Primary jugular venous ectasia: A rare cause of neck mass

Jung Eun Kim, M.D., Wang-Soo Lee, M.D., Eun Jeong Cho, M.D., Sang-Wook Kim, M.D., and Chee Jeong Kim, M.D.

Department of Internal Medicine, College of Medicine, Chung-Ang University, Seoul, Korea

Venous ectasia, also called phlebectasia or venous aneurysm, is an isolated saccular or fusiform dilatation of a vein. Ectasia of the internal jugular vein was once considered rare, but is increasing in apparent frequency due to the wide use of noninvasive diagnostic modalities. A 57-year-old woman was referred for right neck discomfort that had developed 1 month earlier. She complained of a non-painful right neck swelling, located anteromedial to the sternocleidomastoid muscle. Computed tomography and color Doppler ultrasonography showed a 2×1.7-cm right internal jugular venous ectasia. The size of the jugular venous ectasia decreased after compression with a probe and increased during the Valsalva maneuver. Here, we report the first Korean case of primary internal jugular venous ectasia, which presented as an asymptomatic right neck swelling. (Korean J Med 77:124-127, 2009)

Key Words: Aneurysm; Dilatation; Jugular veins; Neck

INTRODUCTION

Ectasias, or aneurysms, are localized dilatations of any vessel such as an artery, vein, or lymphatic channel. Although arterial aneurysms are commonly encountered in medical practice, venous ectasias are rare, and those in the neck are even rarer, with approximately 200 cases reported worldwide. The rarity of jugular venous ectasia is due to the lower pressure of the superior vena cava reservoir. Venous ectasia is also known as phlebectasia, venous cyst, aneurysmal varix, venous aneurysm, and venectasia. Harris reported the first case in 1928, which described an infant with a congenital venous cyst of the superior vena cava system. Jugular venous ectasias are classified as congenital (primary) or acquired (secondary) types. Some reports suggest that venous ectasias are developmental defects due to a weakening of elastic fibers in the vascular wall. Jugular venous ectasias are usually discovered in childhood, and adult cases, as in this patient, are very rare.

We report the first Korean case of internal jugular venous ectasia in an adult woman and describe its etiology, pathology, diagnosis, complications, and management.
CASE REPORT

A 57-year-old woman presented with right neck swelling for 1 month. A mass was located in the right neck, anteromedial to the sternocleidomastoid muscle. The patient had no pain, tenderness, or voice change (Fig. 1). She had a left-sided simple mastectomy 13 years earlier secondary to early breast cancer. The patient had no history of trauma, irradiation, or neck surgery. The family history was non-contributory. Physical examination showed that she was of average build and had a soft, non-pulsating mass that increased in size during the Valsalva maneuver, with a blood pressure of 125/80 mmHg and pulse rate of 78 beats/min. On auscultation, no bruit was heard in the neck. In addition, the breath sounds in both lung fields were clear and regular heart sounds with no murmur were heard. A chest X-ray showed no active lung lesion and the 12-lead electrocardiogram indicated a normal sinus rhythm. The serum VDRL, antinuclear antibody, anti-dsDNA, and rheumatoid factor were all negative and C-reactive protein was within the normal range. Other routine laboratory examinations were normal. Computed tomography (CT) revealed a 2×1.7-cm right internal jugular venous ectasia (Fig. 2). Color Doppler ultrasonography showed no flow limitation (Fig. 3). The size of the ectasia decreased after compression with the probe and increased during the Valsalva maneuver. Based on these findings, we diagnosed primary jugular venous ectasia. No specific treatment was offered, but we observe the patient periodically in the outpatient department and she has no aesthetic complaints, clinical symptoms, or hemodynamic instability.

DISCUSSION

Anatomically, four types of venous ectasias have been described: intracranial, neck and chest, visceral, and upper and lower extremities. Interestingly, most jugular venous ectasias
are right-sided. Paleri et al. hypothesized that anatomical variation in the right venous system plays an important role in the right-side dominance. More direct continuity exists between the shorter right brachiocephalic vein and the superior vena cava than between the left brachiocephalic vein and superior vena cava. The right internal jugular vein is also larger, and the right-sided valves are located at a higher level than those on the left side. Moreover, the incidence of valvular competence is lower in the right jugular vein and higher in the right subclavian vein when compared to the left. Therefore, any increase in intrathoracic pressure is transmitted more easily to the right jugular vein than to the left, which explains the right dominance in this and other cases.

The etiology of venous ectasia has not been fully

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Figure 2. Axial CT images show fusiform enlargement of the right internal jugular vein (arrows in A through C) without thrombus. (D) Left internal jugular vein.

Figure 3. An ultrasonogram with Doppler imaging studies shows aneurysmal dilatation of the right internal jugular vein (arrows in A through C) without flow limitations (B). (D) Left internal jugular vein.
established, although several theories have been suggested. Schatz et al. proposed that endophlebosclerosis and endophlebohypertrophy were the main factors in the development of venous ectasias, analogous to the role of atherosclerosis in the arterial system. Other hypotheses have been proposed, such as inflammation, mediastinal irradiation, trauma, congenital anomalous reduplication of the jugular vein, and increased tone in the anterior scalenus muscle. Numerous histopathologic findings have also been noted in venous ectasias. Matsuura et al. described a reduction in elastic fibers, while Gilbert et al. noted a normal venous structure and a normal elastic tissue component. Venous ectasias are distinct from varicose veins histologically. The latter have a thickened vascular wall and muscle layers with increasing fibrous tissue, while the former show loss of the elastic layer; absence of muscle cells, media, and adventitia; and thinning of the vein wall.

Jugular venous ectasias are usually soft-round or fusiform masses with smooth margins. They are located principally in the lower third of the neck at the anterior border of the sternocleidomastoid muscle. The most common clinical presentation is neck swelling, increasing in size during straining, coughing, sneezing, the Valsalva maneuver, and bending. Other symptoms include painful swelling, progressive enlargement, slight dyspnea, bruit, and voice change.

Several tools are used for diagnosing jugular venous ectasias, including ultrasonography, venography, CT, color Doppler imaging, and direct needle aspiration. Although venography was once regarded as the gold standard diagnostic technique, it is invasive and has the potential for dangerous complications. Therefore, noninvasive neck CT and ultrasonography with color Doppler, as was used in this case, are currently preferred as diagnostic methods. Rarely, jugular venous ectasias can be complicated by spontaneous rupture, congestive heart failure, thrombus formation, thrombophlebitis, or pulmonary embolism. They must be differentiated from cysts or tumors of the upper mediastinum, lymphoceles, hemangiomas, and hygromas.

The management of jugular venous ectasia is controversial. Surgical indications include an undefined mass, symptomatic unilateral jugular venous ectasia (progressive swelling, pain, cosmetic problem), and a high risk of thrombosis. Some studies have suggested surgery even in asymptomatic cases due to the potential complications of rupture, risk of thrombus formation and subsequent pulmonary thromboembolism, and emotional stress. However, we chose a conservative option, as in other asymptomatic cases, since she had no symptoms and we follow her regularly to monitor potential complications.

REFERENCES