

Superior vena caval syndrome due to substernal goitre

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ABSTRACT: Superior vena caval syndrome is usually due to malignant disease, however, benign cases do occur. Two patients with superior vena caval syndrome due to substernal goitre are reported, and the use of phleboscintigraphy and/or vena caval phlebography are advocated. A review of potentially helpful diagnostic procedures is given.

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Obstruction of the superior vena cava leads to a relatively constant complex of symptoms, the most common findings being oedematous swelling and cyanosis of the head, neck and upper extremities, accompanied by dilated collateral veins of the neck, thorax and upper abdomen. The syndrome is more pronounced in acute obstruction, as is the case in malignancy, when venous collaterals have not had time to develop [1]. The syndrome is observed primarily in malignancies, few cases being the result of benign disease [2].

Between January 1, 1980 and November 1, 1987 superior vena caval syndrome (SVCS) was diagnosed in our hospital in 40 cases, 35 of which were the result of malignant disease. In one patient aged 93 yrs, no specific diagnosis was established. Two patients had thrombosis of the superior vena cava. In this paper we report two patients with SVCS due to benign substernal goitre.

Case 1

A 74-yr-old man was admitted to the hospital for evaluation of progressive dyspnoea and hoarseness. He had never experienced pulmonary problems before. In the past, he had smoked cigarettes for a limited number of years (± 10 pack-years). He had noticed a swelling of the neck. Years previously an asymptomatic substernal goitre had been diagnosed incidentally.

On examination he appeared well. Blood pressure was 180/110 mmHg, pulse rate 80 beats per min, regular. A stridor was heard. There was cyanosis of the lips, and the neck veins were markedly distended. Distended venous collaterals were visible on the anterior chest wall. The thyroid gland could not be

palpated and no abnormal cervical lymphadenopathy was found.

Laboratory studies showed no abnormalities apart from an ESR of 58 mm. Results of thyroid function tests were all normal. Arterial blood gas analysis showed an arterial oxygen tension (P_{aO_2}) of 7.8 kPa (normal 8.7-13.1 kPa), oxygen saturation was 93%. A chest X-ray revealed a marked mediastinal widening deviating the trachea to the left.

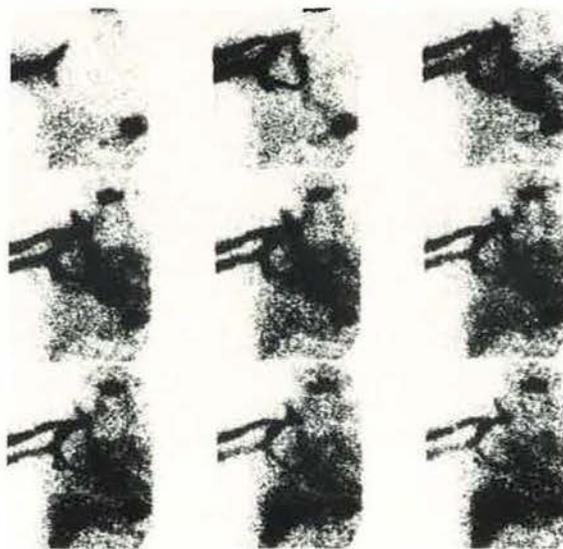


Fig. 1. - Radionuclide superior vena cavogram of case 1 showing obstruction of the superior caval vein and a collateral circulation including the lateral thoracic and intercostal veins. Compare with figure 3 (normal flow).

A radionuclide superior vena cavogram demonstrated superior vena caval obstruction with an extensive collateral circulation including the lateral thoracic and intercostal veins (fig. 1). At bronchoscopy

tracheal narrowing was seen, but no epithelial abnormalities. A computerized tomography (CT) scan of the chest revealed the existence of a large mediastinal mass containing coarse calcifications in continuity with the right thyroid lobe, with narrowing of the trachea. The left thyroid gland was also enlarged but to a lesser extent (fig. 2). A diagnosis of SVCS caused by substernal goitre was established.

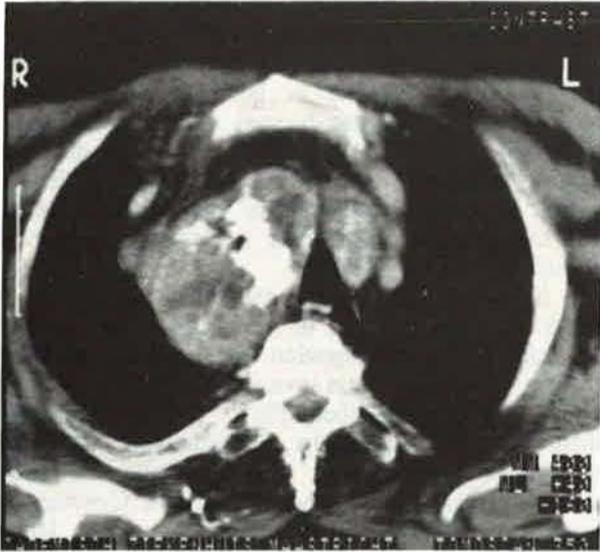


Fig. 2. - CT-scanning of the superior mediastinum of case 1 demonstrating a large well-defined tumour containing gross calcification, deviating and narrowing the trachea.

Subsequently a subtotal right-sided hemithyroidectomy was performed through a Kochers collar incision; the substernal extension of the tumour made it necessary to perform a median sternotomy as well. The resected mass weighed 222 g, the pathological diagnosis was multinodular adenomatous goitre with foci of calcification and recent haemorrhage, without signs of malignancy.

Post-operative recovery was uneventful. A repeated phleboscintigraph one month post-operatively showed a normal blood flow, collaterals were no longer visible (fig. 3).

Case 2

A 73 yr-old-woman was known to have a partially substernal non-toxic goitre for several years, non-insulin dependant diabetes mellitus and chronic bronchitis. In April, 1985 hyperthyroidism was diagnosed. I^{123} thyroid scanning showed an enlarged thyroid gland, with diffuse patchy uptake and substernal extension. She refused surgery or treatment with radio-iodine, so thyroid suppressive drugs were prescribed. In September, 1987 she was admitted to the hospital complaining of fatigue, dysphagia, dyspnoea and swelling of the face.

On physical examination she was somnolent. An inspiratory stridor was heard. A large goitre was palpated without bruits and no abnormal cervical lymphadenopathy was found. The neck veins were markedly distended bilaterally, and a superficial venous pattern was present over the upper anterior chest wall.

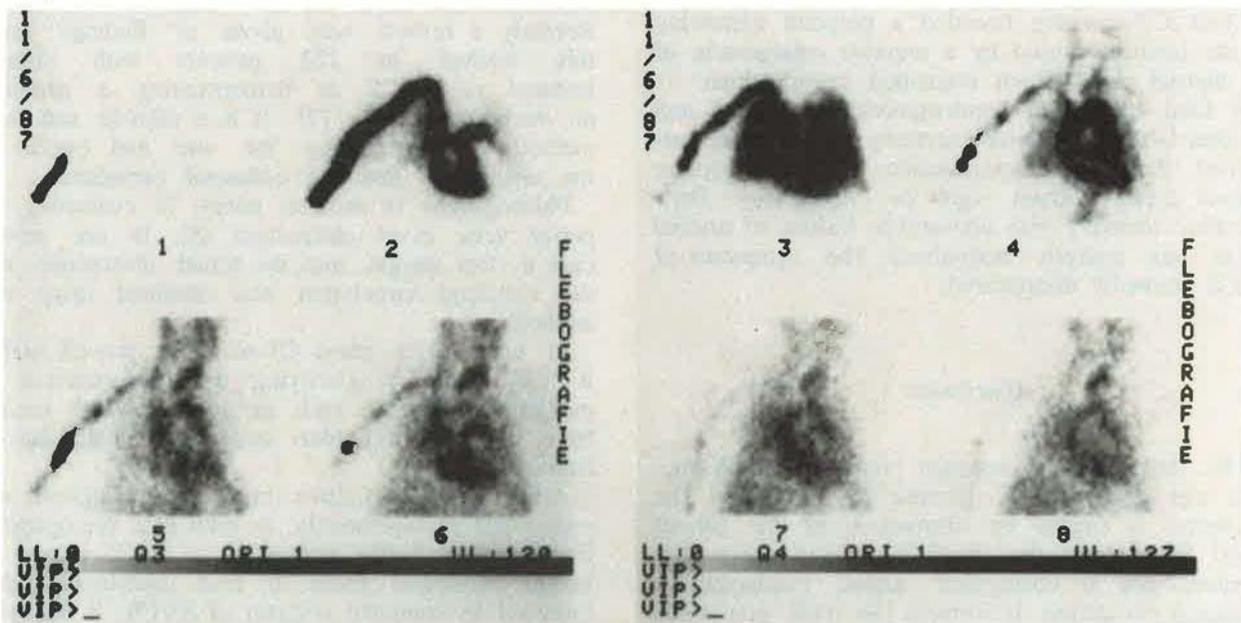


Fig. 3. - Radionuclide superior vena cavogram of case 1 after resection of the substernal goitre. There is a normal flow of the injected activity through the subclavian and superior caval vein, the right ventricle (2), the lungs (3) and the left ventricle and the aorta (4).

Laboratory investigations, including thyroid function tests were normal. Arterial blood gas analysis demonstrated a pH of 7.37, arterial carbon dioxide tension (P_{aCO_2}) of 8.4 kPa, P_{aO_2} of 5.1 kPa, (normal: pH 7.45, P_{aCO_2} 4.5–5.9 kPa, P_{aO_2} 8.7–13.1 kPa).

The chest X-ray showed a large anterior mediastinal mass. A phlebogram, produced by simultaneous injection of contrast in the arm veins bilaterally, gave evidence of complete obstruction of the superior vena cava and extensive collateral venous circulation via the lateral thoracic and intercostal veins (fig. 4).



Fig. 4. — Contrast phlebogram of case 2 after simultaneous injection of contrast medium in both arms, showing obstruction of the blood flow in both subclavian veins and an extensive collateral circulation.

Chest CT-scanning revealed a pinpoint narrowing of the trachea, caused by a massive enlargement of the thyroid gland which contained calcifications.

A total right-sided hemistrulectomy (230 g) and subtotal left-sided hemistrulectomy (210 g) were performed. Pathological examination showed a nodular colloid goitre without signs of malignancy. Post-operative recovery was uneventful. Values of arterial blood gas analysis normalized. The symptoms of SVCS gradually disappeared.

Discussion

The first case of superior vena caval obstruction was described by HUNTER in 1757 [3]. The syndrome is caused by obstruction of the venous blood flow from the head, neck and upper extremities and if obstruction arises insiduously a collateral circulation is formed. The most commonly encountered collaterals are the lateral thoracic, intercostal and internal mammary veins, and the azygos vein, which is the principal collateral pathway.

During recent decades there has been a remarkable change in the incidence of the most common causes of SVCS. In a review of the literature in 1949 [4], the most common causes were primary thoracic malignancy (33%), aortic aneurysm (30%) and chronic mediastinitis (19%). In a review of 274 cases in 1954 [5], luetic aortic aneurysm or tuberculous mediastinitis was reported in 40%. Over the years a gradual decrease of SVCS caused by benign diseases has occurred. In a recent survey [6] a malignant disease is listed in up to 97% of the cases. Usually this implies a pulmonary malignancy, malignant lymphoma or metastatic disease of a tumour localized elsewhere.

SVCS as a result of benign conditions such as tuberculosis or syphilis is becoming increasingly rare. A marked increase is seen in iatrogenic causes of SVCS. Recently, the increase in use of intravenous techniques such as Swann-Ganz catheters, cardiac pacemaker electrodes or parenteral alimentation catheters has resulted in a growing number of reports of benign SVCS.

It has been stated that immediate treatment of SVCS is warranted, but in most cases there is time for additional investigation to establish a more specific diagnosis: non-invasive methods such as X-ray, contrast phlebography, phleboscintigraphy, duplex doppler sonography, CT-scanning and magnetic resonance imaging can determine the exact size and localization of the obstruction. Cytology of sputum, bronchial washings, pleural effusions, lymph node or bronchial biopsy, and thoracotomy can ascertain a pathological diagnosis.

In our first case phleboscintigraphy, or radionuclide venography of the superior vena cava, was helpful in demonstrating a total obstruction of the superior vena cava and the collateral circulation. Recently a review was given of findings using this method in 123 patients with clinical features of SVCS or demonstrating a parahilar or mediastinal mass [7]. It is a reliable and safe method of determining the site and extent of the obstruction and the collateral circulation.

Phlebography is another means of evaluating superior vena caval obstruction [8]. In our second case a clear insight into the actual obstruction and the collateral circulation was obtained using this method.

In both cases chest CT-scanning proved useful. It is of value in clarifying the characteristics of mediastinal tumours, such as goitres, which usually have well-defined borders and often contain calcification [9].

Although no definitive histological diagnosis was established pre-operatively, in both cases pre-operative findings were highly suggestive of SVCS caused by benign substernal goitre. In both patients thyroidectomy led to complete reversal of SVCS. It has been reported that in case of a toxic multinodular goitre, I^{131} therapy can reduce tumour mass [10]. Our second patient refused this therapy in 1985. In 1987

because of the life-threatening narrowing of the trachea emergency surgery was warranted.

SVCS is in most cases the result of malignant disease, but benign cases do occur. With the range of modern diagnostic procedures a specific pretreatment diagnosis can be established.

References

1. Simpson JR, Perez CA, Presant CA, van Amburg AL. – Superior vena caval syndrome. In: Oncologic emergencies. J.W. Yarbo and R.S. Bornstein eds, Grune & Swatton Inc., New York, 1981, pp. 43–72.
2. Lochridge SK, Knibbe W, Doty DB. – Obstruction of the superior vena cava. *Surgery*, 1979, 85, 14–24.
3. Hunter W. – The history of an aneurysm of the aorta with some remarks on aneurysm in general. *Med Observ Ing*, 1757, 1, 323–357.
4. McIntire FT, Sykes EM Jr. – Obstruction of the superior vena cava: a review of the literature and report of two personal cases. *Ann Intern Med*, 1949, 30, 925–960.
5. Schechter MM. – The superior vena cava syndrome. *Am J Med Sci*, 1954, 227, 46–56.
6. Schraufnagel DE, Hill R, Leech JA, Pare JAP. – Superior vena caval obstruction. Is it a medical emergency? *Am J Med*, 1981, 70, 1169–1174.
7. Sy WM, Lao RS, Seo JS. – Scintigraphic features of superior vena caval obstruction due to substernal non-toxic goitre. *Br J Radiol*, 1982, 55, 301–303.
8. Dyet JF, Moghissi K. – Role of venography in assessing patients with superior vena caval obstruction caused by bronchial carcinoma for bypass operations. *Thorax*, 1980, 35, 628–630.
9. Shepard J-AO. – Computed tomography of the mediastinum. *Clinics in chest medicine*, 1984, 5, 291–305.
10. Siderys H, Rowe GA. – Superior vena cava syndrome caused by intrathoracic goiter. *Am Surg*, 1970, 36, 446–450.

RÉSUMÉ: Le syndrome de la veine cave supérieure est généralement considéré comme résultant d'affections malignes; quoique des cas bénins puissent survenir. Deux patients atteints d'un syndrome de la veine cave supérieure dû à un goitre rétro-sternal font l'objet de cette présentation. On plaide pour l'utilisation de la phlébo-scintigraphie et/ou de la phlébographie veineuse cave. Présentation d'une revue de la littérature sur le syndrome de la veine cave supérieure et résumé des méthodes de diagnostic potentiellement utiles.