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 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.

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

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 cavoangiography, performed prior to balloon angioplasty, showed almost complete obstruction of the inferior vena cava at the level of the diaphragm.
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-tissue 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).
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 inferior 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, perireteric 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
"Aorta-in-Aorta" Sign on Chest Radiograph Representing Enlarged Left Superior Intercostal and Hemiazygos Veins

Aortic knob
Membranous obstruction
Collateral pathway
Left superior intercostal vein
Hemiazygos vein

57% of cases, the aorta-in-aorta sign is caused by an obstruction (aortic knob), membranous obstruction (collateral pathway), or a left superior intercostal vein (left superior intercostal vein) or hemiazygos vein (hemiazygos vein) that is present in the aorta-in-aorta sign.