Primary Retrosternal Goiter Mimicking as Teratoma of Anterior Mediastinum: A Diagnostic Enigma

Ramana Reddy Naru, Manilal Banoth, Mohammed Basheeruddin Inamdar, Aruna K. Prayaga[1]

Departments of Surgical Oncology and [1]Pathology, Sri Venkateswara Institute of Medical Sciences, Tirupathi, Andhra Pradesh, India

Abstract

Primary intrathoracic goiters arising from aberrant thyroid tissue are rare, representing <1% of all retrosternal goiters (RGs). We report a rare case of primary RG in a male patient with symptoms of mediastinal mass lesion alone. By careful examination and with the help of radiological imaging, we could pick up a thyroid swelling with a mediastinal mass which led to a diagnostic dilemma, as the thyroid swelling was reported as a multinodular goiter and the mediastinal mass was misdiagnosed as teratoma. We subjected the patient to a functional imaging with a thyroid scintigraphy, which revealed a thyroid swelling with retrosternal extension that was managed by surgery (total thyroidectomy and mediastinal mass excision) with only a Kocher's neck crease incision.

Keywords: *Ectopic thyroid, Kocher's incision, mediastinal mass, nuclear imaging, primary retrosternal goiter, thyroidectomy*

Introduction

Retrosternal goiter (RG) was first described by Albrecht von Haller in 1749 as the extension of the thyroid tissue below the upper opening of the chest. Since then, RG has always been considered a challenge for the surgeons, because of the difficulties that may be encountered during surgical removal.[1] The definition of RG is still not precise and varies between different authors. Describing RG as substernal or retrosternal when ≥50% portion of the mass is located in the mediastinum is the most commonly accepted definition. The presence of RG is documented in 2%–19% of all thyroidectomies.[2] This wide range in the incidence is mainly due to the variation in the definition of RG. Primary intrathoracic goiters arising from aberrant thyroid tissue are rare, representing <1% of all RGs.[3] Its progressive extension may lead to compression of the surrounding structures such as trachea, esophagus, nerves, and great vessels, which leads to disastrous symptoms such as dyspnea, dysphagia, hoarseness of voice, and facial plethora. An identified RG is an absolute indication for surgery.

We report a rare case of primary RG in a male patient with symptoms of mediastinal mass lesion alone. By careful examination and with the help of radiological imaging, we could pick up a thyroid swelling with a mediastinal mass which led to a diagnostic dilemma, as the thyroid swelling was reported as a multinodular goiter and the mediastinal mass was misdiagnosed as teratoma. We subjected the patient to a functional imaging with a thyroid scintigraphy, which revealed a thyroid swelling with retrosternal extension that was managed by surgery (total thyroidectomy and mediastinal mass excision) with only a Kocher's neck crease incision.

Case Report

A 63-year-old gentleman presented to the General Medicine Department with dry cough and occasional dysphagia of 1-month duration. On imaging, a thyroid swelling extending into the superior mediastinum was detected. There were no features of hypothyroidism and hyperthyroidism. There was no history of radiation exposure and no family history of thyroid disorders or swellings. Examination revealed a vague nodule on the right side in the region of thyroid measuring 2 cm × 2 cm, and the lower border was not clinically palpable. Chest X-ray revealed a widening of superior mediastinum. Computed tomography (CT) scan of the chest revealed a heterogeneously enhancing multinodular mass lesion measuring 9.8 cm × 4.9 cm × 6.5 cm

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Address for correspondence:
Dr. Ramana Reddy Naru, Department of Surgical Oncology, Sri Venkateswara Institute of Medical Sciences, Tirupathi - 517 501, Andhra Pradesh, India.
E-mail: ramanareddynaru77@gmail.com

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in the neck anterior and lateral to trachea. The mass was extending inferiorly into the superior and middle mediastinum, and calcifications were noted within. Trachea was grossly deviated to the right. The lesion was splaying the origin of right brachiocephalic trunk and left common carotid artery which had a common origin. The mass was also compressing the arch of aorta, and precarinal lymph nodes were also present measuring 2.2 cm × 1.2 cm. CT scan impression was given as teratoma mediastinum with multinodular goiter of thyroid [Figure 1]. As we strongly suspected a thyroid swelling with a retrosternal extension, the patient was advised a functional imaging to rule out the presence of two separate disease entities. We proceeded with ^{99}M-technetium-thyroid scintigraphy which did not show a significant uptake in the thyroid bed, which was suggestive of subacute thyroiditis. Furthermore ^{99}M-technetium sestamibi (MIBI) scintigraphy revealed a mild increased radio tracer uptake in both lobes of thyroid gland, and the mediastinal part documented on CT scan also showed uptake of MIBI [Figure 2]. Impression was subacute thyroiditis in a case of multinodular goiter with a retrosternal extension into the anterior mediastinum. Fine needle aspiration (FNA) cytology from neck swelling revealed an adenomatous goiter with thyroiditis. Thyroid profile and other blood investigations were within the normal limits.

Surgery was undertaken with a consent for sternotomy if needed but with only a neck crease (Kocher's) incision. A total thyroidectomy and superior mediastinal mass excision were done. Intraoperative findings were multinodular goiter involving both lobes of thyroid with the largest nodule of size 3 cm × 3 cm in the left lobe with a retrosternal extension and we found a separate mediastinal mass which was not continuous with the thyroid swelling of size 6 cm × 8 cm present in the superior mediastinum. The final histopathology from both thyroid and mediastinal mass revealed a similar microscopic picture and an impression of adenomatous goiter was given [Figures 3 and 4]. Postoperative period was uneventful. The patient was discharged on day 4.

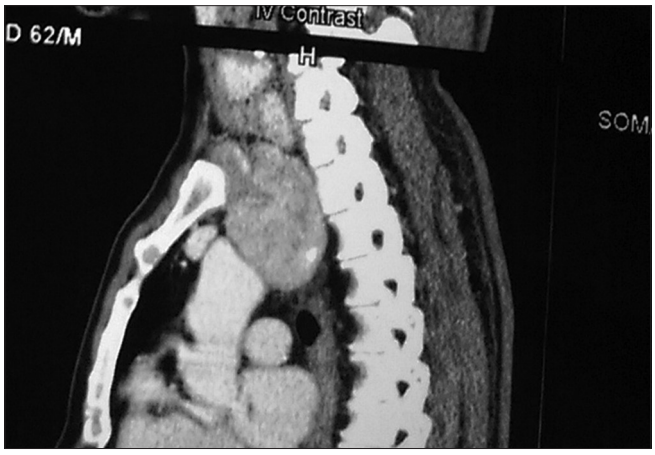

Figure 1: Sagittal section of computed tomography scan showing multinodular goiter and a separate large retrosternal mass with calcification

Discussion

Mediastinal ectopic thyroids with no connection to normal thyroid gland are very rare, accounting for <1% of all cases.[3] The definition of RG is still not precise and varies between different authors. Describing RG as substernal or retrosternal when ≥50% portion of the mass is located in the mediastinum is the most commonly accepted definition. The presence of RG is documented in 2%–19% of all thyroidectomies.[2] This wide range in the incidence is mainly due to the variation in the definition of RG. Diagnosis of RG is most frequently made in the fifth or sixth decades of life, with a female/male ratio of 4:1. RGs can be classified as either primary or secondary. Primary intrathoracic goiters arise from aberrant thyroid tissue which is ectopically located in the mediastinum, receive their blood supply from mediastinal vessels, and are not connected to the cervical thyroid. They are rare, representing <1% of all RGs.[4] Secondary RGs develop from the thyroid located in its normal cervical site. These secondary RGs are, characteristically, in continuity with the cervical portion of the gland and receive their blood supply, depending on cervical vessels, almost always through branches of the inferior thyroid artery. Therefore, for the patients to be operated for substernal goiter, the differentiation of primary and secondary is clinically important.

About 20%–40% of RGs are symptomatic. The most common symptoms are related to compression of the airways and the esophagus and are represented by dyspnea, choking, inability to sleep comfortably, dysphagia, and hoarseness. The diagnosis of RG is based upon clinical history, clinical examinations, and imaging findings. Chest X-ray, CT scan, and magnetic resonance imaging examination are beneficial in preoperative evaluation of

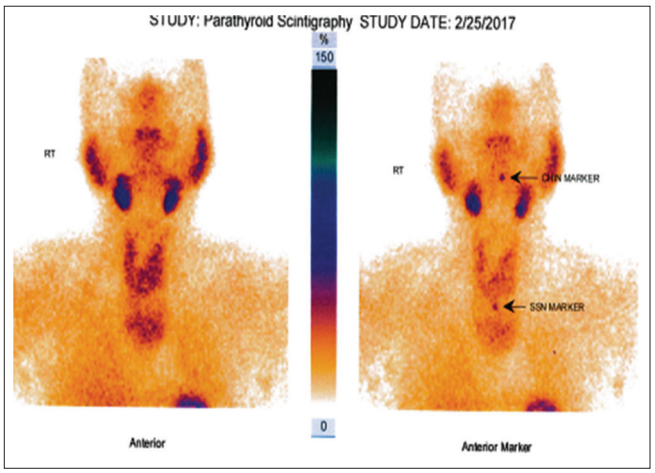

Figure 2: Scintigraphy with ^{99}M-technetium sestamibi scan revealed uptake in the thyroid gland and a second noncontiguous area of similar uptake confirming an ectopic retrosternal thyroid

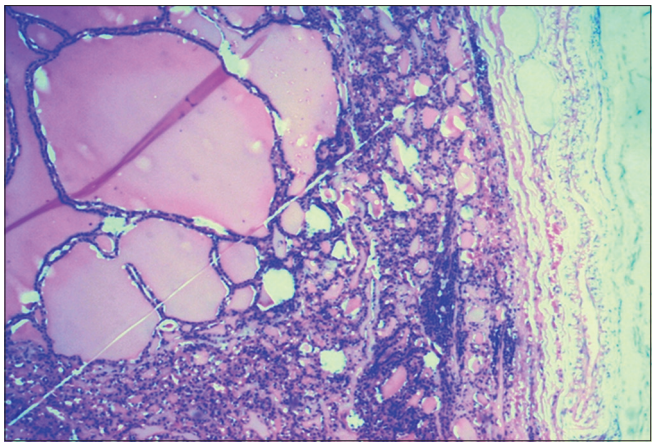

Figure 3: Microphotograph (×4) from thyroid swelling showing multiple thyroid follicles of varying sizes filled with colloid

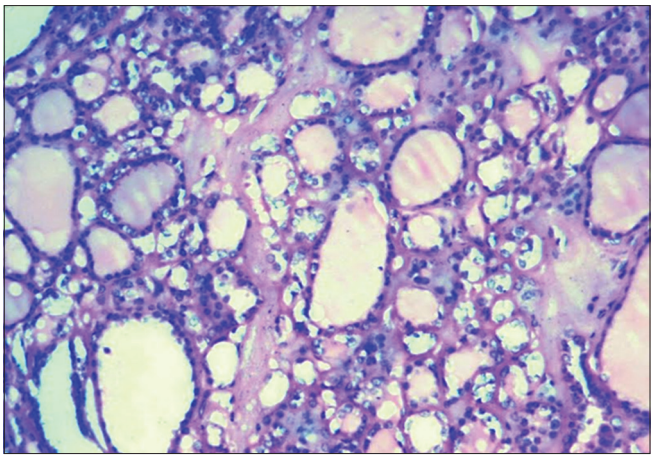

Figure 4: Microphotograph (×10) of the mediastinal mass also showing colloid-filled thyroid follicles with its cuboidal lining epithelium

patients thought to have retrosternal thyroid tissue.[5] CT scan is considered as the imaging modality of choice for RGs. Thyroid scintigraphy with I-131 or ^{99}M-technetium is highly sensitive and specific for detecting normal and ectopic thyroid tissues.

An FNA of a RG may be helpful when a large cervical component exists, but this is not usually recommended, because it is technically difficult, may be dangerous to perform, and can miss the true pathology.

The surgical strategy for treating RG is somewhat different from the one used for the goiter due to anatomic and physiologic dissimilarities. This makes retrosternal thyroidectomy technically challenging. The surgery is technically demanding, with greater associated chances of injury to native structures. Sternotomy or lateral thoracotomy should be considered once the following situations appear: (1) RG is too large to be removed through thoracic inlet; (2) the RG blood supply originates in the chest; (3) RG growing into the mediastinum causes anatomic variations in the location of the recurrent laryngeal nerve and the parathyroid glands; and (4) venous congestion due to compromised drainage may cause severe bleeding. Based on this, we consent with Neves *et al*.[6] in that most RGs can be resected through a transcervical approach, but those extending beyond the aortic arch into the posterior mediastinum are better dealt with by sternotomy or lateral thoracotomy.

The ideal surgical approach for RG is the subject of significant debate.[7] Most RGs can be removed through a cervical approach, while a partial or total sternotomy should be performed only in a minority of patients, ranging between 1% and 11%.[8] Postoperative complications include recurrent laryngeal nerve injury, airway complications, hematoma, and hypocalcemia. The incidence of these complications is variable between different centers. For recurrent laryngeal nerve injury, it ranges between 2.7% and 5.1%, transient hypocalcemia between 3.4% and 13.3%, and hematoma between 2.7% and 3.4%.[9] Parathyroid devascularization is more common with resection of a substernal goiter and autotransplantation can prevent permanent hypoparathyroidism.[10] Our patient had a smooth postoperative recovery apart from transient hypocalcemia which was managed with oral calcium supplementation.

Conclusion

In conclusion, incidentally picked up thyroid swelling with an RG may present as a diagnostic challenge as initial imaging had given a picture of two separate disease entities but after careful examination workup with radiological and functional imaging and confirmation with postoperative histopathology, it was confirmed as a case of primary RG. Primary RGs have a higher incidence of symptoms related to compression, progression of the disease, difficulty to follow-up, the risk of malignancy, and development of acute airway obstruction, which are all the indications to opt for surgery. In such cases, proper evaluation with radiological imaging, nuclear imaging to overcome the diagnostic difficulty, and a proper surgical planning is needed. Cervical incision alone is nearly always adequate, with negligible operative mortality and minimal complications.

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Conflicts of interest

There are no conflicts of interest.

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