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**Case report,**

# A giant intrathoracic goiter

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**Abstract:**
Intrathoracic goiter is a relatively rare benign finding, leading often to dyspnea and dysphagia. Imaging methods are of a great importance for the diagnosis. Surgery and total thyroidectomy are the treatment of choice. We present a case of a 72-year-old woman with dysphagia, shortness of breath and fatigue. We observed an increase in both thyroid lobes with retrosternal evolution, descending to the pulmonary artery to the right, and to the aortic arch to the left, with compression of the trachea, brachiocephalic veins and vena cava superior. We performed total thyroidectomy through combined cervical-anterior thoracic right-sided access. Surgical treatment for intrathoracic goiter with or without clinical symptoms is always indicated and should be performed as soon as possible.

**Key words:** intrathoracic goiter, thyroidectomy, vena cava superior syndrome, dyspnoea, dysphagia.

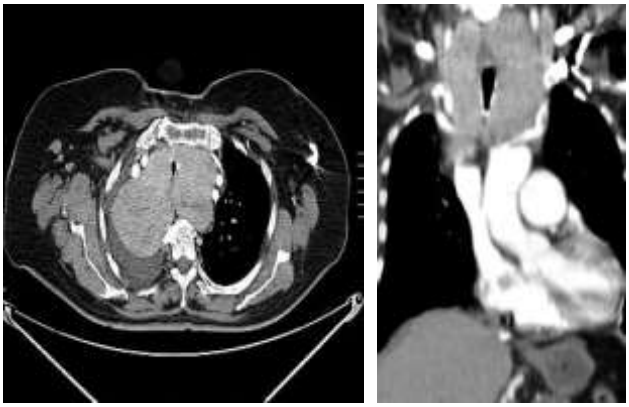
## Introduction:

Giant goiters are of interest both from a diagnostic and from a surgical point of view. Secondary retrosternal (intrathoracic) goiters develop as a result of the descended growth of mediastinal goiters [1]. Ultrasound of the neck and thorax, conventional roentgenography and computed tomography are the first imaging techniques to the proper diagnosis. If possible, transcutaneous transthoracic ultrasound-guided trucut biopsy is appropriate [2]. It is optional and can be complicated, and is usually used when malignancy is suspected in imaging studies. As invasive methods, fibrogastroduodenoscopy and flexible tracheobronchoscopy are used to assess the compression of the trachea and esophagus and their probable infiltration. Clarification of the hormonal status of the thyroid gland is required. Mediastinal and intrathoracic goiters are a rare pathology and their treatment is extremely surgical [3]. They are challenging and can cause serious technical intraoperative difficulties [4, 5].

## Clinical case:

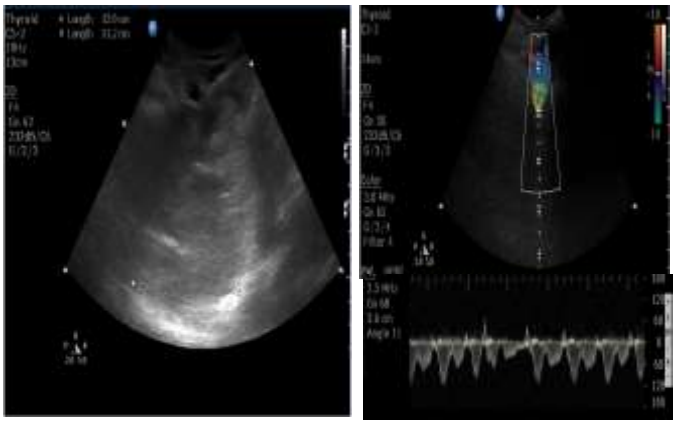
A 72-year-old woman was consulted at the Department of Gastroenterology regarding dysphagia to solid foods for about 4 months. She reported limited dietary intake and 3 kg weight loss. Furthermore, the patient reported shortness of breath and easy fatigue, dating back to 6 years, which she attributed to long-term hypertension. During the physical examination, the patient was eupnoic with a normal abdominal status. We observed cervical venous stasis, palpable edema in the neck and chest with dilatated collaterals and telangiectasias of the anterior chest wall. As an additional finding, an enlarged and compacted left part of the thyroid gland was palpated. No laboratory abnormalities were observed and TSH levels were in reference ranges. Abdominal ultrasound and fibrogastroduodenoscopy were performed, showing no pathological processes and no endoscopic stenotic section of the esophagus, including no external compression. To exclude thyroid pathology, which could explain the dysphagic and dyspnea symptoms, and due to

observed vena cava superior syndrome, the patient underwent a computed tomography (CT) examination (Figures 1A and 1B). Both parts of the thyroid gland were visualized enlarged with an inhomogeneous structure with benign characteristics. The cranio-caudal size of the right lobe was 122 mm and of the left lobe was 104 mm. The right lobe was intrathoracically located, paratracheally and paraesophageally to the level of the right main bronchus. On the left, the formation reached the aortic arch paratracheally and preesophageally. Compression of the tracheal lumen up to 5 mm from both sections was observed. The caudal part of the thyroid gland reached the bifurcation of the trachea. Compression of the lumen of the superior vena cava was visualized with the development of a small pleural effusion on the right.



**Figures:1 A and 1B. CT of the mediastinum in the axial and coronary views with evidence of retrosternal tumor formation**

The patient was referred for diagnostic and therapeutic clarification to the Surgery department. The performed fibrotracheobronchoscopy revealed mobile vocal cords and a severely hyperemic trachea, which was swollen and difficult to pass through a 6 mm bronchoscope. No infiltration was detected. The vessels of the mucosa were dilatated. By echocardiography we found no signs for thrombosis of the superior vena cava - accelerated Doppler blood flow in the area above the compression of the superior vena cava with preserved cardiac and respiratory modulation. Thoracic ultrasound (Figures 2A and 2B) again observed the giant bilateral goiter with retrosternal evolution, descending to the right to the level of the pulmonary artery and to the left to the aortic arch. Compression of the brachiocephalic veins bilaterally and the proximal segment of the superior vena cava were observed. Diagnostic and therapeutic pleural puncture was performed on the right with evacuation of a clear pleural effusion (1000 ml) with the characteristic of exudate with the presence of erythrocytes, mesothelial cells, lymphocytes, eosinophilic and neutrophilic cells. The patient was considered for radical surgical treatment.



**Figures 2: A and 2B. Thoracic ultrasound and Doppler ultrasound of giant intrathoracic goiter with development of superior vena cava syndrome**

A total thyroidectomy of the described formation was performed by bimanual access - Kocher cervicotomy and submammary anterolateral thoracotomy on the right (Figures 3, 4, 5). Histologically we observed micro- and macrofollicular goiter with degenerative changes, areas with connective tissue growth, hemorrhage, groups of macrophages with brown pigment, deposition of cholesterol crystals, focal deposition of calcium salts.

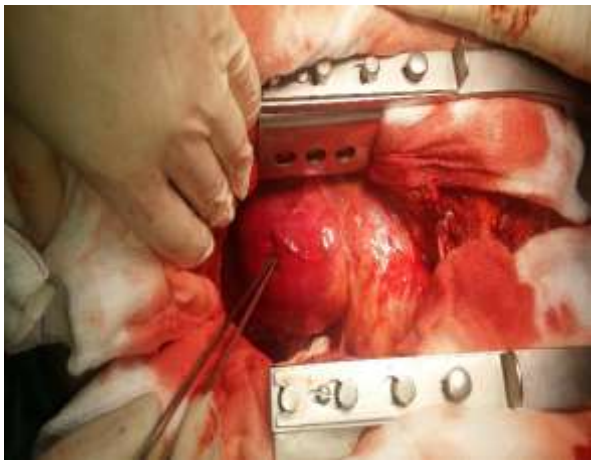


**Figure 3. Right lobe by right thoracic access**

The patient was discharged on the 11th day of the operation without complications in the postoperative period.

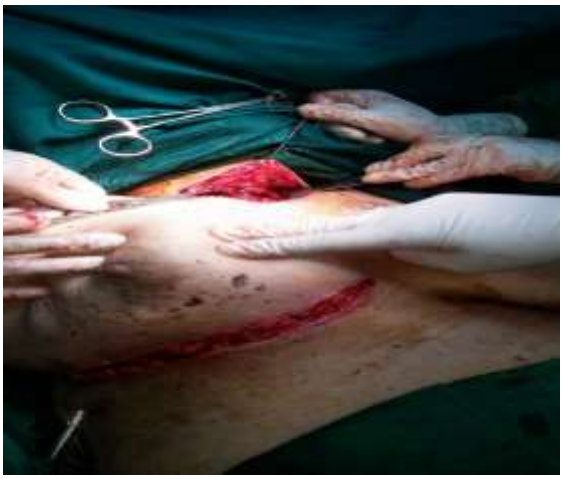


**Figure 4. Cervicotomy with right anterior submammary thoracotomy**

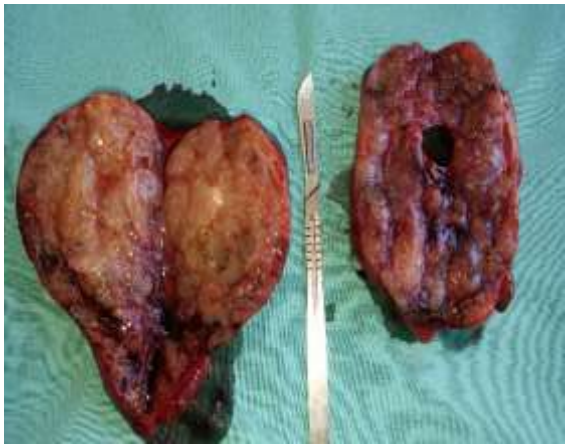


**Figure 5. Gross specimen**

## Disscusion:

Retrosternal, substernal, or intrathoracic goiter was first described by Haller in 1749 [6]. Intrathoracic goiters were observed in 3% to 20% of patients undergoing thyroid resection [7, 8]. The natural development of thyroid goiter is characterized by progressive growth, including to the thoracic cavity as mediastinal and intrathoracic tumors, which raises a diagnostic dilemma [9]. Retrosternal (intrathoracic) gioters are classified into primary and secondary. The primary ones are rare and have neither a fibrous nor a parenchymal connection with the cervical part of the gland. Secondary goiters, which occur in over 99% of cases, are formed as a result of descending growth of the thyroid gland and are supplied with blood by the cervical branches of the thyroid arteries [1, 8, 10]. Etiologically, 85% -95% of retrosternal thyroid formations are benign (goiter) and occur as a result of iodine deficiency [8]. The majority of mediastinal goiters are diagnosed in the sixth decade predominantly in women (female to male ratio 3:1) [7, 11].

Palpable cervical mass is observed in 80% -90% of patients. The development of clinical symptoms in patients with intrathoracic goiters is associated with compression of the airways or esophagus. Dyspnoea and dysphagia, which we observed in our case, as well as sleep disturbance and hoarse voice, are among the most common symptoms described in the literature [12]. The first case of obstruction of the superior vena cava was reported by Hunter in 1757 [13]. The vena cava superior syndrome is the clinical expression of its external compression or internal obstruction [14]. The syndrome was initially described as secondary to an infection such as tuberculosis or a syphilitic aortic aneurysm, however, nowadays it is almost exclusively secondary to a malignant process (more than 90% of cases) [15]. In our clinical case, we present a rare etiopathogenetic mechanism for the development of vena cava superior syndrome with subsequent pleural effusion, namely benign giant intrathoracic goiter. Obstruction (internal or external) of the superior vena cava can lead to increased hydrostatic venous pressure, which in turn leads to a limitation of the capacity of the lymph vessels in the parietal pleura with subsequent accumulation of pleural fluid [16]. Pleural effusions, uni- or bilateral, occur in 60% of cases of vena cava superior syndrome. The effusions are small and usually occupy less than half of the affected chemithorax [17]. Swelling of the neck and shortness of breath are among the most typical symptoms, which we also observed in our case [18].

Most patients with giant goiter have normal thyroid function [11]. Due to the dysphagia and possible compression of the esophagus, fibroesophagoscopy or contrast esophagography should be performed to rule out infiltration, especially in malignant mediastinal goiters. Preoperative laryngoscopy determines the location and movement of the vocal cords. Imaging is the most effective diagnostic method for verifying intrathoracic goiter and other formations in the posterior meastinum and determing the location and spread of the lesion with the corresponding involvement of surrounding organs or vessels. Mediastinal mass or developed structural distorsion (eg, partial deviation of the trachea from the midline or its compression to varying degrees) is often detected by chest roentgenography [12]. From imaging studies, computed tomography provides the most detailed information regarding anatomical connections and mediastinal and

intrathoracic goiter distribution, defines its well-defined boundaries and frequent focal calcifications, cystic areas and relatively high attenuation with prolonged enhacement after intravenous administration. Perrot et al reported that 38% of goiters were located in the prevascular region, 33% were located between the vessels and the trachea, and 27% were located retrotracheally [19]. The differential diagnosis of intrathoracic goiter includes lymphadenopathy, bronchial cyst, arterial aneurysm, neurogenic tumor, pheochromocytoma, hernia and others. Experience with MRI is limited, so CT remains an integral part of planning a surgical approach [20]. CT is the key method to diagnose superior vena cava syndrome and to develop the best therapeutic strategy in each case [18].

Surgical treatment is the treatment of choice in the presence of mediastinal and retrosternal goiter with or without clinical symptoms. Total thyroidectomy is mostly preferable in patients with a life expectancy of more than 10 years [12]. Surgical intervention is performed mainly by cervicotomy [1]. In this case, we demonstrated additional extracervical resection (submammary anterolateral thoracotomy on the right), which avoids damage to surrounding tissues and allows en block resection. Extended surgical access is required in only 2% to 3% of patients with thyroidectomy, in whom the goiter reaches intrathoracic vessels, displaces or compresses the aortic arch, leads to vena cava superior syndrome, or is observed malignant transformation [12]. Complications of surgical treatment include recurrent laryngeal nerve palsy, tracheomalacia, dysphonia, laryngeal edema, hypocalcemia, nausea, vomiting, and others [8].

## Conclusion:

Although rare, intrathoracic goiters lead to a diverse clinical presentation and a number of complications. Surgical treatment is the treatment of choice and aims at total thyroidectomy. Proper and timely diagnostic approach, careful planning and precise surgical techniques are key to achieving a successful therapeutic outcome.

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