Retrosternal goiter with posterior mediastinal extension surgery in a patient with superior vena cava obstruction through a classic neck incision; Case report

Peer review status:
No

Corresponding Author:
Dr. Attiya Al-zahrani,
Consultant in chief, Department of General Surgery, Al-Hada Armed Forces Hospital, Al-Hada Armed Forces Hospital, Taif 21947
Saudi Arabia - Saudi Arabia

Submitting Author:
Dr. Mohammad Othman,
Consultant Obstetrician and Gynaecologist, King Abdullah Medical City, Maternity and Children Hospital, Madinah, Saudi Arabia, 84 Bradfield Road, M32 9LE - United Kingdom

Other Authors:
Dr. Samah Khayat,
Surgical resident, Department of General Surgery, Al-Hada Armed Forces Hospital, Al-Hada Armed Forces Hospital, Taif 21947
Saudi Arabia - Saudi Arabia
Dr. Alaa Al-Juaid,
Surgical resident, Department of General Surgery, Al-Hada Armed Forces Hospital, Al-Hada Armed Forces Hospital, Taif 21947
Saudi Arabia - Saudi Arabia
Dr. Arif Khorchid,
Senior registrar, Department of General Surgery, Al-Hada Armed Forces Hospital, Al-Hada Armed Forces Hospital, Taif 21947
Saudi Arabia - Saudi Arabia
Dr. Basem Othman,
Minimal invasive surgery consultant, General Surgery Department, Al-Hada Armed Forces Hospital, Al-Hada Armed Forces Hospital, Taif 21947
Saudi Arabia - Saudi Arabia

Article ID: WMC004890
Article Type: Case Report
Submitted on: 08-May-2015, 08:51:06 AM GMT Published on: 08-May-2015, 12:02:46 PM GMT
Article URL: http://www.webmedcentral.com/article_view/4890
Subject Categories: SURGERY
Keywords: Retrosternal Goiter, Substernal Goiter, Goiter, Intrathoracic Goiter, Thyroidectomy, Sternotomy, Cervicothoracotomy

How to cite the article: Khayat S, Al-Juaid A, Khorchid A, Othman M, Othman B, Al-zahrani A. Retrosternal goiter with posterior mediastinal extension surgery in a patient with superior vena cava obstruction through a classic neck incision; Case report. WebmedCentral SURGERY 2015;6(5):WMC004890
Copyright: This is an open-access article distributed under the terms of the Creative Commons Attribution License (CC-BY), which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Source(s) of Funding:
None

Competing Interests:
None Known

Additional Files:
Figure 1
Figure 2
Figure 3
Figure 4
Figure 5
Retrosternal goiter with posterior mediastinal extension surgery in a patient with superior vena cava obstruction through a classic neck incision; Case report

Author(s): Khayat S, Al-Juaid A, Khorchid A, Othman M, Othman B, Al-zahrani A

Abstract

Intrathoracic goiters are usually located anteriorly, in the superior or anterior mediastinum, and are termed substernal or retrosternal goiters. Posterior mediastinal goiters are rare, about 10% of all intrathoracic goiters. Patients with retrosternal goiter usually have a visible or palpable cervical mass. In addition, tracheal deviation may be present with compression symptoms.

Posterior mediastinal goiter with mediastinal compressive symptoms is an indication of surgery. Lateral thoracotomy is an alternative approach for intrathoracic goiter extending into the posterior mediastinum.

We report a case of 75 years old gentleman, referred with right neck swelling, difficulty in swallowing and breathing and changes of voice. Total thyroidectomy was performed. Histopathology report showed multi nodular goiter with no evidence of malignancy. This patient had both types of extension. Both were delivered successfully through a classic neck incision without sternotomy or combined cervicothoracotomy. It would be reasonable to consider surgical management for such symptomatic goiters if there were no contraindications.

Introduction

Retrosternal goiter was first described by Haller in 1749 [1, 2]. From that time, there were controversies concerning its definition. The most common definition was proposed by deSouza and Smith, as that in which more than 50% of the total bulk of thyroid tissue resides below the thoracic inlet [1, 3, 4].

The intrathoracic thyroid adenoma or goiter is mostly located in the anterior mediastinum, about 8%-15% are in the posterior mediastinum [1, 3, 5, 6]. It is derived from embryonic thyroid tissue and developing into isolated thyroid tumor within the mediastinum or descending into the retrosternal loose tissue space from neck, which may cause various compressive symptoms when it reaches a certain size [1, 7, 8]. Most of the anterior mediastinal goiters can be removed by a transcervical approach, but posterior mediastinal goiters may require additional extracervical incisions [1, 4, 9].

Retrosternal goiter refers to the thyroid mass grows along dermal sternum from the neck to the substernal portion, descending below the thoracic inlet [4, 6]. It is characterized by slow progression and a longer course of illness [4, 5]. If the substernal goiter compresses the adjacent esophagus, trachea, nerves and blood vessels, then the corresponding symptoms would occur [4, 5, 8, 9]. These symptoms included anhilation and wheezing secondary to tracheal compression, superior vena cava syndrome caused by superior vena cava compression, hoarseness caused by recurrent laryngeal nerve compression, and Horner syndrome caused by peripheral adrenergic nerve compression [2, 10, 11]. Some patients may be asymptomatic, and the abnormalities were detected by physical examination [3, 6, 12].

Retrosternal nodular goiter usually results from simple goiter [5, 6, 13]. Although bilateral glands are often involved, the large lesions are usually located in unilateral gland [3, 12, 14]. Large substernal nodular goiter often causes compression of surrounding structures, secondary hyperthyroidism and malignant changes [3, 4, 12, 14]. Therefore, surgery will be indicated when the diagnosis is confirmed. However, if it is treated with surgery, the operative bleeding risk was high [4, 6]. Most of the cases are operated upon via a cervical or combined cervical-thoracic approach. Substernal goiter resection performed through cervical approach is minimally invasive with less potential complications [1, 4, 6]. The patients don’t require thoracotomy and rehabilitate fast postoperatively. In contrast, combined cervical-thoracic approach pose more risk of intraoperative damages and complications, as well as slower postoperative rehabilitation [1, 4, 6, 8].

The mediastinum constitutes a compartmentalized septum or partition that vertically divides the thorax [9,
It is anatomically bound on the lateral side by the parietal pleural reflections along the medial aspects of both lungs, superiorly by the thoracic inlet, inferiorly by the diaphragm, anteriorly by the sternum, and posteriorly by the anterior surfaces of the thoracic vertebral bodies [9, 11, 13, 16].

Swallowing, gravity and thoracic negative pressure help the growing goiter direct into the chest cavity [4, 9]. Anatomically speaking, goiter in the chest cavity generally grows to the position of relatively low resistance. At first, the tumor will grow into the anterior superior mediastinum between trachea and sternum, forming the common retrosternal thyroid goiter [1, 4, 6]. Because there are thymus (which may atrophy), left and right brachiophchalpic veins and superior vena cava in the front, aortic arch and its three branches (phrenic nerve and vagus nerve have smaller resistance) in the middle left of retrosternal space, tumor growth will be resisted there [1, 4]. Right posterior mediastinum has relatively low resistance than left posterior mediastinum, and it helps form right posterior mediastinal goiter. The primary intrathoracic goiter only accounts for 0.2 ~ 1% of all the intrathoracic goiters, it affects females more often (male: female = 1 : 3 or 1 : 4). Its causes are totally different from the ones of secondary intrathoracic goiter [1, 4, 9, 12]. During the embryonic developmental period of thyroid gland, part or all of the thyroid blastoma leaves primordium and is pulled into the thoracic cavity by the descendent heart and great vessels, then continues to develop in the thoracic cavity, forming the final primary intrathoracic goiter [1, 2, 7, 12]. Because of different originations, secondary posterior mediastinal goiter is often continued with the cervical thyroid gland, with blood supply from inferior thyroid artery and its branches while primary posterior mediastinal goiter maintains little or no connection with the cervical thyroid gland, and has a blood supply derived from intrathoracic arteries [3, 7, 12].

Radiographic image is the most effective and necessary diagnostic method for intrathoracic goiter[1, 13]. CT scan is the most common one for preoperative evaluation. On CT films, intrathoracic goiter usually manifests as a clear boundary mass, its density varies due to the amount of iodine contained: when the amount of iodine in the mass is low, its density is close to the soft tissue of chest wall, and when the amount of iodine is high, its density could be greatly higher than soft tissue [1, 4, 9, 13]. In addition, its density can be uneven due to colloid cysts and calcified plaque. Radionuclide scan is also one of the common diagnostic methods, but it is not so effective when compared with its usage in thyroid goiter of other regions because the intrathoracic goiter does not always uptake iodine [1, 5, 7, 12].

Fine-needle aspiration cytology to exclude the presence of carcinoma is not recommended because intrathoracic goiters are not easily accessible and the procedure may lead to life threatening complications [1, 3, 7]. When mediastinal mass lesions are diagnosed using imaging techniques, image interpretation requires accurate assessment of the lesion origin, area of existence, extension and inner structures [3, 4, 6, 7].

The surgical approach used to gain access to RSG is commonly based on intraoperative findings [1, 7]. The use of CT scanning to aid in this decision has always been limited, but now, it is gaining popularity among many surgeons. Several have emphasized on the importance of CT in preoperative planning of surgery for RSG. Extension to the level of the aortic arch and beyond was found to greatly influence the need for a thoracic approach [1, 2, 7, 10].

The differential diagnosis of intrathoracic goiter is of great variety, it should be differentiated from lymphadenopathy, branchial cleft cyst, arterial aneurysm, neurogenic tumour, pheochromocytoma, spinal cord injury, hiatus hernia, etc [1, 7, 9].

Case Report

75 years old Saudi gentleman, presented to endocrine surgery clinic in Al-Hada Military Hospital, Taif, Saudi Arabia. Patient known to have bronchial asthma, he was referred to the with right neck swelling, difficulty in swallowing and breathing and changes of voice. On examination, the face was congested with dilated with congested neck and upper chest veins (Fig. 1). Air entry was decreased on the right side of the chest but there was no tracheal deviation. There was a 5x6 cm lump on the right side of the neck which was mobile with swallowing. Biochemically, he was euthyroid. Chest X-ray (Fig.2) showed a well-defined huge mediastinal soft tissue dens lesion with tracheal compression. CT with contrast showed heterogeneous bilateral neck masses. The right side mass was 7x5 cm pushing the pharynx to the left and the right carotid laterally (Fig. 3). Added to that, the left side mass was 11x6 cm with intra-thoracic extension down to the carina compressing and pushing the trachea to the right side (Fig.3). There was compression on the superior vena cava (Fig. 4) with multiple collaterals on the upper anterior chest wall. There was no cervical or mediastinal lymphadenopathy. Total thyroidectomy was performed through a classical transverse cervical...
symptoms may be more acute, and the patient's
duration of symptoms may range from
potentially fatal tracheal obstruction. Rarely, jugular
hemorrhage into the goiter which may cause sudden
compression of the cervical sympathetic chain may be
present too, and may be indicative of a malignant
dysphagia results from
primarily due to maneuvers that force the thyroid into
facial flushing and choking on lying down, occurs
common symptoms. A positive Pemberton's sign,
initially, many patients are asymptomatic, but later
obstructive symptoms and signs may develop, due to
compression and displacement of trachea, bronchi,
esophageal compression [1, 2, 4, 9]. Features of
phrenic nerve paralysis, Horner's syndrome due to
obstructive symptoms and signs may develop, due to
progressive enlargement of the
gland, the path of least resistance is inferiorly into the
thorax [1, 5, 7, 9]. The weight of the goiter as well as
normal swallowing and respiratory motion favors
mediastinal displacement [1, 9]. They are usually
located anteriorly, in the superior or anterior
mediastinum, and are termed substernal or
retrosternal goiters. Their incidence in the general
population is about 1:5000. Retrosternal goiters are
seen in 5-16% of all thyroidectomies. Most of them are
benign, although thyroid cancer is identified in a small,
but definite number (2.5-15%) of cases. Posterior
mediastinal goiters are rare, comprising only about
10% of all intrathoracic goiters [1, 4, 9].

Initially, many patients are asymptomatic, but later
obstructive symptoms and signs may develop, due to
compression and displacement of trachea, bronchi,
esophagus or large veins [1, 7, 9, 13]. Patients with
retrosternal goiter usually have a visible or palpable
cervical mass on presentation. In addition, tracheal
deveiation may be present. Exertional, nocturnal or
positional dyspnea is the most common complaint,
seen in 30-60% of cases [2, 11, 16]. Stridor, wheezing,
cough, dysphonia or hoarseness as a result of
recurrent laryngeal nerve compression are other
common symptoms. A positive Pemberton's sign,
facial flushing and choking on lying down, occurs
primarily due to maneuvers that force the thyroid into
the thoracic inlet. A variety of other symptoms can be
induced by obstructive goiter. Dysphagia results from
esophageal compression [1, 2, 4, 9]. Features of
phrenic nerve paralysis, Horner's syndrome due to
compression of the cervical sympathetic chain may be
present too, and may be indicative of a malignant
process. Occasionally, patients suffer acute
hemorrhage into the goiter which may cause sudden
potentially fatal tracheal obstruction. Rarely, jugular
vein thrombosis, cerebrovascular steal syndrome or
even superior vena cava syndrome have also been
reported. The duration of symptoms may range from
months to decades. The onset of obstructive
symptoms may be more acute, and the patient's
dysphagia may result in recurrent aspiration
pneumonia [4, 9, 12]. The prevalence of
hyperthyroidism ranges from 0% to nearly 50%.
Posterior mediastinal goiters should be differentiated
from other mediastinal masses by appropriate work-up.
Thyroid function tests must be measured in any
patient with a goiter or mediastinal mass suspected to
be enlarged thyroid. Substernal goiters can be seen
on chest x-ray as a superior mediastinal widening,
often unilateral, with or without tracheal deviation or
narrowing. Cervical and thoracic computed
tomography is the most valuable imaging technique for
evaluating mediastinal and cervical masses and
diagnosing enlarged thyroid as the cause of that mass
[1, 2, 9, 12]. On CT, mediastinal goiter should show
high attenuation values due to iodine content, similar
to normal thyroid. Nodular elements may show
combinations of hypodensity and calcification [3, 7].
The mediastinal goiter is usually continuous with the
thyroid tissue seen in the neck. Iodine-123 agents should not be given routinely due to probability of
inducing or exacerbating hyperthyroidism in this
category of patients [7]. If contrast agent
administration is required, a patient with subclinical or
overt hyperthyroidism should be prepared by
antithyroid drugs to prevent thyroidal iodine
organization. Thyroid ultrasound is not as accurate in
the retrosternal region as in the anterior neck because of
inaccessibility to the ultrasound transducer.
Although thyroid radionuclide imaging with iodine-123
may define areas of autonomous function in large
cervical goiters [2, 7]. It is not so useful or even
misleading in patients with intrathoracic goiter,
because some of them take up radioidine poorly, and
the radioactivity is attenuated by interference from the
sternum, clavicles, mediastinum tissue and blood pool.
Pulmonary function tests, spirometry with flow-volume
loops, may be abnormal even when the patient is
asymptomatic. A barium esophagogram may be
helpful in confirming esophageal compression from a
goiter as the cause of dysphagia [2, 7, 9]. Surgical
selective approach for excision of posterior
mediastinal goiters is now recommended by most
surgeons for symptomatic obstructive goiters. Many
patients require surgery for symptomatic relief and to
reduce the risk of acute exacerbation of compressive
symptoms from hemorrhage or inflammation of the
intra-thoracic gland. Observation rather than surgery is
recommended for elderly asymptomatic patients who
are not a good candidates for surgery, with slow
growth, especially if inferior edge of mass is above
brachiocephalic vein [4, 6, 7, 9, 12].
The typical anterior substernal goiter can almost
always be removed through a cervical incision,
although a partial or complete sternotomy may facilitate removal of a large gland [1, 9]. Although many posterior mediastinal goiters can be removed through a cervical incision, most surgeons have adopted a selective approach. Thoracotomy or a combined cervicothoracic approach has been advocated for larger lesions, when the mass is mainly intrathoracic with little or no cervical component or when the mass is malignant [1, 2, 9].

**Conclusion**

Posterior mediastinal goiter with mediastinal compressive symptoms is an indication of surgery. Lateral thoracotomy is an alternative approach for intrathoracic goiter extending into the posterior mediastinum making presentation atypical. Posterior mediastinal goiter can be differentiated from other posterior mediastinal masses by appropriate investigation, while computed tomography is the most valuable technique that may facilitate earlier diagnosis. In our case, the patient had both types of extension; retrosternal on the right side and to the posterior mediastinum on the left side, both were delivered successfully through a classic neck incision without sternotomy or combined cervicothoracotomy. It would be reasonable to consider surgical management for such symptomatic goiters if there were no contraindications.

**References**


