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Case Report

Left brachiocephalic vein vascular ring on CT: A rare venous anomaly ☆

Varun Taruvai, BS, Pierre Maldjian, MD*

Department of Radiology, Rutgers New Jersey Medical School University Hospital, 150 Bergen Street, Newark, NJ 07103, USA

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ABSTRACT

We present a case of a vascular ring formed by the left brachiocephalic vein. A left brachiocephalic vein ring or circum-aortic left brachiocephalic vein is a rare congenital vascular variant. Although it is usually an incidental finding on chest imaging studies, left brachiocephalic vein anomalies, particularly the ring variant, can be clinically significant during procedures requiring installation of transvenous implantable devices such as pacemakers. In this report, we illustrate the appearance on computed tomography of this rare anomaly and discuss an embryological hypothesis for the etiology.

Introduction

Normally, the left brachiocephalic vein (LBCV) courses obliquely downward anterior to the aortic arch before joining the right brachiocephalic vein to form the superior vena cava. Rarely, this vein may take an anomalous course. In one rare variant, known as a circum-aortic LBCV or double LBCV, the LBCV divides into 2 branches, one coursing anterior and one posterior to the ascending aorta. Typically, an anomalous left brachiocephalic vein is an incidental finding on chest imaging modalities. In this article, we report a case of incidental identification of a LBCV ring on CT and discuss its clinical significance as well as an embryological hypothesis for the etiology.

Case report

A 62-year-old man with a past medical history significant for hypertension presented to the emergency department after a syncopal episode associated with chest pain and shortness of breath. A CT angiogram of the chest was performed due to concern for aortic dissection. The CT study showed no abnormalities of the aorta but assessment of the great vessels revealed that the LBCV split into 2 branches. The first branch traversed the expected course of the LBCV, anterior to the aortic arch across the anterior mediastinum to drain into the superior vena cava (Fig. 1A). The second branch followed an aberrant course passing posterior to the ascending aorta through

Abbreviations: LBCV, left brachiocephalic vein; CT, computed tomography.

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* Corresponding author.
E-mail address: vt212@njms.rutgers.edu (V. Taruvai).

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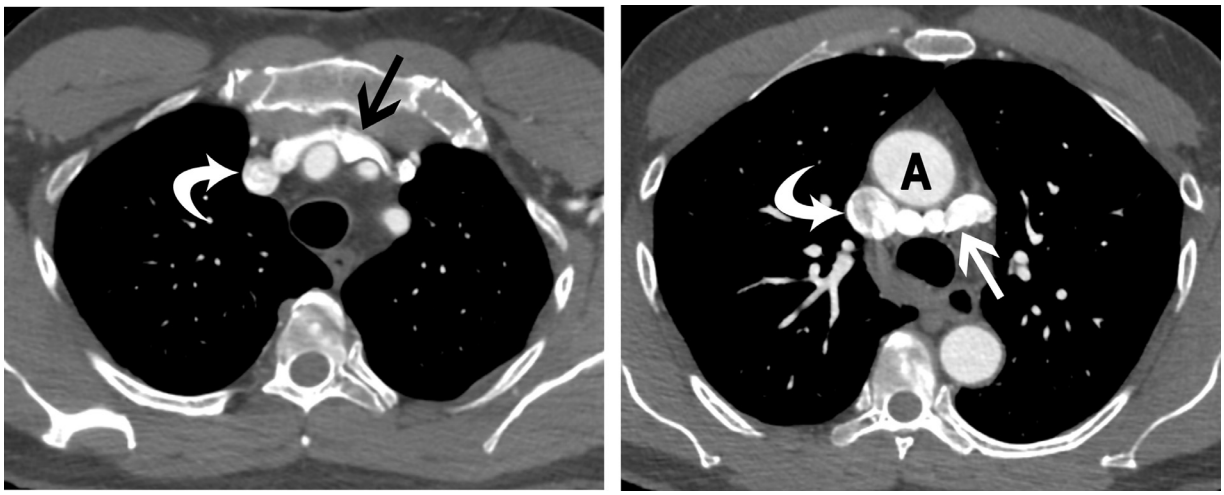

Fig. 1 – A 62-year-old male with a circumaortic left brachiocephalic vein, CT of the chest with administration of iodinated contrast. (A) Axial view shows anterior limb of left brachiocephalic vein (straight arrow) in anterior mediastinum coursing to the superior vena cava (curved arrow). (B) Axial image at a slightly lower level shows posterior limb of left brachiocephalic vein (straight arrow) coursing posterior to ascending aorta (A) to the superior vena cava (curved arrow).

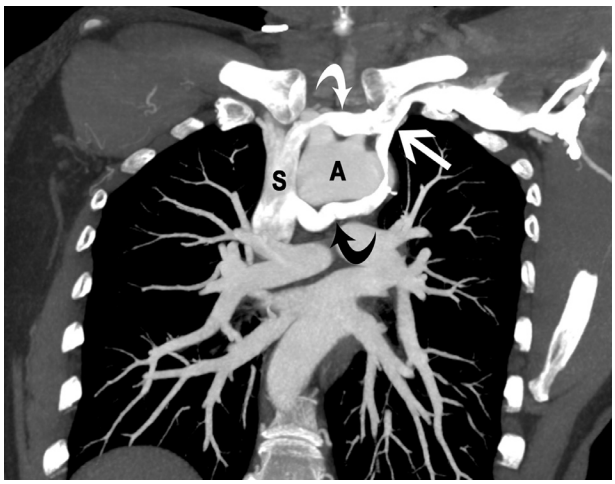

Fig. 2 – A 62-year-old male with a circumaortic left brachiocephalic vein. Maximum intensity projection (MIP) coronal image shows left brachiocephalic vein (straight arrow) splitting into a superior branch (curved white arrow) that courses superior to the aortic arch (A) and an inferior branch (curved black arrow) that passes below the aortic arch (A). Note that the inferior limb bends to nearly a right angle as it courses beneath the arch. Both branches drain to the superior vena cava (S).

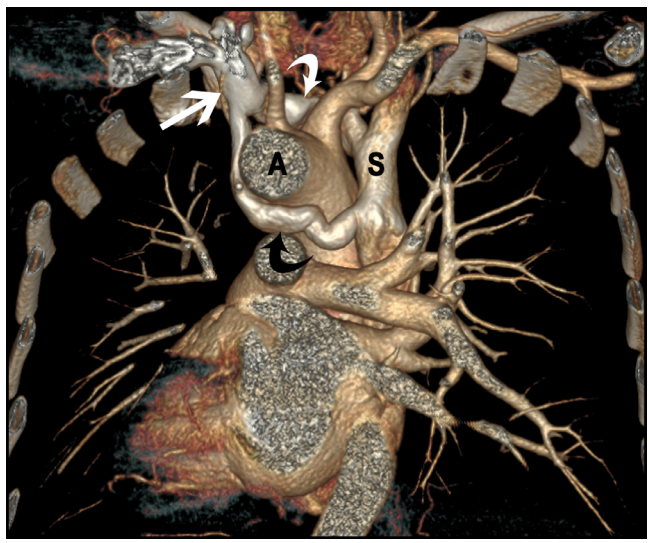

Fig. 3 – Volume rendered view from posterior perspective shows left brachiocephalic vein (straight white arrow) with superior (curved white arrow) and inferior (curved black arrow) branches forming a vascular ring around the aorta (A).

the aortopulmonary window (Fig. 1B). Coronal maximum intensity projection demonstrated that both branches drained to the superior vena cava (Fig. 2). Volume rendered views and cinematic rendering of the great vessels further illustrate the appearance of the anomalous LBCV (Figs. 3 and 4). No other abnormalities of the heart, lungs, or great vessels were observed. Further workup, including laboratory testing, was unremarkable. The patient was discharged and given an outpatient appointment for follow up.

Discussion

Normally, the left brachiocephalic vein (LBCV) courses obliquely downward anterior to the aortic arch before joining the right brachiocephalic vein to form the superior vena cava. Rarely, this vein may take an anomalous course. LBCV anomalies comprise 0.2-1% of all congenital cardiac anomalies [1]. Further classification of anomalous LBCV formation is based on variants in the course of the aberrant vessel. Among these, a circum-aortic or double LBCV is exceedingly unusual,

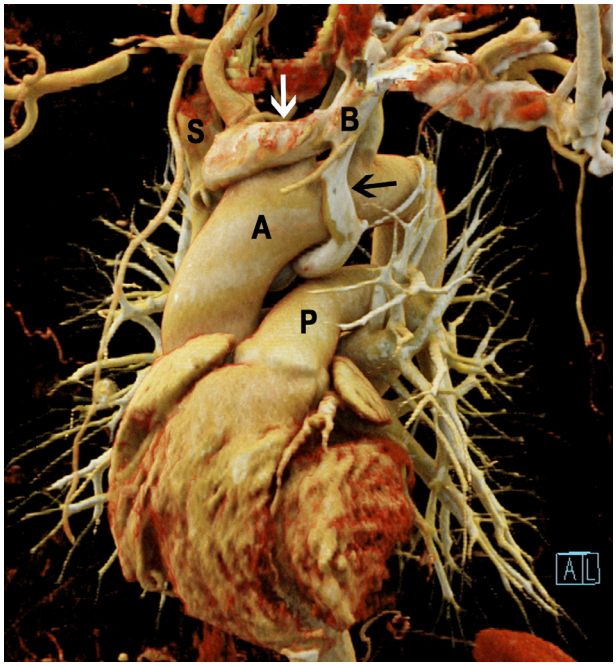

Fig. 4 – Cinematic rendering from anterior perspective shows the left brachiocephalic vein (B) dividing into superior (straight white arrow) and inferior (straight black arrow) limbs encircling the aorta (A). Note that inferior limb passes through the aorto-pulmonary window. P, pulmonary artery; S, superior vena cava.

making up only 0.7% of all anomalous LBCV cases [2]. LBCV anatomic abnormalities can occur in association with other congenital cardiac defects, most commonly tetralogy of Fallot [3]. Notably, no other cardiac structural abnormalities were observed in this case of anomalous LBCV. Prior retrospective studies on demographic characteristics of LBCV cases demonstrated a mean age of diagnosis of 4 years and 9 months with a male predominance of 1.5:1, but these studies are limited by sample size due to the rarity of this anomaly [1,4]. Risk factors for anomalous development of the LBCV are unknown, but are possibly similar to those associated with other congenital cardiac malformations, such as uncontrolled maternal diabetes, intrauterine alcohol or tobacco exposure, or chromosomal abnormalities [3].

Anomalies of the LBCV are usually asymptomatic and encountered incidentally during imaging of the chest. However, the LBCV ring variant can have clinical consequences, especially during procedures involving transvenous catheter insertion or during placement of implantable electronic cardiac devices. In one report, insertion of a cardiac pacemaker was complicated as the pacemaker lead entered the inferior limb of a double LBCV and was unable to negotiate the sharp angle in its course. Removal and reinsertion of the lead through the upper LBCV branch was necessary for successful placement [5]. Note that in our case the inferior branch of the anomalous LBCV bends to nearly a right angle before draining to the superior vena cava potentially impeding passage of a wire lead (Fig. 3).

Regarding the embryogenesis of the LBCV one theory, known as the Adachi hypothesis, proposes that during fetal development 2 precardinal anastomoses exist, one dorsal and one ventral to the aortic sac. During the seventh embryonic week, the dorsal precardinal anastomosis regresses, leaving the ventral anastomosis to form the normal LBCV [6,7]. Failure of the dorsal anastomosis to regress results in a double LBCV or LBCV vascular ring as in our case. Also supporting this hypothesis is an additional LBCV variant, the retro-aortic or subaortic LBCV, where only the posterior component remains passing through the aortopulmonary window, presumably due to regression of the ventral anastomosis [8].

In conclusion, a LBCV ring is a rare congenital vascular malformation, usually an incidental finding on imaging studies. Its morphology lends support to one embryological hypothesis regarding formation of the LBCV.

Human and animal rights

Informed consent was obtained for reporting of the following case. No experimentation on humans or animals was performed for this report.

Patient consent

Written, informed consent for publication of this case was obtained from the patient.

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