

“ Aorta-in-Aorta ” Sign on Chest Radiograph Representing Enlarged Left Superior Intercostal and Hemiazygos Veins¹

Yon Mi Sung, M.D., Kyung Soo Lee, M.D., Tae Sung Kim, M.D.

We recently encountered a patient with membranous obstruction of the inferior vena cava in whom the left superior intercostal and hemiazygos veins were dilated. At chest radiography, the dilation simulated the presence of a second aortic knob and descending thoracic aorta lateral to the originals, and an “ aorta-in-aorta ” appearance was thus created.

Index words : Venae cavae, abnormalities
Venae cavae, stenosis or obstruction

The aortic nipple is a dot-like projection extending laterally from the aortic arch. Enlargement of the nipple is usually due to dilation of the left superior intercostal vein as it heads for the left innominate vein lateral to the aortic arch. Where the superior vena cava - or, occasionally, the inferior vena cava - is obstructed, the aortic nipple becomes prominent and enlarged (1 - 4); according to Friedman et al. (1), a nipple larger than 4.5 mm in diameter is abnormal.

Where they are dilated posterolateral to the descending thoracic aorta, the hemiazygos and accessory hemiazygos veins may lead to a new interface adjacent to the descending thoracic aortic interface (5). We recently encountered a patient with membranous obstruction of the inferior vena cava in whom the left superior intercostal and hemiazygos veins were dilated. At chest radiography, this simulated the presence of a second aortic knob and descending thoracic aorta lateral to the originals, and an “ aorta-in-aorta ” appearance was thus created.

Case Report

Over a two-week period, a 57-year-old woman experienced nausea and blood-tinged vomiting. Physical examination revealed caput medusa of several collateral veins in the anterior abdominal wall and extensive varicose veins in both lower legs, and the edge of the liver was palpated two centimeters below the right costal margin. A chest radiograph depicted a soft-tissue bulge lateral to the aortic arch and a descending interface lateral to the presumed original descending aorta (aorta-in-aorta sign) (Fig. 1A). CT of the thorax and upper abdomen demonstrated marked distension of the left superior intercostal vein (Fig. 1B) and the azygos, hemiazygos, and accessory hemiazygos veins (Fig. 1C). The intrahepatic inferior vena cava was partially opacified by intraluminal calcification (Fig. 1C), and the dilated left superior intercostal vein formed a venous arch lateral to the aortic arch and headed for the left innominate vein (Fig. 1B).

Esophagogastroduodenoscopy indicated that esophageal varices were present, and ultrasonography-guided biopsy of the liver revealed inactive liver cirrhosis. Inferior vena cavography, performed prior to balloon angioplasty, showed almost complete obstruction of the inferior vena cava at the level of the diaphragm

¹Department of Radiology, Samsung Medical Center, Sungkyunkwan University School of Medicine

Received December 20, 2001 ; Accepted March 4, 2002

Address reprint requests to : Kyung Soo Lee, M.D., Department of Radiology, Samsung Medical Center, Sungkyunkwan University School of Medicine, 50 Ilwon-dong, Kangnam-ku Seoul 135-710, Korea.

Tel. 82-2-3410-2518 Fax. 82-2-3410-2559

E-mail: kslee@smc.samsung.co.kr

(Fig. 1D). There was a 13.5-cm-H₂O pressure gradient between the right atrium and the inferior vena cava, but after balloon angioplasty, this decreased to zero-cm-H₂O.

Discussion

In our patient, continuous round and tubular soft-tis-

sue lesions lateral to the true descending aorta and aortic arch, and due to dilation and enlargement of the left superior intercostal and hemiazygos/accessory hemiazygos veins, formed the aorta-in-aorta sign.

The left superior intercostal vein arises from the confluence of the left second, third, and fourth intercostal veins, and in three-quarters of all cases communicates with the accessory hemiazygos vein at its lower end (2).



Fig. 1. A 57-year-old woman with membranous obstruction of the inferior vena cava.
A. Chest radiograph shows a round bulge (arrows) lateral to the aortic arch (arrowheads). Also note a descending interface (open arrows), continuous with the round bulge, lateral to the original descending aortic interface (curved arrows).
B. Continuous enhanced (7-mm collimation) helical CT scans obtained at the level of the aortic arch show the accessory hemiazygos vein communicating with the left superior intercostal vein (arrows). Merge of unopacified blood flow of the left superior intercostal vein to the left innominate vein resulted in a nodular low-attenuation in the innominate vein (arrowhead).
C. CT scans obtained at the level of the liver dome show markedly distended azygos and hemiazygos veins (arrows) posterior to the descending aorta. Also note partially opacified intrahepatic inferior vena cava containing internal calcification (arrowhead), suggesting membranous obstruction of the inferior vena cava.
D. Inferior vena cavogram shows gradual tapering and almost complete obstruction (arrow) of the inferior vena cava at the level of diaphragm.

At the level of the third or fourth thoracic spine, the left superior intercostal vein turns anteriorly around the aortic arch to join the left innominate vein (3). The point at which the left superior intercostal vein is adjacent to the aortic knob has been named the aortic nipple (2), and this is visualized in 0.4 to 9.5 percent of normal chest radiographs (1, 3).

At chest radiography, a dilated hemiazygos/accessory hemiazygos vein may appear as a lobular paraspinal density (5), while an aneurysm of the hemiazygos vein is seen as a retrocardiac paraspinal density (6). In an anomalous inferior vena cava with accessory hemiazygos continuation, the abnormality may manifest as a tubular paramediastinal mass continuous with the left paraspinal density (7). To our knowledge, however, no report in English has described the accompaniment of the aorta-in-aorta sign by dilation of the left superior intercostal and hemiazygos/accessory hemiazygos veins.

An enlarged hemiazygos/accessory hemiazygos venous system, including the left superior intercostal vein, is usually the result of increased blood flow or venous pressure (3). This may be due to congestive heart failure, increased portal venous pressure with collateral blood flow, obstruction or a congenital anomaly of the vena cava with collateral venous return via the azygos and hemiazygos system, or anomalous pulmonary venous drainage (3). Dilated azygos and hemiazygos veins may be associated with obstruction of the superior vena cava (1 - 4) or, occasionally, of the inferior vena cava (2).

Membranous obstruction of the inferior vena cava is a rare, congenital or acquired cause of Budd-Chiari syndrome (8). With the obstruction of normal flow in the in-

ferior vena cava, blood from the lower extremities and pelvis returns to the right side of the heart through deep and superficial venous collateral vessels; there may be involvement of the azygos and hemiazygos veins, periureteric and gonadal vein collaterals, or a superficial or portal pathway (9).

In summary, dilated left superior intercostal and hemiazygos veins may, at chest radiography, simulate a further aortic knob and descending thoracic aorta lateral to the originals. The phenomenon is known as the aorta-in-aorta sign.

References

1. Friedman AC, Chambers E, Sprayregen S. The normal and abnormal left superior intercostal vein. *AJR Am J Roentgenol* 1978;131:599-602
2. McDonald CJ, Castellino RA, Blank N. The aortic "nipple": the left superior intercostal vein. *Radiology* 1970;96:533-536
3. Ball JB, Proto AV. The variable appearance of the left superior intercostal vein. *Radiology* 1982;144:445-452
4. Carter MM, Tarr RW, Mazer MJ, Carroll FE. The "aortic nipple" as a sign of impending superior vena caval syndrome. *Chest* 1985;87:775-777
5. Castellino RA, Blank N, Adams DF. Dilated azygos and hemiazygos veins presenting as paravertebral intrathoracic masses. *N Engl J Med* 1968;20:1087-1091
6. Hayward I, Forrest JV, Sagel SS. Hemiazygos vein aneurysm: CT documentation. *J Comput Assist Tomogr* 1989;13:1072-1074
7. Haswell DM, Berrigan TJ. Anomalous inferior vena cava with accessory hemiazygos continuation. *Radiology* 1976;119:51-54
8. Zamboni P, Pisano L, Mari C, Galeotti R, Feo C, Liboni A. Membranous obstruction of the Budd-Chiari syndrome. *J Cardiovasc Surg* 1996;37:583-587
9. Sonin AH, Mazer MJ, Powers TA. Obstruction of the inferior vena cava: a multiple-modality demonstration of causes, manifestations, and collateral pathways. *RadioGraphics* 1992;12:309-322

