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Substernal goitre: a rare cause of pulmonary hypertension and heart failure

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Abstract
Benign substernal goitres usually extend into the upper anterior mediastinum and are easily extractable through a cervical approach. Very infrequently these tumours extend into the thoracic cavity causing compression of mediastinal structures. The authors report a case of pulmonary hypertension and severe cardiac failure secondary to a long-standing substernal goitre, and support the surgical management of this disease.

Key words: Goitre, Substernal; Hypertension, Pulmonary; Heart Failure, Congestive; Surgery

Introduction

Benign substernal goitres rarely cause compression of intra-thoracic structures and most are due to malignancy. A few cases of superior vena cava syndrome and one case of pulmonary perfusion defects have been previously reported.[1–4] Diagnosis is usually suspected on computed tomography (CT) scan or magnetic resonance imaging (MRI) and confirmed using scintigraphy.[5,6] The treatment of substernal goitre is surgical and is easily extractable through a cervical approach. However in two to four per cent of cases a partial upper sternotomy is required to totally excise the tumour.[5,6] We report a case of pulmonary hypertension and severe cardiac failure secondary to a long-standing substernal goitre.

Case report

A 72-year-old woman presented to the emergency room for severe respiratory distress, orthopnea, ascites and lower limb oedema. Physical examination revealed signs of severe right-sided heart failure with rapid, weak pulse, large disintended jugular veins accentuated during inspiration (positive Kussmaul's sign), hepatomegaly, peripheral oedema, and distant heart sounds. The right lung was clear, and the neurological examination was normal.

Chest roentgenogram showed a huge mass occupying the whole of the left hemithorax with total collapse of the lung. Echocardiography showed severely dilated right cardiac cavities with severe pulmonary hypertension (85/45 mmHg). Within the next two hours her respiratory status deteriorated requiring endotracheal intubation and ventilatory support.

During the past 10 years the patient had started experiencing increasing fatigue, dyspnoea on exertion, and lower limb oedema. A chest radiograph carried out nine years earlier (1988) revealed a mass occupying two thirds of the left hemithorax (Figure 1). A CT scan of the neck and chest performed in 1995 showed a large mediastinal tumour extending into the left pleural cavity (Figure 2). The radiologist then concluded that there was a possible substernal goitre. The patient and her family refused surgery and no further investigations were performed. During the past three months symptoms worsened until her present admission.

A new CT scan showed the same mediastinal tumour which had enlarged and which extended into the left side of the chest with compression of the pulmonary artery and veins. The left lung was almost totally collapsed and displaced towards the diaphragm. During the examination, the patient arrested in the radiology suite. Following successful resuscitation she was transferred to the operating theatre for emergency excision of the mediastinal tumour.

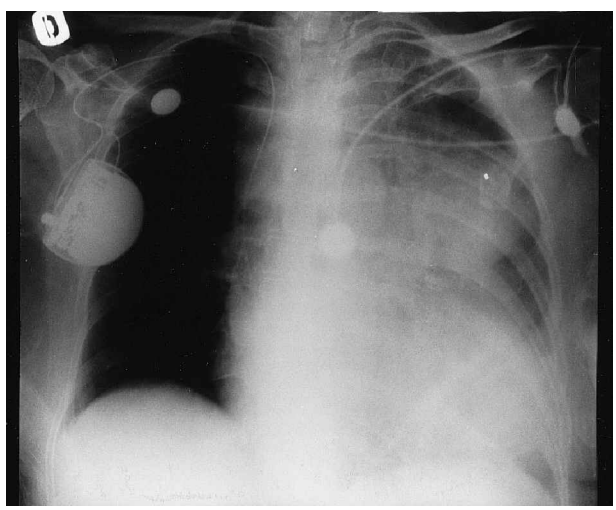

Fig. 1
Chest radiograph carried out nine years earlier (1988) showing an intrathoracic tumour.

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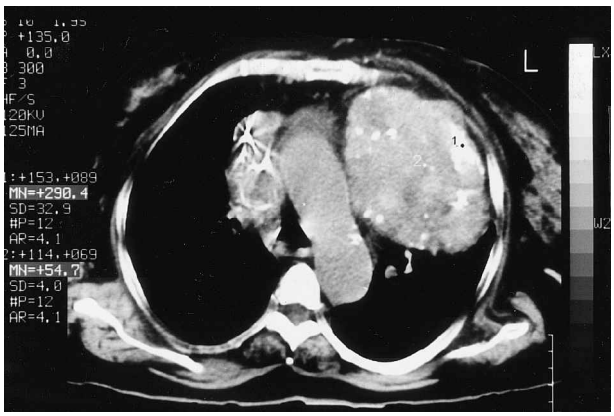

FIG. 2

CT scan of the neck and chest carried out two years earlier showing the thoracic tumour with intra tumoral calcifications.

Through a median sternotomy, exploration of the chest revealed a large solid mass occupying the anterior mediastinum and most of the left pleural cavity. The heart was displaced inferiorly and the right cardiac cavities were severely dilated. The tumour was easily separated from the adjacent structures namely the pericardium, the great vessels, the neck veins, the lung, the left phrenic nerve, and the chest wall. Following excision of the bulky thoracic tumour the left lung was inflated to its normal volume. It was then possible to continue the dissection superiorly towards the neck obviating the thyroid origin of the mass. A frozen section confirmed the diagnosis of benign thyroid multinodular goitre. The operation was terminated by a total thyroid lobectomy on the left side and a subtotal on the right side. Both recurrent laryngeal nerves and the parathyroid glands were preserved. At the end of the procedure pulmonary artery pressure dropped dramatically from 85/40 mmHg pre-operatively to 50/21 mmHg.

Post-operatively the patient required five days of mechanical ventilation and was discharged 10 days later. Pathological examination confirmed the diagnosis of benign multinodular goitre. Control echocardiography at the time of discharge showed a significant decrease in the size of the right cardiac chambers as well as normal pulmonary artery pressure. At one month follow-up she reported no symptoms and echocardiography was satisfactory.

Comment

Thyroid tumours encountered in the thorax are of two types: primary substernal goitres which develop in the mediastinum in relation to aberrant intra-thoraric thyroid tissue and secondary substernal goitres which develop in the neck and extend downwards into the mediastinum. Primary substernal goitres are rare and constitute only one per cent of all substernal goitres.[5,6]

Substernal goitres usually extend into the superior anterior mediastinum and in the great majority of cases are easily excised through a cervical approach without the need for a sternal split. However in two to four per cent of cases a partial upper sternotomy is required to totally excise the tumour.[5,6] It is extremely rare that substernal goitres cause compression of intra-thoracic structures and most are due to malignancy. Few cases of superior vena cava syndrome and one case of pulmonary perfusion defect have been previously reported.[1–4] Diagnosis is usually suspected on CT scan or MRI and confirmed using scintigraphy.[5,6] Unfortunately, in our case cardiac arrest and haemodynamic instability precluded further investigations prior to surgery.

Our patient presented for a substernal goitre with a documented 10-year history of intra-thoracic development and extension. The benign nature of the tumour allowed the slow and progressive development of symptoms related to compression of surrounding vital structures (heart, left lung, left pulmonary artery and veins).

The treatment of substernal goitre is surgical, consisting of either total or subtotal thyroidectomy. Mack[5] provided five reasons in support of surgical management: (1) there is no other treatment for long-standing large multinodular goitres. (2) I^{131}, the alternative to operation for patients with large thyrotoxic goitres, can precipitate acute reactions in the elderly that can result in respiratory distress. (3) A long history of having a large multinodular goitre precluded neither malignancy, hyperfunction, nor complications such as tracheal or oesophageal compression. (4) Malignancy occurs in a significant number of these lesions which are inaccessible to needle biopsy. Up to 16 per cent of patients with substernal goitres have been found to have malignant thyroid disease. (5) Nearly all substernal goitres can be removed through a cervical incision.

In our patient, total excision of the tumour was curative, supporting the absolute indication for surgery in substernal goitres.[5,6]

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