms the adult VA distally and the BA proximally. If there is lack of connection of the basilar artery with posterior cerebral artery then may develop to the vertebrobasilar agenesis. And it result antegrade flow within the BA. Possibly the absence of the proximal and distal connections of the BA might cause the similar result that's mechanism unknown.[6] The final part of 1 vertebral artery (VA) aplasia or hypoplasia is a common anatomical variation and in this case, there is a

small connection of vertebral artery with basilar artery or it continues as the posterior inferior cerebellar artery .[6]Agenesis of both VAs precede to synchronous hemispheric and vertebrobasilar ischemic cerebrovascular disease. [7]The aplasia of bilateral VA is common in male than female. It most likely depends on genetic factors and diseases are obvioused usually over 30 years of age.[8] Rete mirabile means ("wonderful net") in Latin. Vertebrobasilar arteries agenesis with rete formation is unusual. Bilateral segmental agenesis of carotid and vertebrobasilar arteries with rete formation have been seen in 10 cases.[9] Initial symptoms of bilateral Vertebrobasilar artery agenesis are different and unspecified. Primary symptoms are vertigo, vomiting, headaches, dizziness diplopia, blindness, ataxia, balance problem, and weakness in both sides of the body. The angiography is a good method for diagnosis.

Case Report:

This 40-year-old hypertensive lady was admitted to the Neurosurgery department of Sylhet Women's Medical College Hospital with the complaints of severe headache and altered conscious level for 2 days. During admission her Glasgow Coma Scale (GCS) was E3,M6,V4 (13/15). Her pupils were bilateral equal and reacting to light. She had right sided limbs weakness. Right upper limb muscle powers were 3/5 and right lower limb muscle powers were 2/5. Her blood pressure was 180/120 mm of Hg and heart rate was 86/minute. Her CT scan of head revealed intraventricular hematoma (mostly in left lateral ventricle) with minimum mass effect. Cerebral CT angiogram showed complete absence of basilar artery both vertebral arteries. Both posterior cerebral arteries were supplied by both internal carotid arteries. Her raised blood pressure was controlled by adding tablet Olmesartan 20 mg twice a day. Her intracerebral hemorrhage was treated conservatively. Repeat CT scan of brain (after 2 weeks) showed the hematoma was in resolving state. Her conscious level was improved to GCS 15/15 within 2 weeks. She was discharged from the hospital after 15 days. During follow up after 2 months her right sided limbs weakness improved to 4/5. She had a good control of bladder and bowel at that time. She never experienced any headache after getting discharged from hospital.

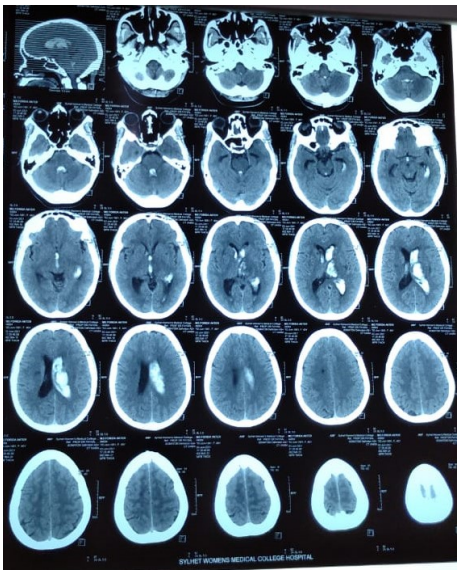

Figure 1: CT scan of brain shows intraventricular hemorrhage.

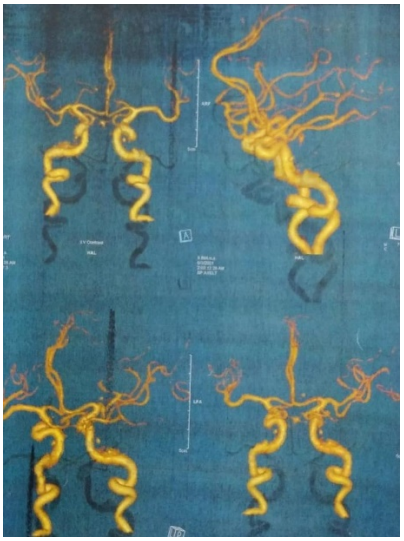

Figure 2: Cerebral CT angiogram shows complete agenesis of Vertebral and Basilar arteries.

Discussion:

I.M. Burger et al. presented 3 cases.[6]The first case was a 69 year old lady who had history of hypertension, migraine and premature birth was observed for a right sided tinnitus that was pulsatile. After doing magnetic resonance imaging and angiography, there was no connection between the vertebral artery and the basilar artery. The VA of the right side terminated as a branch of the occipital artery and the left sided VA continued as the right PICA. The BA was separated from its normal blood sourcesbecause of no connection within both PCA andVA and was supplied by the left persistent trigeminal artery only.

Second case was a 65 year old gentleman who was observed for Wegener granulomatosis and right sided deafness. Magnetic resonance imaging and angiography, which reported on either sides, there was continuation of the VA as the PICA and no connection to the BA. The right sided posterior communicating artery (PcomA) that was separated from the PCA which was providing the BA and the left sided PCA arised from the left ICA (terminal part) .Due to no connection within both sided PCAs and VAs, the BA was separated from its normal blood sources and was provided only by the right sided PcomA.

The third case was a 51 year old gentleman with history of coronary disease, hypertension, hyperlipidemia, and smoking and 1 year history of episodic dizziness with a throbbing pain in both ears, aggravated by rotation of the head to the right or left. Magnetic resonance imaging and angiography revealed little seeing of the BA and VA but DSA showed the extracranial part of both sided VA that caliber was small. The vermian branch of the right sided PICA was originated from the continuation of the right sided VA, and the hemispheric branch originated from the right sided AICA. There was connection of the proximal part of BA with the small and irregular segment of the V4 of left sided VA which revealed opacification forward moving up to the origin of both sided AICAs. The vermian and hemispheric branches of the left sided PICA originated from the V4 segmentof left VA. Through a large PcomA, the leftsided ICA contributed to the BA which itself continued as the left sided PCA.

Fong Y. Tsai et al presented a 36 year old male got admitted with history of left frontotemporal headache and numbness of the right face and body with right sided blurring of vision.[10]He had history of episodes of thrombophlebitis of the right leg and a right saphenous vein ligation was performed. After performing transfemoral carotid and arch angiograms which showed absence of both sided VAs with occipital-basilar anastomosis. The right common carotid arteriogram revealed negative but the left common carotid arteriogram revealed a infarction in the branches of left sided posterior cerebral artery with collaterals. During hospitalization he developed thrombophiebitis which was medicated with heparin and oral anticoagulant agents.

Patel D M et al presented two unusual cases about blood supply of posterior cranial fossa.[11]The first case was a 52 year old right handed lady got admitted with symptoms of dizziness and neck pain for several months that had progressed. MRI and MRA showed a right internal carotid artery stenos is, and an end arterectomy was performed. MRA also indicated posterior circulation was severely hypoplastic and there was atresia of theboth sided vertebral arteries and terminated as the posterior inferior cerebellar arteries. Three-dimensional (3D) angiography revealed atresia of bilateral VAs with lack of a BA. Both sided fetal posterior cerebral arteries ensured circulation posteriorly with vertebral arteries which terminating into right and left sided PICA with anterior vertebrobasilar system that extends cephalad to unite with the right PComAs.

The second case was a 69 year old, chain smoker lady presented with short term memory loss and he found that she had aneurysm of right sided cavernous ICA. She has history of raised blood pressure, hyperlipidaemia, peripheral vascular disease, incontinence of urine, and stents kept in left sided arteries of lower extremity. Cerebral CT angiogram including neck revealed a petrous carotid artery aneurysm (3 x 2 mm) with a persistent right trigeminal artery. Mainly, posteriorly circulation was provided through a right sided fetal posterior cerebral artery. The 3D angiogram (arteriogram) showed atresia of the bilateral vertebral arteries and absence of a basilar artery. There was anterior vertebrobasilar system that extends cephalad to unite with the right PComAs.

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