CASE REPORT: RETROSTERNAL MULTINODULAR GOITER AND SUPERIOR VENA CAVA SYNDROME

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Abstract

Retrosternal enlargement of the thyroid can cause compression of several mediastinal structures. Tracheal obstruction and superior vena cava syndrome are rare complications of retrosternal goiter. We report a 60 old-year woman, complaining of hard dyspnea and with oedema of the face and the neck. She was treated for hyperthyroidism but not regurlarly. Alongside with other many examinations she had an ultrasonography of thyroid gland. The result was a giant thyroid with heterogeneous nodules, with necrotic area, in its both lobes and compressing phenomena over trachea and jugular vein. This thyroid extended to the superior mediastinum, until the tracheal bifurcation, an image confirmed by thoracic computed tomography. Thyroid needle aspiration biopsy didn't give any data for atypism and malignance. The patient had surgical intervention, total thyroidectomy. Histological examination confirmed multinodular goiter. One month after surgery, the patient had total remission of compressive signs and now, she feels well and is taking hormone replacement therapy regularly.

Keywords: retrosternal multinodular goiter; superior vena cava syndrom.

NJË RAST ME STRUMË MULTINODULARE RETROSTERNALE DHE SINDROMI I VENËS CAVA SUPERIOR.

Abstrakt

Hyrje: Struma multinodulare me shtrirje retrostrenale mund të shkaktojë kompresion të strukturave mediastinale. Obstruksioni i trakesë dhe sindromi i Venës Cava Superior janë komplikacione të rralla të kësaj strume.

Në shërbimin e Mjekësisë Interne paraqitet një grua, 60 vjeç, e cila ankonte për dispne të rëndë, cianozë, edema të fytyrës dhe qafës. Një e dhënë me rëndësi nga anamneza e të sëmurës ishte se vuante prej disa muajsh nga tiroidja për të cilën mjekohej jo rregullisht. Kjo gjë bëri që bashkë me shumë ekzaminime të tjera të bëhet dhe ekografia e gjëndrës tiroide, ku rezultoi një tiroide gjigande me formacione heterogjene, me zona nekrotike në të dy lobet e saj, dhe me fenomene komprimuese mbi trakenë dhe venat jugulare. Kjo tiroide shtrihej deri në mediastinin superior, imazh që u konfirmua dhe nga CT e toraksit. Citopunksioni i saj nuk dha të dhëna për fenomene të atipizmit qelizor e nuklear dhe as për mitoza patologjike. E sëmura iu nënshtrua ndërhyrjes kirurgjikale nga specialiste me pervojë dhe u bë tiroidektomi totale. Dhe biopsia rezultoi në favor

të strumës multinodulare. Sot e sëmura është në gjendje të mirë dhe i nënshtrohet rregullisht mjekimit me hormonoterapi zëvendësuese.

Fjalë kyçe: Struma multinodulare retrostrenale, sindromi i Venës Cava Superior.

Introduction

Retrosternal enlargement of the thyroid can cause compression of several mediastinal structures. The incidence of retrosternal goiters among patients with thyroid goiters is reported to range from approximately 5-45%(1). Tracheal obstruction and superior vena cava syndrome (SVC) are rare complications of retrosternal goiter. This syndrome usually presents with oedema and lividity of the face with enlargement of jugular and superior limbs' veins.
Malignancy is the most common cause (93 - 97%) of SVC syndrome (2). There are also rare cases reported of SVC syndrome associated with Graves' disease and multinodular goiter. Retrosternal goiter is estimated to 5 - 24 % causes of a mediastinal mass (3).

Case presentation.

In the Department of Internal Medicine, a 60 old-year woman came, complaining of hard dyspnea, retrosternal pain, cyanosis, hoarse voice. She had oedema of the face and neck with tortuous jugular vein and collateral circulation in both arms. She was having this situation for some days. By her carefully anamnesis, an important data, was the fact she had been suffering from thyroid disorders, for some months, without being cured regularly. Thyroid function tests were in the normal range. Enlarged, regional, lymph nodes were not palpable. Body temperature was 36.6°C, the heart rate was 82/min, and blood pressure was 135/85 mm Hg. The blood count was as follows: haematocrit: 49.5%, haemoglobin: 16.4g/dl, platelets: 380.000/mm^3, WBC: 11600/mm^3. Erythrocyte sedimentation rate was 12 mm in the first hour. The ECG was remarkable for sinus tachycardia, without specific ST-T changes.
Ultrasonography of thyroid gland has shown a huge thyroid with heterogeneous nodules, with necrotic area, in its both lobes, resulting in an enlargement of the gland and compressing phenomena over trachea and jugular vein. This thyroid extended to the superior mediastinum, until the tracheal bifurcation, with compression of the brachiocephalic vessels, an image confirmed by thoracic computed tomography (CT). (Fig.1) Thyroid needle aspiration biopsy didn't give any data about cell, nuclear atypism and pathologic mitosis.

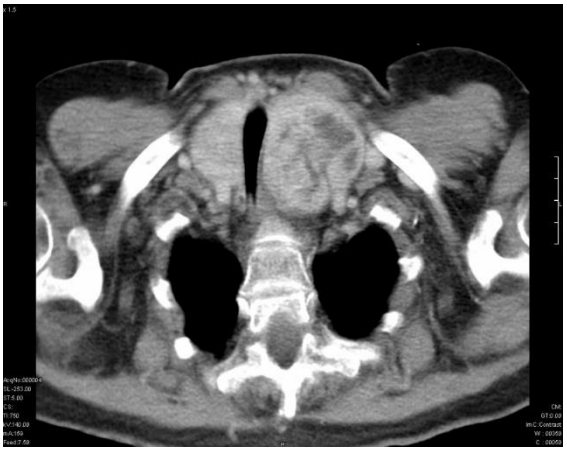

Figure. 1. CT scan of the chest. Giant retrosternal nodular thyroid gland with narrowed trachea.

The diagnosis of giant multinodular retrosternal euthyroid goiter and compression of mediastinal structures was made. The patient had surgical intervention, total thyroidectomy (Fig.2,a-b) .Histological examination confirmed nodular goiter without signs of malignancy. One month after surgery, the patient had total remission of compressive signs and now, she feels well and is taking hormone replacement therapy regularly.

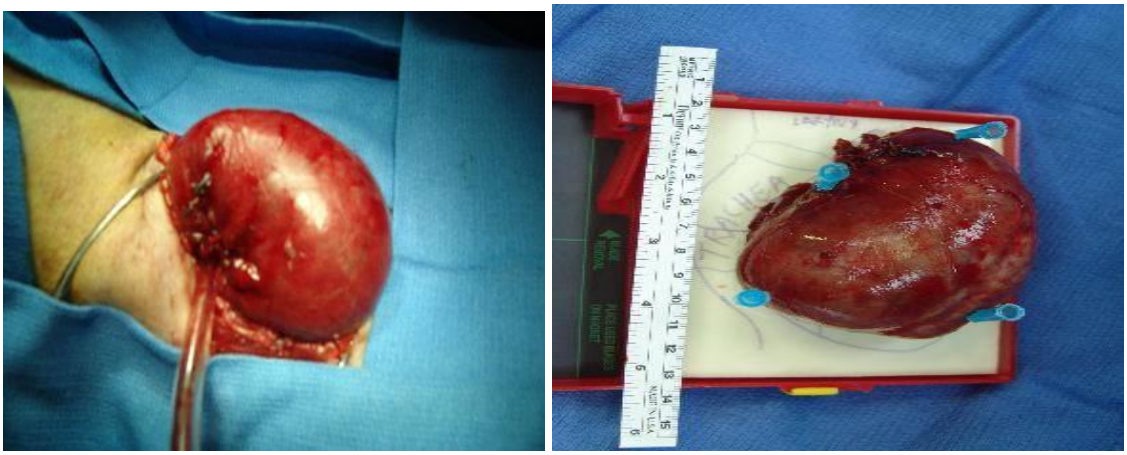

Figure 2 (a,b). Thyroid gland as surgical specimen after thyroidectomy.

Discussion

The term goiter refers to an enlargement of the thyroid. The clinical manifestations of goiter vary with the size and location of the goiter and can be potentially serious (1).
In patients with substantial enlargement of one lobe or asymmetric enlargement of both lobes, the trachea, esophagus, or blood vessels may be displaced or, less often, compressed (2). Bilateral lobar enlargement, especially if the goiter extends posterior to the trachea, may cause

either compression or concentric narrowing of the trachea or compression of the esophagus or jugular veins (3).

With some goiters, there is growth of one or both lobes through the inlet into the thoracic cavity which can result in obstruction of any of the structures in the inlet. Such goiters are called retrosternal. Retrosternal goiter was first described by Haller in 1794. Retrosternal goiter incidence varies widely depending on its definition but can be found in up to 45% of all thyroidectomies (4). Diagnosis is more common in the fifth or sixth decade of life with a female to male proportion approximately 3–4:1 (1) and have a familial component in up to 30% of cases (5).

Most retrosternal goiters are in the anterolateral mediastinum, (6) but about 10 percent are located primarily in the posterior mediastinum (7). Some patients have very low-lying thyroid glands located at the level of the thoracic inlet; in them even minimal enlargement can cause symptoms.

Etiologically, 85%–95% of retrosternal thyroid masses emerge as benign goiter (3). However, the incidence of thyroid cancer in large series of patients with retrosternal goiter has ranged from 10 to 16 percent (4,6).

Among benign retrosternal goiters, the following causes were noted in one large series (7):

- Multinodular goiter — 51 percent
- Large follicular adenoma — 44 percent
- Chronic autoimmune thyroiditis — 5 percent

The majority of patients with obstructive cervical goiters have had a visible goiter for many years. Most patients with retrosternal goiter (77 to 90 percent in two series) also have visible goiters (7,8) .

The most common complaint in patients with obstructive cervical or retrosternal goiter is exertional dyspnea, which is present in 30 to 60 percent of cases (5,7,9) . This symptom usually occurs when the tracheal diameter is under 8 mm (10). In some patients with substernal goiter, dyspnea is primarily positional or nocturnal, and it occurs primarily during maneuvers that force the thyroid into the thoracic inlet such as reaching and bending (11). When tracheal compression becomes severe (luminal diameter less than 5 mm), stridor or wheezing occurs at rest (10,11). This upper airway wheezing must be distinguished from asthma. An upper respiratory illness may exacerbate upper airway obstruction.

Obstructive goiter rarely, can induced jugular vein compression or thrombosis, cerebrovascular steal syndromes, or even the superior vena cava syndrome (12,13). However, retrosternal goiter as a cause is extremely rare. The incidence of retrosternal goiter causing SVC syndrome is 3.2% (14) and may be asymptomatic for a long period due to the slow but steady growth of the gland on account of venous collateral development (15). Compression of the superior vena cava by a mediastinal malignancy is the most common cause of SVC syndrome (16).

Substernal goiters are often seen on chest x-ray as a mass that causes tracheal narrowing or deviation or as superior mediastinal widening. However, the extent of the goiter and its effect upon surrounding structures can be assessed better by computed tomography (CT) or magnetic resonance imaging (MRI) (5,12,17). Thyroid ultrasonography, although more accurate than CT for defining thyroid anatomy in the anterior neck, is not satisfactory for imaging of posterior neck structures or the substernal region.

Surgery is the treatment of choice for any goiter causing obstructive symptoms, and for significant retrosternal goiters whether or not obstructive symptoms are present (7,12,15).

The majority of obstructive and retrosternal goiters can be excised through a standard collar incision. However, to remove a very large retrosternal goiter, or invasive cancer, partial or complete sternotomy or even thoracotomy may be required (12).
Radioiodine (131-I) may be useful as nonsurgical treatment for obstructive or retrosternal goiter. Despite the apparent success of radioiodine, surgery is still the treatment of choice because of concern that radiation thyroiditis might result in worsening of airway obstruction and the possibility of carcinoma if the goiter is mostly substernal (17). Radioiodine may be useful in patients who refuse surgery or are poor surgical candidates, particularly if the retrosternal or obstructive goitrous tissue is functional on thyroid radionuclide imaging.

As conclusion, based on the presented case, in patients with SVC syndrome should have a careful thyroid exam and early diagnosis and proper treatment of SVC syndrome due to thyroid pathologies may prove to be lifesaving.

Conflict of interest: The authors have no conflicts of interest to declare.

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