



Superior vena cava syndrome caused by a benign intrathoracic goiter

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ABSTRACT

Intrathoracic goiters are defined as the extension of the thyroid gland into the mediastinum. Superior Vena Cava (SVC) syndrome due to the compression of major vessels can be seen in these patients. Most cases of SVC syndrome occur as a complication of malignancy. A 75-year-old female, non-smoker, was admitted with complaints of the midline neck swelling for the past 45 years, mild puffiness of face, breathlessness on lying down and dry cough for last 5 years. On clinical examination, there was a massive grade IV thyromegaly. Neck computed tomography revealed a heterogeneous, hypertrophic, nodular thyroid gland with multiple calcification and mediastinal extension with narrowed trachea. Intraoperatively, there was a huge retrosternal thyroid gland compression of the right brachiocephalic vein, the brachiocephalic trunk from behind the vessel and the right carotid artery along with the right internal jugular vein. Pathological examination showed a nodular colloid goitre without signs of malignancy. When SVC syndrome is secondary to benign thyroid disease, total thyroidectomy should be performed.

Key words: Vena cava superior syndrome, goiter, thyroidectomy

Introduction

The first case of superior vena caval obstruction was described by HUNTER in 1757 [1]. Superior vena cava (SVC) syndrome is a critical condition in which an intrathoracic mass lesion compresses the SVC and promotes the development of head and upper extremity edema and cyanosis. Either a luminal obstruction or an extrinsic compression can impair the flow in the thin-walled low-pressure SVC [2]. Approximately 73% to 97% of SVC syndromes are caused by malignancy [3] and benign giant retrosternal goitre can also lead to this syndrome.

Case Presentation

A 75-year-old female, non-smoker, was admitted with complaints of midline neck swelling for the past 45 years, mild puffiness of face, breathlessness on lying down and dry cough for last 5 years. She did not report a history of fever, palpitations, syncope, weight loss, abdominal distension, edema feet, bone pain or stridor. There was also no history of diabetes, hypertension or coronary artery disease. The family history was unremarkable for endocrinopathy or malignancy. Physical examination revealed a pulse of 92/min, blood pressure of 100/70mmHg and respiratory

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Figure 1. On clinical examination there was a huge grade IV thyromegaly.



Figure 2. Heterogeneous, hypertrophic, nodular thyroid gland with multiple calcification and mediastinal extension with narrowed trachea.

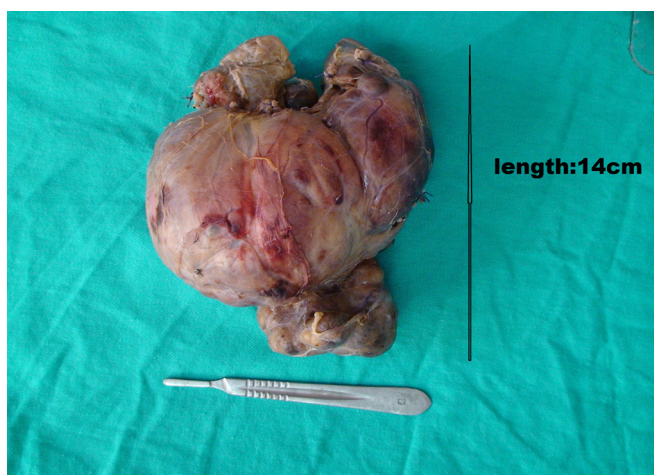


Figure 3. Total thyroidectomy (1030gr) specimen.

rate of 29/min. The face was plethoric and puffy. Distended veins were seen over the chest and arms. The right jugular vein was distended but non-pulsatile. On clinical examination there was a huge grade IV thyromegaly, 12 x 8 cm of size with retrosternal extension (Figure 1). There was no lymphadenopathy. Her pulse oximetry revealed a low oxygen saturation (89%), which improved with 2 L of intranasal oxygen. The ECG was remarkable for sinus tachycardia and nonspecific ST-T changes. Investigations revealed that hemoglobin levels, WBC, ESR, thyroid function tests, liver and renal function tests and thrombophilia work-up were all normal. An anterior-posterior chest radiograph showed a widened superior mediastinum. The ultrasonography showed retrosternal goitre with heterogeneous parenchyma and nodules with cystic-necrotic degeneration. Neck computed tomography revealed heterogeneous, hypertrophic, nodular thyroid gland with multiple calcification and mediastinal extension with narrowed trachea (Figure 2). The trachea was severely compressed and reduced to a slit at the T4 vertebral level. The thyroid mass showed extension into posterior mediastinum. The left lobe was enlarged measuring approx. 8.4 x 7.2 cm at the level of thoracic inlet, and it extended inferiorly almost to the level of aortic arch; the right lobe was also enlarged with intrathoracic extension measuring approx. 8.8 x 8.1 cm at the level of the confluence of brachiocephalic veins. Fine-needle aspiration was difficult to identify by palpation and the patient was referred to a radiologist for FNA under ultrasonographic guidance. Fine-needle aspiration cytology smears, which showed thyrocytes and rare macrophages with a colloid background, were nonspecific. There was no evidence of malignancy. Total thyroidectomy (1030gr) was performed through a Kocher's collar incision (Figure 3). Intraoperatively, there was a huge retrosternal thyroid gland compression of the right brachiocephalic vein, brachiocephalic trunk from behind the vessel and right carotid artery along with right internal jugular vein. Pathological examination showed a nodular colloid goitre without signs of malignancy. Postoperative recovery was uneventful. Values of arterial blood gas analysis normalized. The symptoms of SVCS gradually disappeared.

Discussion

Intrathoracic goiters cause adjacent structure compression more frequently than the cervical goiters, due to the limited space of the thoracic cage. Compression of the trachea, oesophagus, vascular and neural structures may cause dyspnoea, dysphagia, superior vena cava (SVC) syndrome, subclavian vein thrombosis, hoarseness, and Horner's syndrome [4]. SVC syndrome is a critical condition in which an intrathoracic mass lesion compresses the SVC and promotes the development of head and upper extremity edema and cyanosis. Either a luminal obstruction or an extrinsic compression can impair the flow in the thin-walled low-pressure SVC [2]. Approximately 73% to 97% of SVC syndromes are caused by malignancy [3]. Tumor and lymph node masses, aortic aneurysms, bronchogenic and esophageal carcinomas cause extrinsic compression [2]. Benign causes, such as iatrogenic superior vena cava thrombosis secondary to venous catheterisation or pacemaker implantation, may also result in sudden onset of symptoms [5]. On the contrary, a slowly growing substernal thyroid can be asymptomatic due to venous collateral formation [6]. The most commonly encountered collaterals are the lateral thoracic, intercostal and internal mammary veins, and the azygos vein, which is the principal collateral pathway. Huge retrosternal goitre can also lead to this syndrome; however, the retrosternal extension of huge goitre was ruled out by clinical and ultrasonographic examination [7]. Compression of the azygous vein will increase superficial venous distension [8] as in our patient.

The most common symptoms are dyspnea, facial congestion, venous distension of the chest wall, arm swelling, and facial plethora [9]. As the disease progresses, the symptoms may include hoarseness, periorbital edema, dysphagia, headaches, dizziness, syncope, lethargy, and chest pain. Other findings may include confusion and laryngeal and/or glossal edema. These symptoms may be worsened by positional changes such as bending forward, stooping, or lying down. Complications include thromboembolism, excessively high venous pressure with cerebral and life-threatening laryngeal edema [10,11].

The diagnostic modality of choice is contrast CT scan of chest and neck [2]. Computed tomography

(CT) scanning provides an effective, noninvasive evaluation of the superior vena cava and its collateral circulation. CT scanning provides anatomic details of the mediastinal and thoracic organs, allowing the identification of the cause and extent of the obstruction.

Based on the presented case and the literature, it should be remembered that the benign thyroid pathologies are treatable causes of the SVC syndrome. Early diagnosis and proper treatment in the cases of SVC syndrome due to thyroid pathologies and some other treatable conditions may prove to be life saving. Patients with SVC syndrome should have a careful thyroid exam and the diagnosis should be considered when a solid thyroid nodule is present [12]. When SVC syndrome is secondary to thyroid disease, total thyroidectomy should be performed.

Conflict of interest statement

The authors have no conflicts of interest to declare.

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