

Case Report

Hypoplastic internal carotid artery, renal artery stenosis, vertebral fusion anomaly: rare association-a case report

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ABSTRACT

Hypoplastic internal carotid artery has been reported in about 100 patients, most being incidentally diagnosed. Association with other anomalies rarely described. Authors report 6-year-old male presenting failure to gain weight and precordial bulge from past 6 months, past history was significant-diagnosed to have right dysplastic kidney at 8 months age. Detailed investigations revealed left hypoplastic internal carotid artery, vertebral segment anomaly, right dysplastic kidney due to right renal artery stenosis. Child was treated medically and was clinically better at latest follow up. All cases with dysplastic kidney need to be searched for vertebral and carotid anomalies, left ventricular dysfunction.

Keywords: Dysplastic kidney, Internal carotid hypoplasia, Left ventricular dysfunction, Renal artery stenosis, Vertebral segment anomaly

INTRODUCTION

Hypoplasia of the internal carotid artery (HICA) is a rare congenital anomaly, prevalence of HICA (including agenesis and aplasia) is estimated to about 0.01%.¹ Hypoplasia of internal carotid artery is distinguished from aplasia by the presence of patent but reduced caliber ICA. Aplasia is referred when the ICA and the carotid channel are completely absent.² The mechanism of development of HICA remains unclear. Some authors have suggested secondary regression of ICA following a phase of normal development, whereas others consider it to represent arrest of the development of ICA.³ HICA is usually asymptomatic due to the development of collateral vessels. The collateral circulation may be in the form of a transcranial anastomosis with external carotid artery with persistent embryonic arteries or with anastomotic channels in the circle of Willis.⁴

Uniform narrowing of the luminal diameter of the ICA may not only result from developmental hypoplasia but

also from a variety of acquired conditions such as atherosclerosis, tubular fibromuscular dysplasia, moyamoya disease, dissection and arteritis. HICA must be differentiated from this acquired ICA stenosis because acquired stenosis could require proper treatment such as endarterectomy or anti-coagulotherapy. In congenital hypoplasia, the entire ICA caliber is too small and there is no thickening of the vessel wall. On the contrary, in most acquired diseases, the external caliber of the ICA is normal and internal luminal narrowing is due to wall thickening.⁵

Other important radiologic finding is carotid channel hypoplasia. The carotid canal closely linked to the development of the ICA during embryonic life. The skull base develops between the 5th and 6th weeks of embryonic life and is associated with ICA development. The presence of a developing ICA is essential for the formation of the carotid canal.⁶ Thus the carotid canal is absent in ICA agenesis, small and not well developed in

cases of hypoplasia, as in our cases. In acquired stenosis of the ICA, the carotid canal is normal in size.

The ICAs are derived from portions of the first and third aortic arches and paired dorsal aorta, when the embryo has attained the 3-mm stage.⁷ The roots of the ICAs are formed from the third aortic arches. The dorsal aorta forms the intermediate portions of the ICAs and the distal part of the ICAs are formed from the first aortic arches. Hypoplasia of the ICA depends on the abnormal regression or involution of the first and third aortic arches and dorsal aorta in the 20-24 mm embryonic stage.²

Block vertebrae occur when there is improper segmentation of the vertebrae, leading to parts of or the entire vertebrae being fused. The adjacent vertebrae fuse through their intervertebral discs and also through other intervertebral joints so that it can lead to blocking or stretching of the exiting nerve roots from that segment. It may lead to increased stress on the inferior and the superior intervertebral joints and neurological problems.

CASE REPORT

A 6 years old boy presented with not gaining weight and precordial bulge noticed from 6 months back, investigations for which elsewhere revealed dilated cardiomyopathy with ejection fraction of 35% and increased flow resistance in right renal artery.

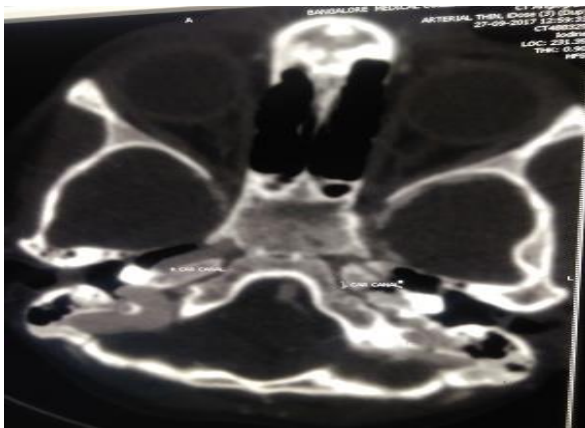


Figure 1: CT head and neck region, transverse section shows hypo plastic left sided carotid canal.

He had significant past history of having diagnosed right dysplastic kidney while evaluating for cloudy urine at 8 months of age. Patient was lost to follow up in between.

Examination revealed: PR-130 bpm with good volume, all pulses were palpable except left carotid artery, BP-118/74 mm Hg (>95th centile).

There were no discrepancies in BP checked in 4 extremities. There was mild precordial bulge and apex beat located in 5th ICS at midclavicular line.

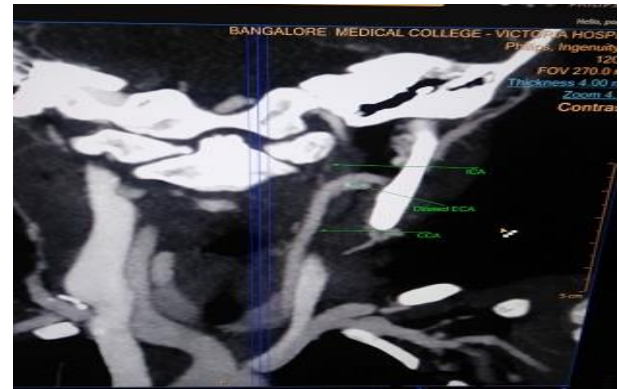


Figure 2: CT head and neck region, coronal section shows hypo plastic internal carotid artery on left side with dilated external carotid artery.

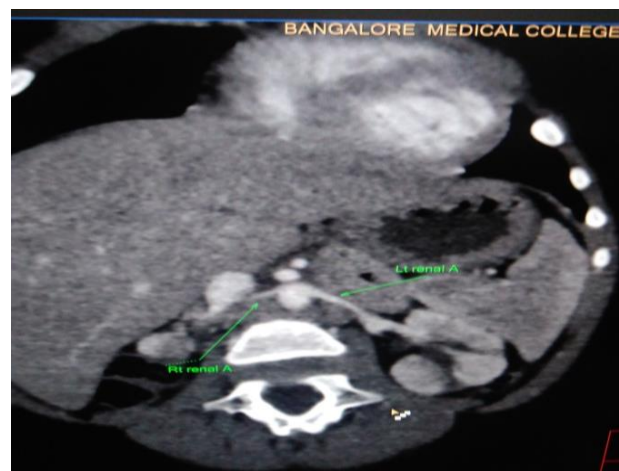


Figure 3: CT thorax region shows reduced caliber of right sided renal artery.

There was no palpable thrill and heart sounds. S1 was soft and P2-normal intensity. All blood investigations being normal- CRP-<0.02, ESR-11, ANA - negative. CT revealed HICA on the left with dilated left external carotid artery. Right sided vessels were normal.

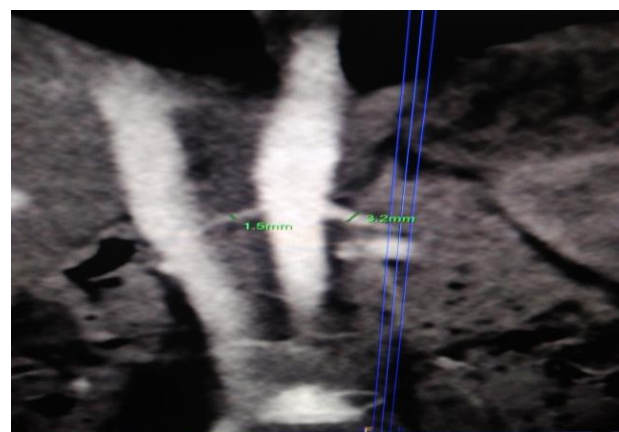


Figure 4: CT thorax coronal section- hypo plastic renal artery right sided.

Anterior cerebral artery was reformatting by anterior communicating artery (ACOM), middle cerebral artery (MCA) by left posterior communicating artery (PCOM).

In bone window, left carotid canal was also hypoplastic. Incidentally, there was also vertebral fusion anomaly-block vertebrae C5, C6 and C7, synostosis of right T4-T5, left T1-T2 and T10-T11. DTPA scan showed reduced differential function on right side (89% left kidney and 11% right kidney). Child was started on oral antihypertensive Tab amlodipine 0.3 mg/kg/day and asked for regular follow up.

DISCUSSION

Congenital absence of the internal carotid artery (ICA) is a rare anomaly that occurs in less than 0.01% of the population. It encompasses agenesis, aplasia, and hypoplasia.¹ About 100 cases of absence or underdevelopment of the internal carotid are reported in the literature, but the real incidence may be higher because they usually do not come to medical attention.⁸

Tode, in 1787 became pioneer to demonstrate carotid agenesis on postmortem examination.⁹ In 1954, the first case of ICA agenesis at cerebral angiography was reported by Verbiest.¹⁰

Lie et al, defined agenesis as complete failure of an organ to develop, aplasia as lack of development (but its precursor did exist at one time), and hypoplasia as incomplete development of the organ.¹¹

Although an exact cause of these developmental anomalies has not been established, all three variations are thought to represent the sequela from an insult to the developing embryo. Postulated causes of unilateral absence have centered on mechanical and hemodynamic stresses placed on the embryo, including effects related to exaggerated folding of the embryo toward one side and constriction by amniotic bands.⁶

The most common type of collateral flow is through the circle of Willis, ACOM and PCOM. Less commonly, collateral flow is provided via persistent embryonic vessels or from transcranial collaterals originating from the external carotid artery (ECA) system.

Although many of these cases remain asymptomatic and go undetected, it is associated with cerebral aneurysms. Six pathways of collateral circulation in association with aplasia or hypoplasia (a/hypoplasia) of the ICA are described.¹¹

Patients with HICA may be completely asymptomatic due to collateral blood supply to the affected hemisphere, or they may present symptoms due to cerebrovascular insufficiency or compression by enlarged collateral cerebral vessels. The increased blood flow through collateral vessels and altered flow dynamics may cause

ACOM and PCOM aneurysms. In these patients, the prevalence of intracranial aneurysm is estimated to about 24-34% while in the general population it is 2-4%.² Hence, they need to be followed up for any new neurological manifestations of cerebral aneurysms, ischemia, other system involvement such as skeletal deformities, renal dysplasia and progression of renal impairment, requirement of renal replacement.

CONCLUSION

HICA is rarely reported in children along with, vertebral anomalies, renal dysplasia and dilated cardiomyopathy. Detailed clinical examination is critical to avoid missing or delayed recognition of associated anomalies like HICA and vertebral anomalies. Underlying genetic basis may have to be evaluated.

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Ethical approval: Not required

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