SITUS INVERSUS;
AN UNUSUAL VARIATION OF CAROTID SYSTEM

CASE REPORT
PROF-1877

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ABSTRACT… An unusual case of situs inversus was observed during routine postmortem of a 60 year old male subject at Government medical college, Amritsar, Punjab, India. On the right side the common carotid artery was found to be absent. The right external carotid artery arose directly from the arch of aorta. The left internal carotid artery had a wide diameter and bifurcated . The right subdivision crossed over compensating for absent right internal carotid artery. The compensation of absent internal carotid artery in situs inversus makes present case unique and such variation has thus far never been reported in literature.The ontogeny and clinical implications of the variation have been discussed.

Key words: Situs inversus, absent internal carotid.

INTRODUCTION

Situs inversus viscerum is a rare congenital variant occurring in about one of 10.000 people[1].The aetiology is uncertain and the pathological condition of transposed organs often misleads the clinician. Agenesis, aplasia, and hypoplasia of internal carotid artery (ICA) occurs in less than 0.01% of the population[2,3]. In this setting, the most common type of collateral flow is through the circle of Willis. Less commonly, collateral flow is via persistent embryonic vessels / transcranial collaterals originating from external carotid artery (ECA) system. Slightly more than 100 cases of congenital absence of the ICA have been reported in the literature[4].The compensation of absent internal carotid artery in the setting of situs inversus makes present case unique and such variation has thus far never been reported in literature.

CASE REPORT

An unusual case of situs inversus was observed during routine postmortem of a 60 year old male subject at Government medical college, Amritsar, Punjab, India. The thorax and abdomen were systemically opened and position of heart, other visceral organs and blood vessels noted. On the right side the common carotid artery was found to be absent (Figure 1).

The right external carotid artery arose directly from the arch of aorta. The brachiocephalic trunk was present on left side giving left common carotid artery and left subclavian artery(Figure 2).

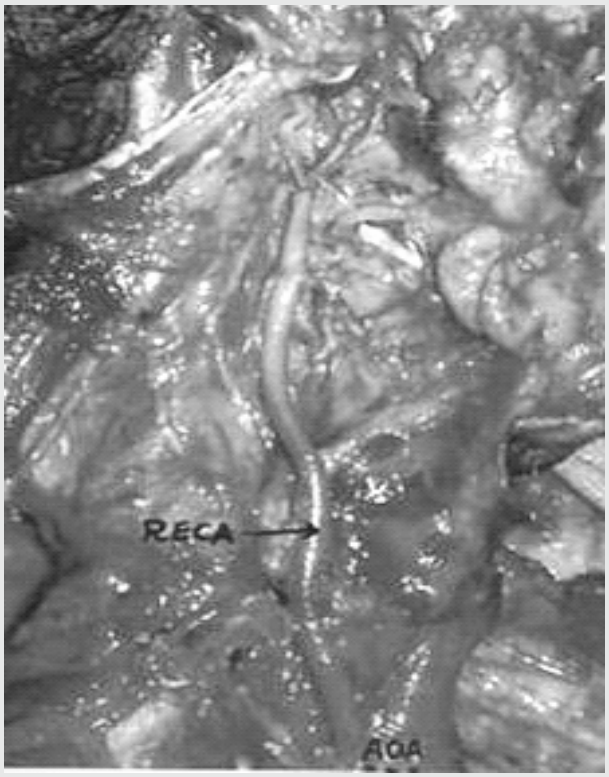

Fig-1. Right external carotid artery arose from arch of aorta and continued as internal carotid artery. Right common carotid artery was absent.(RECA: Right external carotid artery; AOA:Arch of aorta)

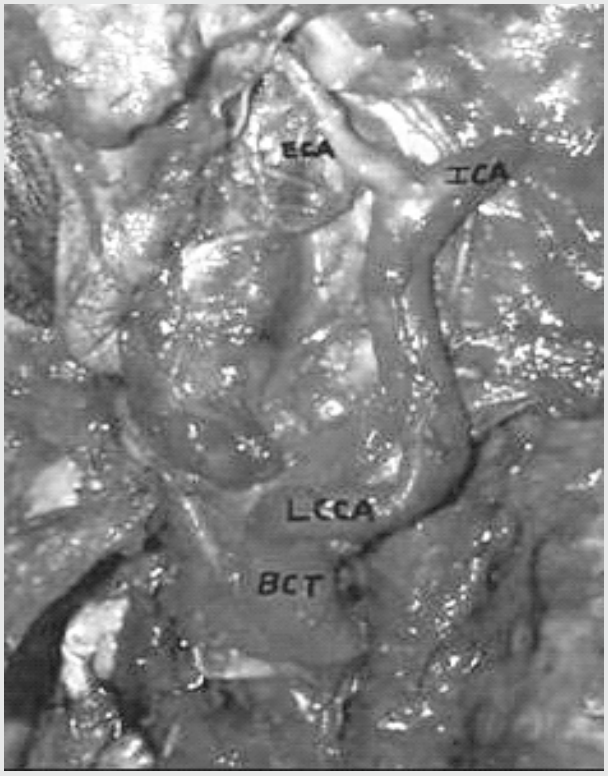

Fig-2. Brachiocephalic trunk on left side giving left common carotid artery and left subclavian artery (BCT: Brachiocephalic trunk; LCCA: left common carotid artery; LSA: left subclavian artery)

The left common carotid artery had a broad diameter and it divided into external and internal carotid arteries. The left internal carotid artery had a wide diameter and after entering base via carotid canal it bifurcated. The right subdivision crossed over to the right side. It passed through the sphenoidal air sinus to reach the foramen lacerum and followed normal course thereafter compensating for absent right internal carotid artery. The left subdivision passed through foramen lacerum of its own side and continued as the internal carotid artery of the left side(Figure 3).

DISCUSSION

Situs inversus was recognized in 1600 and in 1824 the condition was noted in living person for first time[1]. Tode is credited with the first documented case of carotid agenesis, discovered on postmortem examination in

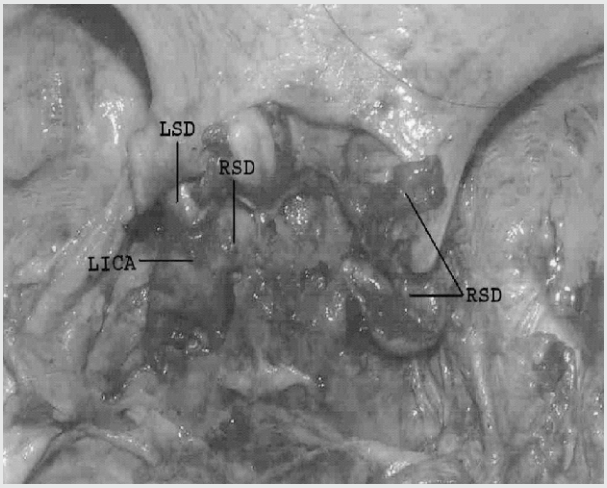

Fig-3. Left internal carotid artery bifurcated; right subdivision crossed to right side compensating for absent right internal carotid artery. Left subdivision passed through foramen lacerum of its own side. (LICA: left internal carotid artery; LSD: left subdivision; RSD: right subdivision).

1787[5]. In 1954, the first case of ICA agenesis at cerebral angiography was reported by Verbiest[4].

Ontogeny: Our knowledge of origin of situs inversus is far from complete. The likely cause in the absence of evidence seems to be the inversion of the viscera in pre-embryonic period. Early in embryonic development the tube like structure that becomes the heart forms a loop towards the left, identifying the left/right axis along which other organs should be positioned. Situs inversus can be seen in families. Postulated causes of unilateral absence of internal carotid artery have centered on mechanical /hemodynamic stresses on embryo, including effects related to exaggerated folding of embryo toward one side and constriction by amniotic bands[6]. To date, an explanation for bilateral absence has not been rendered [6]. Padget's[7] analysis of embryo provides useful insight into development of ICA, revealing its origin from dorsal aorta and third aortic arch at approximately 4-mm to 5-mm embryonic stage, with full development of ICA by 6 weeks (Fig 4).

Although it is generally accepted that ICA originates from third aortic arch, origin of CCA and ECA remains

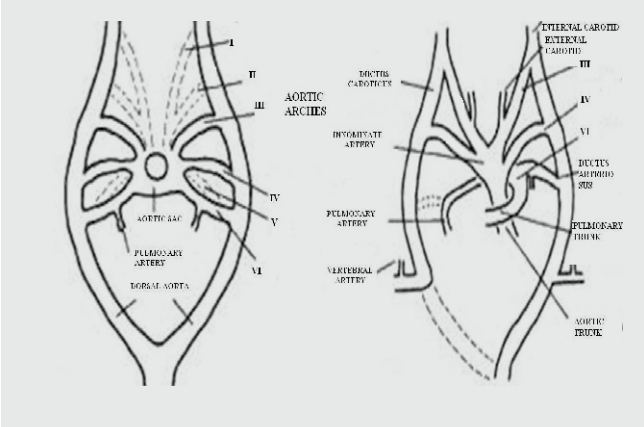

Fig-4. Illustrations of the developing embryo at 6mm and 1mm[8].

controversial[4,8,9]. Some investigators argue that both proximal ICA and the ECA arise jointly from third aortic arch, and thus agenesis of ICA should be accompanied by absence of ipsilateral ECA. Others argue that ECA and CCA can develop normally in setting of ICA agenesis, as the former arises independently from aortic sac[8,9]. The latter seems more plausible, as numerous cases of ICA agenesis exist with normally developed ECA systems.

CLINICAL IMPLICATIONS

Situs inversus may not be diagnosed until later in life. The alterations of left-rightight development can have serious cinical implications including cardiac defects. In some cases situs inversus is commonly associated with serious primary ciliary dyskinesis(sterility),chronic bronchitis and splenic malformations and may be transmitted as an autosomal recessive trait called Kartagener's syndrome[1].

Congenital absence may be unilateral or bilateral, although the unilateral variety is distinctly more common[10]. While the literature supports a nearly 3:1 left sided predominance of ICA absence[6], there is no side predilection when absence of the ICA is associated with an intercavernous anastomosis[5]. While many cases of absence of ICA remain asymptomatic these patients may present later in life with symptoms related to cerebrovascular insufficiency. The estimated prevalence of cerebral aneurysms in the general population is 2% to 4%, but the reported prevalence of aneurysms in association with absence of the ICA is 24% to 34%[2,9]. Increased flow through collateral vessels and altered flow dynamics are cited as plausible explanations for this increased prevalence[2,9]. The increased risk of aneurysm has been listed as an indication for clinical and radiologic surveillance in these patients[9]. Recognition of absence of internal carotid artery has important implications during carotid endarterectomy[11] and transsphenoidal hypophyseal surgery[12], and in the setting of thromboembolic disease.

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Article received on: 24/10/2011 Accepted for Publication: 10/11/2011 Received after proof reading: 03/01/2012

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Article Citation:
Sharma RK, Agnihotri G. Situs inversus; an unusual variation of carotid system. Professional Med J Feb 2012;19(1): 137-140.

Appreciation is a wonderful thing: It makes what is excellent in others belong to us as well.

Voltaire