PEMBERTON'S SIGN AND INTENSE FACIAL EDEMA IN SUPERIOR VENA CAVA SYNDROME DUE TO RETROSTERNAL GOITER

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Abstract

Introduction. Retrosternal goitre enlargement can cause compression of several mediastinal structures, especially the trachea and the superior vena cava. Retrosternal goitre as a cause of superior vena cava syndrome is a rare occurrence. We report the case of a middle aged man that underwent surgery for retrosternal goitre with compression of both innominate veins presenting as superior vena cava syndrome.

Case Presentation. A 50 year old man presented with a 2 year history of cyanosis of the upper limbs, head and neck, marked facial edema, plethora, dyspnea on exertion and choking sensation. Pemberton's sign was present. Computer tomography diagnosed retrosternal goitre at the level of the aortic arch, tracheal compression and important collateral circulation. Endocrine evaluation showed normal thyroid function (fT4 15.8 pmol/L) with low-normal TSH (0.5mU/L), normal calcitonin (<2 pg/mL). The patient underwent successful total thyroidectomy with cervical approach and his symptoms dramatically improved. The facial oedema persisted for the next 3 weeks.

Discussion. Less than 3% of superior vena cava syndromes are secondary to a variety of benign causes. Superior vena cava syndrome caused by slow growing retrosternal goitres is very rare and can be asymptomatic for a long period due to venous collateral development.

Conclusion. Superior vena cava syndrome secondary to retrosternal goitres, a very rare occurrence, is an indication for total thyroidectomy, with low postoperative morbidity and dramatic resolution of symptoms.

Key words: retrosternal goiter, superior vena cava syndrome, Pemberton's sign.

INTRODUCTION

Superior vena cava syndrome describes a clinical scenario resulting from mechanical obstruction that impedes the blood flow through the superior

vena cava. Dyspnoea is a common symptom while characteristic physical findings include venous distension of the neck, facial oedema, plethora and cyanosis (1,2). The signs and symptoms may be aggravated in supine position or by bending forward. More than 95% of cases of superior vena cava syndrome are caused by malignant diseases, while benign causes include granulomatous infections, goitres, aortic aneurysms and thrombus formation (3).

The most common definition for a retrosternal goitre is that the majority of the gland resides within the chest (>50%) (4). No anatomic structure prevents the thyroid gland from growing into the thoracic inlet during swallowing or breathing, due to negative intrathoracic pressure (5). Retrosternal growth of goitres leads to compression and dislocation of adjacent structures. Superior vena cava syndrome occurs in 3.2% of retrosternal goitres and may be asymptomatic for a long period due to the slow but steady growth of the gland on account of venous collateral development (5-7).

CASE PRESENTATION

A 50 year old obese male presented for with a 2 year history of cyanosis of the upper limbs, head and neck, marked facial oedema, plethora, dyspnoea on exertion and choking sensation. Pemberton's sign was present. Clinical examination revealed an enlarged, massive and firm, but painless thyroid gland with lower poles extending inferiorly beyond the clavicles. Routine laboratory tests revealed subclinical hyperthyroidism (fT4 15.89 pmol/L, TSH 0.044 mU/L) and impaired glucose tolerance; during OGTT, blood glucose was 0'-88 mg/dL, 1h - 102 mg/dL, 2h-147 mg/dL. Thyroid ultrasound described a multinodular goiter that extends in the anterior mediastinum. Computer tomography

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(Fig. 1) revealed that the multinodular goiter extends to the aortic arch with compression of the innominate and left subclavian arteries, both innominate veins and the trachea. It also noticed important left periscapular and prepectoral venous collateral circulation with left internal mammary drainage.

The patient was scheduled for surgery. After difficult flexible bronchoscope-assisted tracheal intubation, the patient underwent total thyroidectomy with cervical approach and capsular dissection. Digital mobilization was used for the intrathoracic component of the gland (Fig. 2). Both recurrent laryngeal nerves

were encountered and preserved, the parathyroid gland being preserved *in situ*. Although the gland reached the aortic arch, the cervical approach was comfortable.

The postoperative course was uneventful with dramatic resolution of the symptoms while the facial oedema persisted. The patient was discharged on the second postoperative day. The facial oedema, while reduced, still persisted after 7 days. At three weeks follow-up the facial oedema resolved completely and the patient remained in the care of the endocrinologist (Fig. 3-4). The pathology report revealed a multinodular goitre.

Figure 1. CT scan – compression of both innominate veins.

Figure 2. Surgical specimen.

Figure 3. Before surgery.

Figure 4. Three weeks after surgery.

DISCUSSION

Superior vena cava syndrome in the setting of retrosternal goiter is a very rare occurrence and may be asymptomatic for a long period due to the slow thyroid growth that permits collateral venous development. This is the case of our patient that presented with a two year history of signs and symptoms with evident collateral venous circulation revealed by CT scan.

The Pemberton manoeuvre reported first in 1946 is a useful but overlooked clinical test for latent superior vena cava syndrome caused by a retrosternal mass (8). While keeping both arms elevated our patient developed intense facial plethora and cyanosis in less than 30 seconds. While the chest x-ray may suggest a goitre, the computer tomography study is the gold-standard for pre-operative surgical planning (5).

The diagnosis should be clinically oriented but needs imagistic studies for confirmation. There is no clear consensus regarding the definition of retrosternal goitres. While most authors follow the definition of deSouza and Smith (1983) with more than 50% of the gland below the thoracic inlet, recent papers describe as a retrosternal goitre, any goitre that extends below the thoracic inlet (5). The definition should have an impact on pre-operative planning. For most retrosternal goitres the cervical approach is sufficient and comfortable, others need manubriectomy, full sternotomy or thoracotomy.

The capsular dissection in the avascular plane between the true thyroid capsule and the surgical capsule of the gland, derived from the pretracheal fascia, described and used since the 19th century, is a prerequisite for safe and effective surgery especially in cases of retrosternal goitres (9). The avascular plane developed in the cervical part of the gland is then followed in the mediastinum. In order to digitally mobilize the mediastinal part of the gland, advanced dissection should be carried in the cervical part of the gland and the inferior thyroid pedicle needs to be secured. Although the individualization of the inferior pedicle pleads for a retrosternal goitre without mediastinal blood supply, preoperative imaging studies should clarify the blood supply of the gland beforehand.

The presence of mediastinal vascular supply of retrosternal goitres may impose a thoracic approach. In our case, the mobilization of the mediastinal part of the gland was difficult due to its size and the patient's anatomy – short neck and compression of the gland at the level of the thoracic inlet.

In conclusion, superior vena cava syndrome caused by a retrosternal goiter is a rare occurrence. The syndrome may be asymptomatic or latent for a long period due to the slow growth of the goiter that permits the development of collateral venous circulation. Pemberton's maneuver is useful in such cases to reveal the syndrome. The surgical treatment consists of total thyroidectomy. Resection of retrosternal goiters can be generally accomplished through a cervical approach with minimal morbidity in experienced hands. Capsular dissection in the avascular plane permits safe and successful mobilization of the thoracic part of the retrosternal goiter.

Conflict of interest

The authors declare they have no conflict of interest.

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