RETROESOPHAGEAL LEFT BRACHIOCEPHALIC VEIN IN AN INFANT WITHOUT CARDIAC ANOMALIES

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Abstract
Retroesophageal course of the left brachiocephalic vein is a rare variant seen in patients with congenital heart disease. However, this anomaly without associated cardiac or aortic abnormalities is nearly unheard of, with only one prior case described in the literature. We present an infant with anomalous retroesophageal left brachiocephalic vein that was an incidental finding on computed tomography (CT). We also briefly discuss its embryologic and clinical significance.

Key words: aberrant left brachiocephalic vein, retroesophageal, embryology, congenital, CT

INTRODUCTION
The left brachiocephalic vein (LBCV) usually courses in front of the three branches of the aortic arch, joining the right brachiocephalic vein behind the sternoclavicular joint to form the superior vena cava (SVC). Aberrant variations of the LBCV are uncommon, usually seen in the setting of congenital heart diseases that cause right ventricular outflow obstruction (e.g. tetralogy of Fallot)1. Retroesophageal LBCV is a particularly rare variant. It was first described in a case report by Yigit et al. as an incidental computed tomography (CT) finding in a 15-year old child with no other cardiac anomalies2. Six other cases have subsequently been identified, all in patients with congenital heart disease3-5. This report presents a rare case of retroesophageal left brachiocephalic vein in a patient without other cardiac defects.

CASE REPORT
A 4-month old boy was admitted to the hospital for evaluation of feeding issues. His mother reported that during feeding he appeared to gag and choke and tended to spit up his formula during eating. She felt that he was a slow feeder and tended to have noisy breathing during feeding. She reported that these symptoms were present since birth. An echocardiogram was obtained at birth, which did not reveal any significant abnormality. Routine prenatal ultrasound had also been performed at 18 weeks’ gestation with similarly normal cardiac findings.

Vital signs and pulse oximetry were normal. Auscultation revealed normal respiration with no audible stridor. The patient had no syndromic features with a normal physical examination.

A barium swallow obtained to evaluate his feeding issues suggested a mild indentation on the posterior wall of the esophagus. It also revealed gastroesophageal reflux. An echocardiogram was obtained to rule out a vascular ring. The echocardiogram showed a normal left aortic arch with normal branching of the head and neck arteries. CT angiography was acquired to evaluate the posterior indentation of his esophagus and demonstrated the retrotracheal-retroesophageal course of an anomalous left brachiocephalic vein and confirmed normal aortic arch anatomy [Fig. 1]. The posterior indentation of the esophagus on the esophagram corresponded to the retroesophageal course of the anomalous LBCV on CT [Fig. 2]. The retroesophageal LBCV is deemed unlikely to be causing the patient’s feeding problems due to the relative collapsibility of venous structures and the mild severity of posterior esophageal indentation in this particular case. The patient was diagnosed with gastrointestinal reflux and prescribed lansoprazole. The baby is now seven months old and the feeding has improved with documented weight gain from 3.147 kilos at birth to 7.711 kilos at seven months.

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Aberrant brachiocephalic vein (ABCV) is a rare anomaly. It is usually observed in patients with cardiac defects, with reported incidence of 0.2%-1.0%. The most common association is tetralogy of Fallot, but it is also associated with septal defects and right atrial isomerism. Even with increasing application of noninvasive imaging techniques such as echocardiography, CT, and magnetic resonance (MR) imaging, incidental detection of ABCV remains extremely uncommon. In an analysis of 4805 patients without congenital heart disease, Nagashimi et al reported only one case of ABCV.

The ABCV usually courses anterior to the left pulmonary artery before reaching the left atrium, at which point the vessel turns behind the ascending aorta to meet the right-sided superior vena cava. The first known report of retroesophageal brachiocephalic vein was published by Yigit et al. in 2008 as an incidental CT finding in a 15-year old child without associated cardiac abnormalities. Shortly after, Ming et al. identified this condition on CT in four patients with the following congenital heart diseases: ventricular septal defect (VSD); pulmonary atresia (PA) and VSD; PA and VSD; patent ductus arteriosus (PDA) and VSD. Recently, this variation was also described in patient who underwent magnetic resonance imaging (MRI) after tetralogy of Fallot repair. In the largest prenatal assessment of LBCV, Karl et al. used fetal cardiac ultrasonography to identify 1 out of 1437 fetuses with retroesophageal LBCV. The patient had “complex cardiac malformation” that was otherwise unspecified. In all, seven patients with retroesophageal LBCV had previously been described in the literature, all but one of whom had other cardiac anomalies.
Interestingly, prenatal ultrasound and echocardiography at birth did not reveal the aberrant LBCV. On echocardiography, the LBCV can be visualized with the three vessels and trachea (3VT) view. Recently, the International Society of Ultrasound in Obstetrics and Gynecology (ISUOG) recommended the inclusion of this view in the guidelines for fetal cardiac screening, although it was not used in the assessment of this patient. Rates of visualization of the LBCV on the 3VT view are as high at 94.2%. Identifying abnormal size or absence of the LBCV is important as such features can aid in the prenatal detection of anomalies of systemic and pulmonary veins.

It is important to understand the embryologic development of the brachiocephalic vein. The primordia of the systemic veins first appear as paired precardinal and postcardinal veins that join to form a common cardinal vein draining into the primitive sinus venosus. The left and right precardinal veins are connected by transverse anastomotic channels (the superior and inferior transverse capillary plexuses) that form above and below the fourth aortic arch by week 8. The surrounding arterial systems (aortic arch and pulmonary arteries) develop between week 4 and 7 of fetal development and exert inferior and dorsal pressure on the developing veins. This pressure causes the inferior transverse capillary plexus to regress while venous blood is shunted to the ventral portion of the superior transverse capillary plexus. The superior transverse capillary plexus forms the left brachiocephalic vein, its final position ventral and superior to the aorta facilitated by pressure from the developing arterial systems.

The exact pathogenic mechanism causing aberrant LBCV remains unknown. It has been proposed that abnormal development of the aortic arch or pulmonary arteries might alter the pressure exerted on the primordial veins in such a way to cause sparing of the inferior transverse capillary plexus or dorsal portion of the superior transverse capillary plexus.

The retroesophageal brachiocephalic vein has clinical implications, and, thus, should be highlighted if detected incidentally. Because it travels adjacent to the trachea, esophagus, and descending aorta, the retroesophageal brachiocephalic vein may obscure the surgical view during operations in the posterior mediastinum. Furthermore, inadvertent manipulation of the anomalous vein may lead to dangerous intraoperative bleeding. To the radiologist, the retroesophageal brachiocephalic vein can be mistaken for an enlarged lymph node on non-contrast CT scans. Also, difficulty may be encountered in a left-arm approach when inserting a transvenous pacemaker or central venous line.

In conclusion, retroesophageal brachiocephalic vein is rarely seen in isolation, but remains of practical importance especially in preoperative planning for cardiac or other mediastinal procedures.