

SUBSTERNAL MULTINODULAR GOITER TOGETHER WITH BRAINSTEM PATHOLOGY: MISLEADING MANIFESTATIONS

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ABSTRACT

This study aimed at analyzing the clinical presentation, diagnosis, management and results of the treatment of substernal multinodular goiter. Substernal goiter with its clinical presentation can be misdiagnosed. When a patient with substernal multinodular goiter is presented with symptoms like dysphagia, hoarseness and dyspnea, while ruling out the possible malignant or benign causes, central cranial system originated pathologies should always be kept in mind.

Key Words: Substernal goiter, Dysphagia, Dyspnea, Hoarseness, Brainstem pathology.

INTRODUCTION

A substernal goiter is the extension of a thyroid goiter into the mediastinum (1). The origin of the substernal goiter is believed to be an extension of the cervical gland into the mediastinum, rather than abnormal growth of a mediastinal-based gland. Evidence for this is that the neurovascular supply to nearly all substernal goiters is from a

cervical rather than a thoracic source (2). The incidence of substernal thyroid gland in the general population is difficult to ascertain. The reported incidence in the general population varies from 0,02% to 0,5% based on chest x-ray screening reports (3,4). In patients undergoing thyroidectomy, series having a substernal component have reported incidences of 1% to 20% (5). Presentation of a patient with substernal thyroid gland typically occurs in fifth and sixth decades; in women the incidence is three to four times greater than in men. (6) Thyroid extension into the mediastinum leads to significant growth potential as limited resistance is encountered. Subsequent impingement on the esophagus and trachea leads to the commonly encountered symptoms of dyspnea and dysphagia. Less commonly, great vessel compression can lead to venous obstruction (7). Surgical excision is the treatment of choice for substernal thyroid gland. Literature reports often stress the need for operative intervention as a result of compressive symptoms and the possibility of thyroid malignancy in a substernal goiter. The evaluation should include chest x-ray, thyroid function tests, indirect laryngoscopy to assess vocal cord function, and CT scan. FNAB may be performed if cervical thyroid tissue is easily accessible (7).

CASE REPORT

A 58-year-old male patient presented with hoarseness, dysphagia and dyspnea over a period of two months. He described his dysphagia as difficulty in swallowing solid food. He also had vomiting episodes after meals. On physical examination both of the vocal cords were edematous, the right vocal cord was paralytic and the left cord had limited mobility. Pyriform sinuses were considered abnormal. A hypernodular thyroid gland with its upper level on the thyroid cartilage on the right and extending substernally on the left was palpated. In differential diagnosis pyriform sinus malignancy, thyroid malignancy and supraglottic larynx carcinoma were considered. His medical history was significant for multinodular goiter, hypertension and a 35-pack-year smoking history. Chest x-ray revealed tracheal deviation to the left. Thyroid function tests were performed. TSH was very low and T3 and T4 were in normal range. The CT images of the neck of the patient revealed an 8 cm. mass compressing the right pyriform sinus externally and a very large multinodular substernal goiter. (Fig. 1). A radionuclide thyroid scan was also performed and it revealed multinodular goiter with nodules of heterogenous activity. (Fig. 2) Fine needle aspiration of the significant thyroid nodule was negative. In the patient's pharyngoesophagography x-ray studies, it was seen that the right lateral pharyngeal wall, the right pyriform recess, the vallecula and the aryepiglottic fold were deformed (Fig. 3).



Fig. 1: CT scan of the presented case revealing compression of the right pyriform sinus externally and a very large multinodular substernal goiter.

Direct laryngoscopy and total thyroidectomy to the right and subtotal to the left were performed. In direct laryngoscopy all the laryngeal structures and the pyriform sinus mucosa were observed as normal, without evidence of any pathology. During the surgical procedure frozen sections of the right thyroid lobe and the isthmus were examined and the histopathologic result was calcified thyroid tissue with hyalen nodules. After the operation, the patient's symptoms diminished

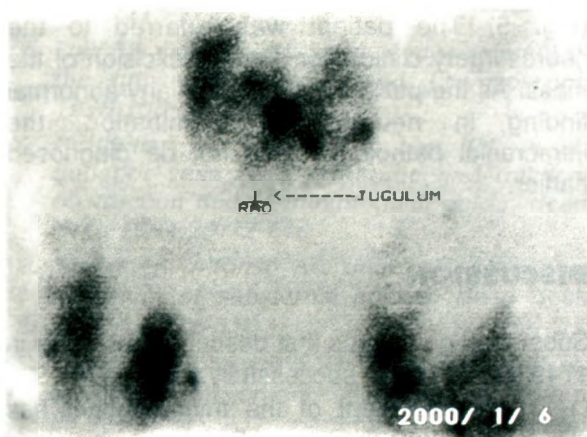


Fig. 2: Radionuclide thyroid scan of the presented case revealed multinodular goiter with nodules of heterogenous activity.

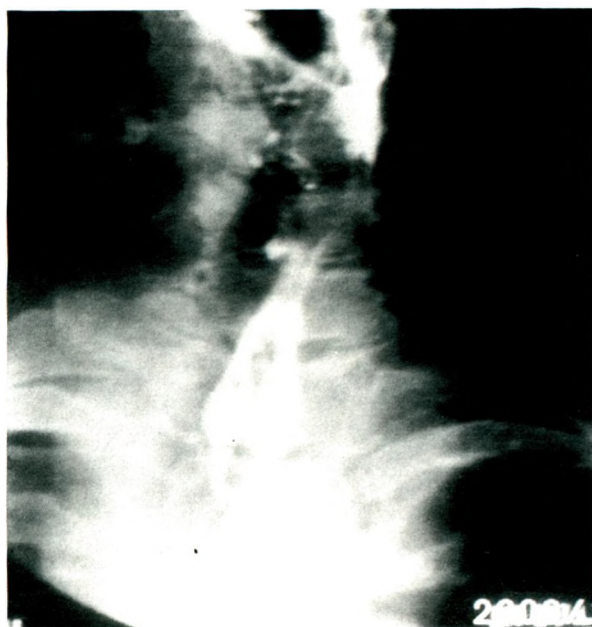


Fig. 3: Preoperative pharyngoesophagography of the presented case. The right lateral pharyngeal wall, the right pyriform recess, the vallecula and the aryepiglottic fold are deformed.

but did not disappear completely. Especially dysphagia, nasopharyngeal regurgitation, aspiration symptoms became more prominent. Postoperative control pharyngoesophagography studies showed no difference compared to the preoperative ones (Fig. 4). Aspiration pneumonia arose in the patient tenth day postoperatively, parenteral antibiotic treatment started. During this period the patient started to have complaints of imbalance and headache. In the cranial MRI studies, a brainstem mass recognized which explained the latest and persisting symptoms. (Fig. 5) The patient was referred to the neurosurgery clinics, for surgical excision of the mass. As the patient did not have any abnormal finding in neurological examination, the intracranial pathology could not be diagnosed earlier.

DISCUSSION

Substernal goiter was first described by Haller in 1749 and removed successfully by Klein in 1820 (1-11). Management of the thyroid goiter has evolved through the ages. Surgical excision is the treatment of choice (9). A substernal goiter is

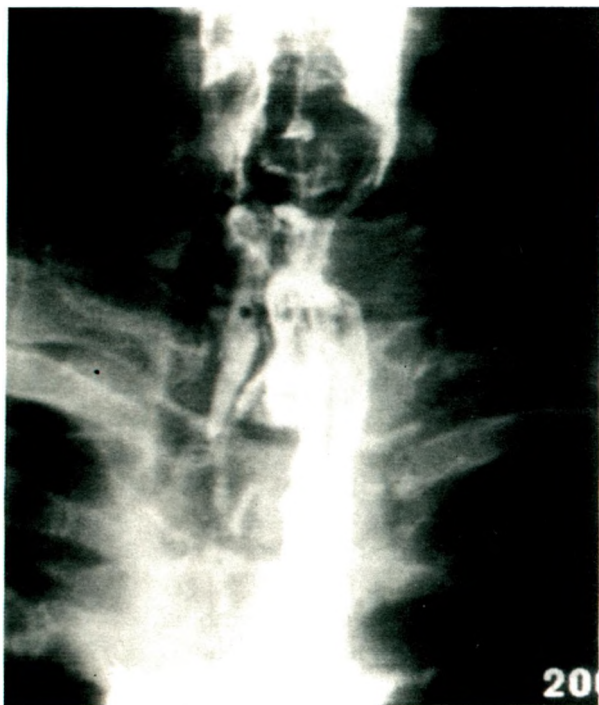


Fig.4: Postoperative pharyngoesophagography of the presented case showing no difference compared to the preoperative pharyngoesophagographies.

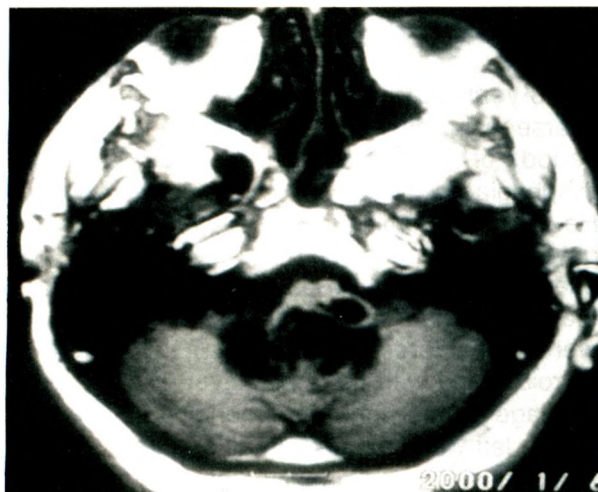


Fig.5: Cranial MRI of the presented case revealing a brain stem mass was recognized explaining the latest and persisting symptoms.

significant for its compressive symptoms. Impingement on the esophagus and trachea leads to commonly encountered symptoms of dyspnea and dysphagia. Less commonly, great vessel compression can lead to venous obstruction (7). The compressive symptoms are secondary to the pressure from an expanding thyroid mass. Because of the bony confines of the thoracic inlet and the limited space of the upper mediastinum, the thyroid compresses the trachea much more readily and causes luminal compromise with a smaller mass, when compared with a cervical goiter (8). Compression from a mediastinal mass tends to affect the trachea before affecting the esophagus (11). Review of the literature puts the risk of malignancy in substernal goiter between 6% and 21% (9-10). The evaluation of a substernal goiter should routinely include a chest x-ray, CT scan of the neck, thyroid function tests, indirect and direct laryngoscopy. Fine needle aspiration biopsy may also be performed. Chest x-ray and CT scan are the only reliable radiographic modalities that help to define the presence and extent of the disease. On the chest x-ray, tracheal deviation is the most often documented finding. Although the vast majority of patients are euthyroid, thyroid function tests should be obtained to identify patients at risk for perioperative thyroid storm. Laryngoscopy both direct and indirect should be performed to assess vocal cord function and to rule out other possible malignant conditions. If a lesion is easily

accessible or other risks for thyroid cancer exist, FNAB may be performed on the enlarged portion of the thyroid gland (6).

In patients with tracheoesophageal compression, surgical excision is the treatment choice (12). But before excision central nervous system originated pathologies should also be remembered and ruled out. As in the current case presented here a multinodular goiter may mask a central pathology with its compressive symptoms, hoarseness and dysphagia. Brain stem pathologies also cause symptoms similar to the compressive symptoms of substernal goiter. Although coincidence of these two different entities is unusual, it may happen as in the case presented here. Thorough and careful evaluation is very important in such patients. And it should also be remembered that during evaluation of tracheoesophageal compression, direct laryngoscopy is a very important diagnostic modality. Substernal goiter with its clinical presentation can be misdiagnosed. When a patient presents with symptoms like dysphagia, hoarseness and dyspnea, while ruling out the possible malignant or benign causes, central nervous system originated pathologies should always be kept in mind.

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