CASE REPORT

Bilateral brachiocephalic vein compression: an unusual and rare presentation of multinodular goitre

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SUMMARY

An interesting and rare case where a longstanding multinodular goitre causes sudden onset symptoms of superior vena cava obstruction. The symptoms were caused by bilateral brachiocephalic vein compression against the first rib. The patient's symptoms included upper limb and facial swelling, whereas her lower limbs were not oedematous (figure 1). The patient underwent CT imaging which showed a multinodular retrosternal thyroid filling the space created by the first rib, sometimes known as the thoracic outlet. The goitre expanded sufficiently to compress the brachiocephalic vein on both sides as shown in figures 2 and 3. This prevented venous drainage from the upper limbs and head. The goitre was also large enough to cause deviation of the trachea although there was no airway compromise. This case is interesting as it is very rare for a benign condition such as a multinodular goitre to cause symptoms of superior vena cava obstruction.

BACKGROUND

Large multinodular thyroid goitre is a reasonably common occurrence but bilateral brachiocephalic vein compression leading to symptoms of superior vena cava (SVC) obstruction is an extremely uncommon presentation.^{1–3}

It is very unusual for a non-malignant condition, such as a multinodular thyroid goitre, to cause symptoms of SVC syndrome (SVCS).^{2–4} It is believed that between 90% and 97% of SVCS cases are due to malignant conditions,^{5–6} with ~70% attributed to advanced lung cancer,^{7–8} 12% to lymphoma and 9% to metastatic cancer.⁴ The most common benign conditions causing SVCS include mediastinal fibrosis, granulomatous mediastinitis, thoracic aortic aneurysms, vasculitis, idiopathic thrombosis and procedural related thrombosis (ie, from central vein catheters, pacemakers).^{2–8–9}

The anatomical location of the thyroid gland means that significant enlargement could result in compression and obstruction of numerous vital structures in the neck and superior mediastinum, including in the trachea, oesophagus, internal jugular veins (IJVs), brachiocephalic veins and the SVC. Locally aggressive thyroid tumours can also cause invasion of the recurrent laryngeal nerve, airway or even the great vessels. In very rare cases, invasive thyroid tumours can also invade the great vessels and lead to carotid blow out. In this case, the enlargement was caused by a multinodular goitre (MNG), a benign and non-invasive cause. MNG rarely causes pathological compression of structures, but in this case, enlargement of the

retrosternal goitre within the fixed space formed by the first rib resulted in compression of the brachiocephalic veins.

The brachiocephalic veins are formed through the union of IJV and subclavian vein. The IJVs are responsible for draining blood from the face and neck and the subclavian veins responsible for blood from the upper limbs. Compression therefore results in the retrograde flow of venous blood and an increase in hydrostatic venous pressure. The venous pressure may be increased from 2–8 to 20–30 mm Hg.¹ This subsequently leads to oedema in the face, neck and arms. The brachiocephalic veins unite to become the SVC, and therefore, symptoms of bilateral brachiocephalic vein compression are almost indistinguishable from SVC obstruction.¹

CASE PRESENTATION

An 82-year-old woman presented to the emergency department with severe respiratory distress. She had a raised urea of 8.1 and a respiratory rate >30 breaths/min. The patient was in type 2 respiratory failure, and routine blood tests showed a slightly raised white cell count ($13.2 \times 10^9/L$) and C reactive protein (25 mg/L). The patient had a background of chronic obstructive pulmonary disease (COPD) requiring home oxygen and a severe exacerbation of COPD was diagnosed which required hospital admission. This exacerbation was treated with non-invasive oxygen and antimicrobial therapy. Two days postadmission, the initial symptoms were greatly improved; however, the patient developed new onset swelling in her face and upper limbs over a matter of hours. On inspection, the patient was conscious and generally well, but there was noticeable facial swelling, facial plethora, neck swelling (with fullness in the supraclavicular fossae bilaterally) and upper limbs swelling bilaterally (figure 1). The oedema was non-pitting and there were distended veins in her neck, thorax and upper limbs. Observations were stable, respiratory and cardiovascular examinations were normal and there was no lymphadenopathy.

The patient had a long-standing history of multinodular thyroid goitre, which was under observation by the Ear, Nose and Throat (ENT) team. It was first diagnosed 15 months prior to this presentation due to an incidental finding of a mediastinal mass on a chest X-ray. An outpatient CT was performed which showed a large multinodular thyroid goitre extending retrosternally into the mediastinum. The right lobe initially measured $4.6 \times 1.9 \times 6.6$ cm and the left lobe measuring 5.2×4.7 cm in the axial plane and at least 9 cm in the longitudinal plane.

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Figure 1 Clinical photograph of patient showing facial, neck and upper limb oedema, with sparing of the lower limbs.

The goitre was causing mild tracheal deviation and compression, but this was not causing any problems clinically. There was no evidence of venous compression at the initial scan. It was noted that the patient's respiratory problems were secondary to moderate-severe COPD. The patient had a repeat CT 1 year later, which reported that there was no significant change in the size of the goitre, and that all major vessels in the neck were patent.

The patient had a medical history of severe COPD with home oxygen, mild heart failure, osteoarthritis and a hiatus hernia. She lived at home with her husband and was independent with no carers, though her husband assisted with some of the activities of daily living. Her maximal exercise tolerance was 50 yards. She is an ex-smoker with a pack-year history of ~55 years.

INVESTIGATIONS

Initial routine blood results on admission showed a raised urea and creatinine, plus a slightly raised white cell count and C reactive protein—likely due to her respiratory infection. Other than this, laboratory findings were normal. The patient was clinically euthyroid and thyroid function tests were within the normal range.

Four days postadmission to the emergency department, the patient underwent a CT scan (figures 2 and 3) to find the cause of her facial and upper limb swelling. CT imaging showed a large multinodular thyroid goitre, which was significantly compressing the brachiocephalic vein lumens bilaterally at the level of the thoracic inlet between the thyroid gland and the first rib. There was no evidence of thrombosis and the SVC and proximal IJVs were patent. The scan also showed the goitre mildly compressing and displacing the tracheal lumen to the right, plus moderate bilateral pleural effusions. Mediastinal and hilar

Figure 2 CT scan with intravenous contrast. Coronal section depicting large multinodular thyroid goitre (T), extending retrosternally and causing significant compression of IJVs bilaterally at the level of the thoracic inlet between the thyroid gland and the first rib (A). There is also compression of subclavian veins at the thoracic inlet by the medial end of the first rib (B). There is also tracheal deviation to the right and moderate bilateral pleural effusions. IJV, internal jugular vein.

Figure 3 CT scan with intravenous contrast. Axial section through the superior mediastinum depicting a large retrosternal multinodular thyroid goitre (T). There is bilateral compression of the IJVs between the first rib and thyroid gland (A). Corresponding subclavian vein dilation is also shown (B). The thyroid (T) is more enlarged on the left causing tracheal deviation to the right. IJV, internal jugular vein.

lymph nodes had also enlarged measuring up to 12 mm in short axis, which was reported as likely reactive changes.

OUTCOME AND FOLLOW-UP

Following her symptoms of SVCS and subsequent CT imaging, she was considered for total thyroidectomy to relieve the compressive symptoms. The ENT and anaesthetic teams reviewed the patient and discussed the risks and benefits of surgery with her. Owing to her severe COPD requiring home oxygen and diagnosis of mild heart failure, it was concluded that the risks of surgery outweighed the benefits. Together they decided she would best be managed medically.

The patient was initially treated with intravenous dexamethasone followed by a course of oral prednisolone. Following some

improvement of her symptoms, the patient was discharged home with a plan for 12-monthly ENT follow-up.

DISCUSSION

Patients with multinodular retrosternal thyroid goitres very rarely present clinically with superior SVCS.^{2–4} The vast majority of SCVS cases are due to malignant conditions, particularly lung cancer.^{5–8} Goitre is more likely to cause the compressive symptoms of SVCS if it extends retrosternally,^{1–3} as seen in this case.

SCVS commonly presents clinically with signs of oedema in the upper extremities, head and neck. Patients may also present with facial plethora, dyspnoea, cyanosis and distention of neck and thoracic veins.^{1 2 5 7 9 10} The patient presented with all of these symptoms and signs, which were more apparent in the morning, after lying in the supine position.

Definitive treatment for a patient presenting with significant compressive symptoms is partial or total thyroidectomy.^{11–15} This would be an effective solution to decompress the patient's brachiocephalic veins, relieve the venous pressure and allow the blood to flow back into the right atrium of the heart. For those patients who undergo thyroidectomy for massive goitre, the possibility for tracheomalacia should be considered, although it is rarely reported.¹⁶

Owing to her significant comorbidities, this patient was not deemed fit to undergo surgery. If the retrosternal component of a goitre is small, such goitres are almost always removed through a cervical approach, the standard approach to thyroidectomy. However, in large retrosternal and mediastinal goitres, a thoracic approach may be required and this may be in the form of a manubriectomy, full sternotomy or thoracotomy.¹⁶

Learning points

- Superior vena cava (SVC) obstruction should be considered in a patient with a known history of thyroid goitre presenting with new onset facial and upper limb swelling.
- The brachiocephalic veins unite to become the SVC and therefore, symptoms of bilateral brachiocephalic vein compression are almost indistinguishable from SVC obstruction.¹
- The treatment of choice for a goitre causing compression symptoms is partial or total thyroidectomy. In those not fit for surgery, medical management with a course of steroids with or without radioactive iodine therapy may be considered.

There is evidence that radioactive iodine therapy is a reasonable, safe and effective therapeutic option, especially in those contraindicated for surgery.^{17–19} It may allow some shrinkage of the goitre and also help prevent further progression. This therefore may pose a possible medical treatment option for this patient in the future.

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Competing interests None declared.

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