A Case Report

Incidentally Found Carotid Artery Hypoplasia in Adult Patient

Tanya Sharma[1], Vaibhav Yadav[2], Shivam Sood [3], Mugdha Subhash Mozarkar[4]

[1,]**Dept. of Radio-Diagnosis, MMIMSR Mullana, Ambala, Haryana India 133207**

tanyasharma1593@gmail.com ***Phone number; 7018570031***

[2,]**Dept. of Radio-Diagnosis, MMIMSR Mullana, Ambala, Haryana India 133207**

yadav.vaibhav94@gmail.com ***Phone number; 9971330077***

[3,]**Dept. of Radio-Diagnosis, MMIMSR Mullana, Ambala, Haryana India 133207**

shivamsood97@gmail.com ***Phone number; 8077059568***

[4,]**Dept. of Radio-Diagnosis, MMIMSR Mullana, Ambala, Haryana India 133207**

mugdhamozarkar@gmail.com ***Phone number; 8055955155***

***Corresponding author**

Tanya Sharma *Department of Diagnostic Radiology, MMIMSR, Mullana, Ambala, Haryana, India

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Abstract:

Background: Carotid artery hypoplasia is a rare congenital anomaly, and there are controversies in the leading cause for it. We presented herein an asymptomatic case of left carotid artery hypoplasia found on Carotid artery Doppler , magnetic resonance imaging (MRI) during routine checkup followed by further evaluation by CT angiography . Through this paper, we highlight that the recognition of this anomaly is very important when planning for carotid endarterectomy or trans sphenoidal hypophyseal surgery and in case of an acute brain attack from thromboembolic causes.

Key-words:Internal Carotid artery, Congenital anomalies, Carotid artery Doppler, Magnetic resonance imaging, CT angiography .Hypoplasia.

INTRODUCTION

ICA is one of the most stable arteries in human body, whose hypoplasia is considered very rare. The frequency of Carotid Artery Hypoplasia is not very clear and most of these cases are identified by modalities like ultrasonography, computed tomography (CT) and MRI. [(9)]

Conditions like agenesis, aplasia or hypoplasia of the ICA are rare congenital anomalies occurring in less than 0.01% of population[(1,2)]

“Absence” is referred to a spectrum of developmental abnormalities that include agenesis, aplasia, and hypoplasia of the ICA . Aplasia ˇand agenesis represent total absence of the artery while the term hypoplasia is characterised by narrowing of ICA along its entire course due to incomplete development. In such cases, the anterior cerebral artery (ACA) and middle cerebral artery (MCA) at the side of the absence are usually supplied through the circle of Willis by the contralateral ICA or posterior circulation through the posterior communicating artery (PCOM)[(2)] .

Agenesis of ICA is due to total nondevelopment of ICA, whereas aplasia and hypoplasia of ICA are due to nondevelopment or incomplete development of the ICA respectively , despite the presence of embryonic precursor of the vessel[(1,2,7)].The exact mechanisms of Carotid Artery Hypoplasia remain

unknown. Agenesis or aplasia represent total absence of the artery, unlike hypoplasia, which is characterised by diffuse narrowing of the ICA along its entire course. Unilateral abnormalities of ICA are distinctly more common than bilateral ICA agenesis or hypoplasia [(2).]

Patients are usually asymptomatic but a some of them may present with headache[(3),] transient ischemic attack (TIA)[(2),] Horner's syndrome[(4,5)], or hormonal dysfunction[(6),]. We reported a case of left carotid artery hypoplasia in an asymptomatic patient who was diagnosed on Carotid artery Doppler , CT Angiography, magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) during routine checkup.

CASE REPORT

A 30-year-old hypertensive woman with complaints of headache from 4 days during routine checkup underwent carotid artery's Doppler. She denied any abnormal neurological symptoms. Diagnostic findings on colour-coded carotid duplex imaging include a long segmental small-caliber lumen of left ICA with markedly decreased flow (13% flow volume) in the affected internal carotid artery relative to the contralateral side but without intraluminal lesions. Indirect findings included markedly increased total flow volume in left vertebral artery and a reduced vessel diameter with increased flow resistance in the ipsilateral common carotid artery.

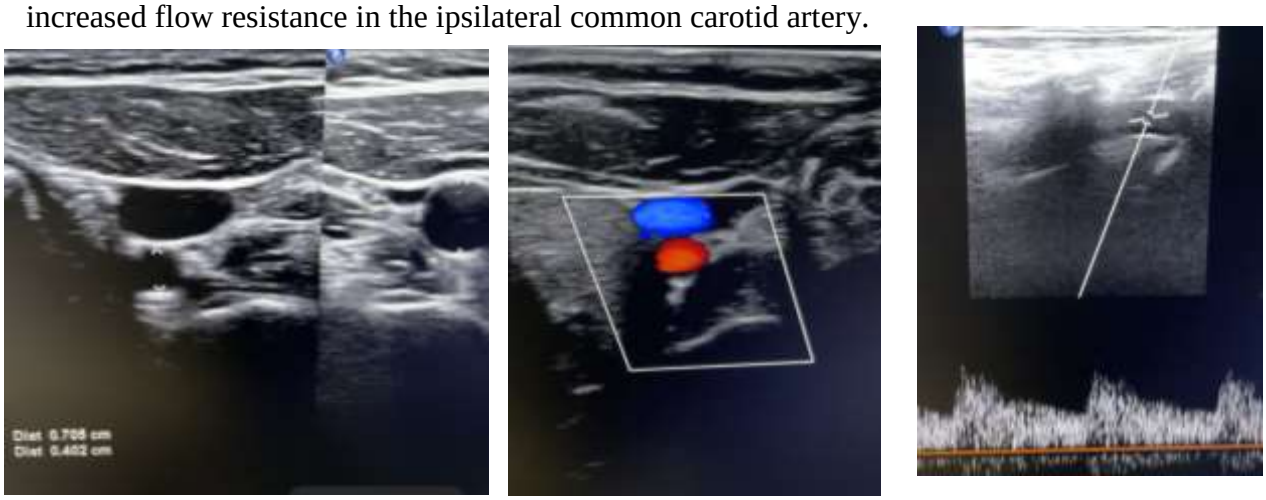


Figure 1: Images (A), (B) and (C)

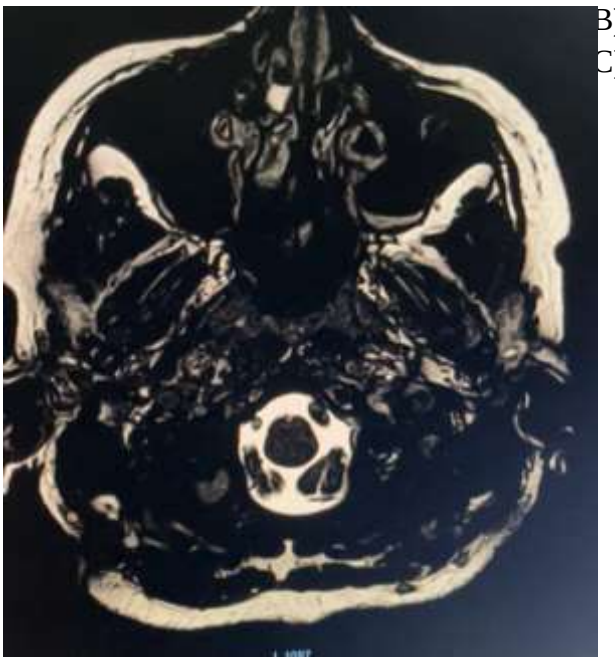
Figure 1: Hypoplasia of the left ICA. (A) B-mode ultrasound image shows small-caliber lumen of left ICA) in the affected internal carotid artery relative to the contralateral side but without intraluminal lesions.(B) colour Doppler image shows diffuse small size of the left CCA .(C) dilated left vertebral artery.\

Indirect findings included markedly increased total flow volume (an increase of 133%) in left vertebral arteries and a reduced vessel diameter TOF MRA of the neck shows diffusely diminished flow-related enhancement within the left ICA (arrow) and MRI and MRA of the brain was performed which showed narrowing of distal cervical and petrous parts of right ICA as compared to the opposite side. The flow void in distal segments of ICA i.e., cavernous and supra-clinoid ICA and left Al segment we're not visualised. Distal left ACA flow void was normal. CECT angiography was further advised . On CECT-carotid angiography , in the expected location of the internal carotid artery a very narrow branch arising from the common carotid artery extending cranially into the narrowed carotid canal and continuing into the cavernous segment on left side was seen suggesting hypoplastic left internal carotid artery. Severely hypoplastic left A1 segment of

ACA and normally opacified A2 segment of left ACA arising from the common trunk was also noted. Prominent left PCOM which seems to supply the left MCA and prominent left vertebral artery were other associated findings .

Figure 2: Images (A), (B) and (C) and (D)

(A)



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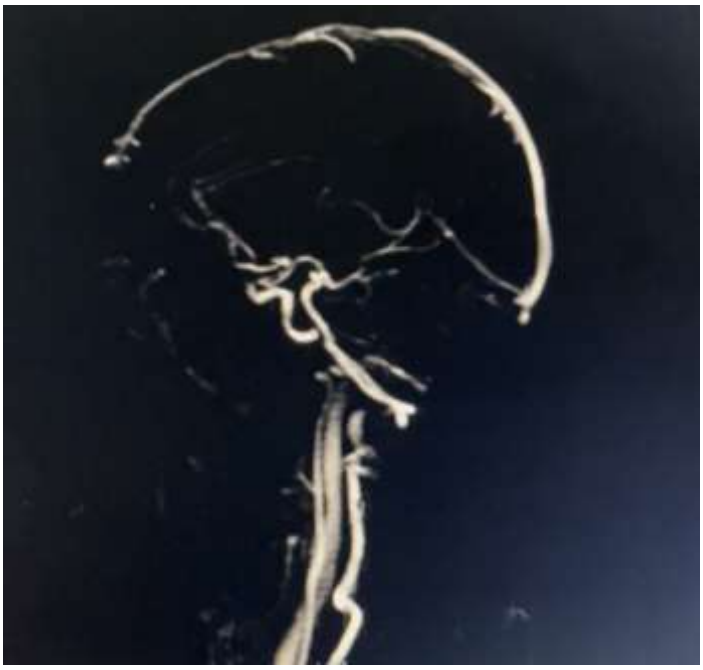
(B)



(C)

(D)

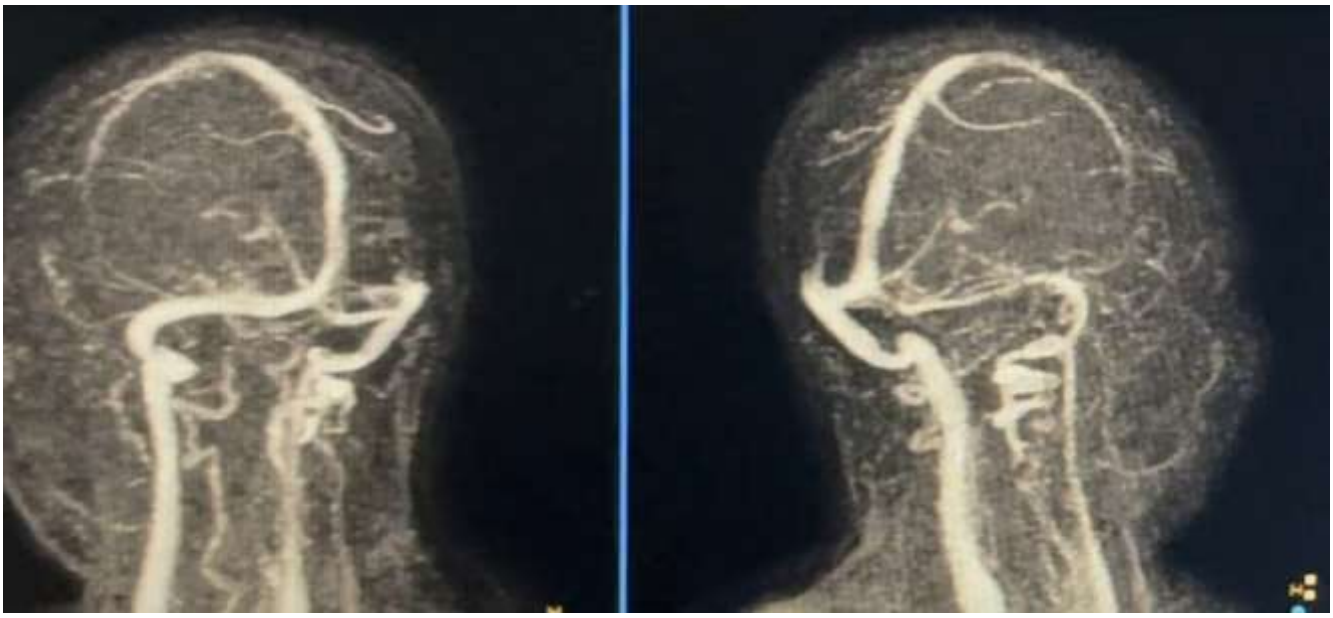
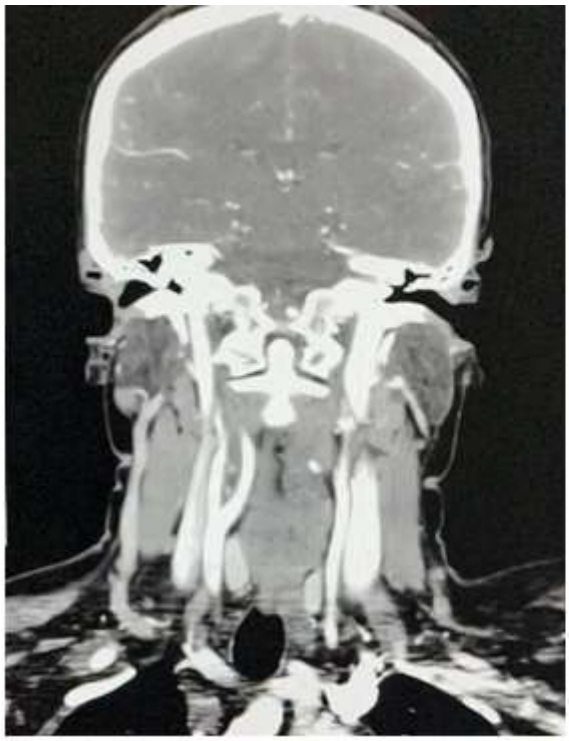
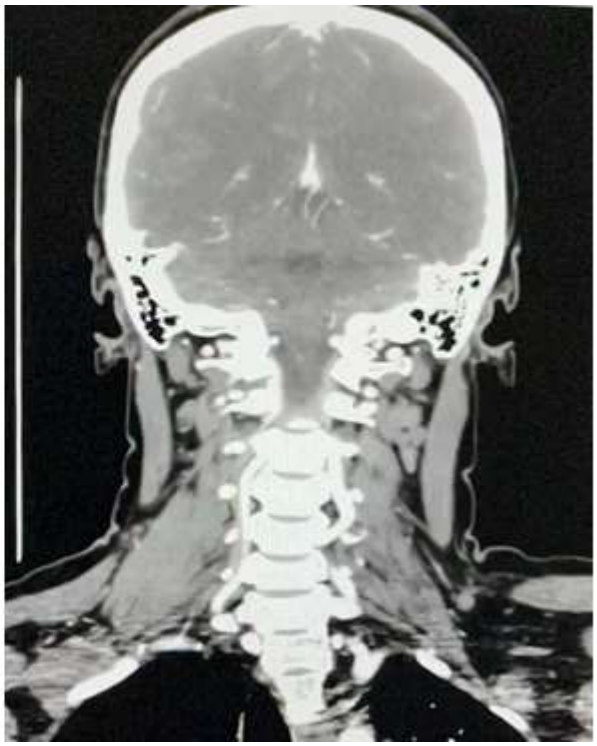


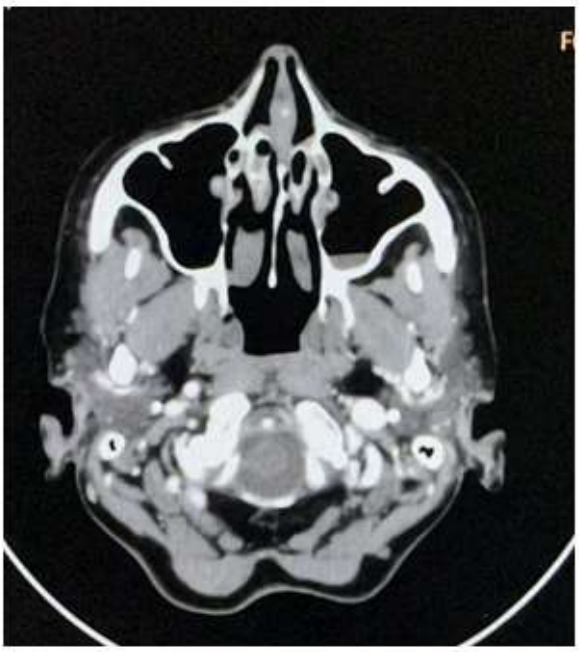
Figure 2 : Hypoplasia of the left ICA. (A) Axial B_FFE image at the level of the petrous ICA shows hypoplasia of the left carotid canal .(B), (C) & (D) Source image from a 3D time-of-flight (TOF) MRA shows diminished flow-related enhancement within the petrous part of the left ICA and diffuse small size of the left CCA .



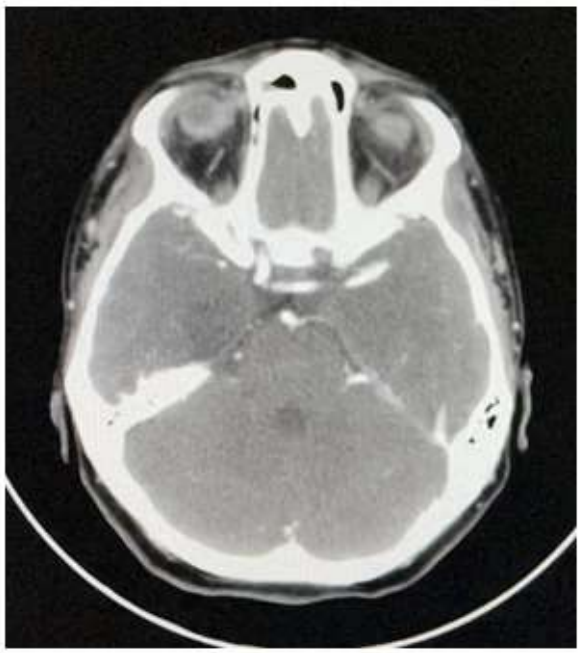
(A)



(B)



(C)



(D)

Figure 3: (A & B) Coronal CT angiogram of the supra-aortic arteries revealed a thinner left common carotid artery (CA) with hypoplastic left internal carotid artery. (C) Axial Brain CT scan revealed a hypoplastic carotid canal on the left side. (D) Axial Brain CT scan revealed a hypoplastic left internal carotid artery .

DISCUSSION

Most cases were identified by ultrasonography, computed tomography (CT) or MRI. The exact mechanisms of these developmental anomalies remain unknown. Therefore, agenesis and aplasia represent total absence of the artery, unlike hypoplasia, which is characterised by diffuse ICA narrowing along its entire course. Unilateral ICA abnormalities are distinctly more common than bilateral ICA agenesis or hypoplasia[2] .

In a study conducted by Tran-Dinh et al, hypoplasia of the left ICA was more often observed, with incidence around 1.5 times that of right ICA hypoplasia .(9) The case in this report also had left ICA hypoplasia.

Most cases of congenital internal carotid artery hyperplasia (CICAH) can be completely asymptomatic as seen in case of our patients . This is because collateral circulation, such as the Willis 'circle, is well developed.[9] Compensations of ICA circulation in CICAH patients was first described by Lie et al and was categorised into 6 types .[10] Type A: in absence of unilateral ICA, the ACA compensates to the ipsilateral ACA and the enlarged PCA to the ipsilateral MCA. Type B: the ipsilateral ACA and MCA are supplied by the ACA. Type C: in case of hypoplasia of bilateral ICA, the anterior circulation of ICA blood supply is compensated by the carotid-vertebrobasilar artery anastomosis of the basilar artery. Type D: blood is supplied to the ipsilateral carotid siphon from the cavernous sinus anastomosis in case of unilateral hypoplasia of ICA,. Type E: the small ACA is supplied by bilateral hypoplasia of ICA and the MCA is supplied by an expanded PCA. Type F: distal collateral circulation is provided through ECA , internal maxillary artery and skull base anastomosis, which is, the microvascular network of the skull base.

In this case, the patient's left PCOM was prominent as compared to right side. Left MCA was normal in caliber and showed normal contrast opacification and seemed to be opacified by prominent left PCOM, which was similar to the type A mentioned above. The patient in our study was well compensated and did not have any clinical symptom .

However, some abnormal symptoms have been reported to be related to CICAH. In general population, the incidence of cerebral vascular aneurysm is about 2% to 4%, but among the CICAH population the incidence rate of intracranial aneurysm raises to about 27.8%.The pathogenesis of aneurysm in CICAH is not well understood, though some studies suggest it might be related to internal carotid agenesis or hypoplasia. Patient with internal carotid hypoplasia is usually accompanied with other intracranial vascular variation, which causes increases in the intravascular blood flow volume and velocity, causing hemodynamic changes, and thus increasing the incidence of intracranial aneurysm . some CICAH patient can also present as recurrent headache, blurred vision, recurrent transient ischemic attack, cranial nerve paralysis, paresis, and Horner's syndrome. However, some rare symptoms have been reported to be related to CICAH, such as posterior fossa malformation, Goldenhar syndrome, arterial lesions, aortic coarctation, PHACE Syndrome and Klippel-Feil syndrome(9) .

Agenesis of ICA may be misdiagnosed as ICA occlusion. To distinguish ICA agenesis from acquired ICA occlusion, computed tomography (CT) scan of the skull base should be performed focusing on the ipsilateral carotid canal(10). This is the carotid canal normally develops in the setting of presence of embryonic ICA at five to six weeks of gestation. Therefore, demonstration of an absent or small carotid canal indicates a congenital absence, and helps differentiate from acquired causes of ICA occlusion or narrowing, such as chronic dissection, severe atherosclerosis, or fibromuscular dysplasia[2,10,11].

Finally, failure to recognise the inter-cavernous collateral pathway can have serious complications during transsphenoidal hypophyseal surgery.

CONCLUSION

Agenesis, aplasia, and hypoplasia of the ICA are rare congenital anomalies. The major collateral pathways include the circle of Willis, persistence of embryonic vessels, anastomosis between ECA and ICA, and intercavernous anastomosis. Although many cases are asymptomatic and incidentally detected, recognition of this anomaly is important in the setting of thromboembolic disease, during planned carotid endarterectomy or transsphenoidal hypophyseal surgery, and detection of associated cerebral aneurysms. Secondly , CT scan of the skull base should be done to identify the hypoplastic carotid canal that helps differentiate ICA agenesis or hypoplasia from acquired causes of carotid occlusion or stenosis.

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