A case of Riedel’s thyroiditis

Riedel’s thyroiditis is a rare disorder that mainly affects middle-aged women. Dense fibrosis disrupts the normal thyroid architecture and replaces the normal thyroid architecture, and can extend beyond the thyroid capsule.

This is the first report of a case of Riedel’s thyroiditis associated with systemic, multifocal fibrosis in a black South African patient.

Case report

A 53-year-old black woman was admitted to Chris Hani Baragwanath Hospital, Johannesburg, with ascites. She had been diagnosed as hypothyroid the previous year, the cause thought to be Hashimoto’s thyroiditis. She had been treated with thyroxine, but at the time of admission was clinically and biochemically hypothyroid (free T4 3.3 pmol/l, TSH >150 mIU/l). Examination revealed a small, firm goitre which moved well on swallowing. She was mildly proptotic. An ascitic tap was performed and the fluid found to be chylous with a triglyceride concentration of 3.17 mmol/l. Urea and electrolyte testing on admission revealed that she was in renal failure (urea 32.4 mmol/l, creatinine 1 676 µmol/l). Serum calcium and phosphate values were within normal limits.

An abdominal ultrasound scan showed her to have bilateral hydronephrosis and a multifibroid uterus. Gynaecological opinion was that the enlarged uterus was unlikely to account for the hydronephrosis. An attempted nephrostomy on the right side failed, but following her second dialysis session, she developed a right-sided hemiplegia and expressive aphasia. A non-contrast computed tomography (CT) scan of the brain showed a watershed infarct with cerebral oedema. Her level of consciousness deteriorated further and she died 22 days after admission.

A postmortem examination, performed with her family’s permission, revealed the following:

1. A destroyed thyroid gland that had been replaced by fibrous tissue, with lymphocyte and plasma cell infiltration, in keeping with Riedel’s thyroiditis (Fig. 1, A, B and C).
2. Extensive perivascular fibrosis of the lungs.
3. Mediastinal fibrosis.
4. Extensive retroperitoneal fibrosis, with fibrosis surrounding the ureters bilaterally and encasing the left kidney (Fig. 1, D).
5. A right perinephric abscess.

Discussion

Fibrous or Riedel’s thyroiditis was originally described in 1896 as a ‘specific inflammation of mysterious nature producing an iron hard tumefaction of the thyroid’. It is an extremely rare condition, making up only 0.03 - 0.98% of surgical thyroid diseases. It is characterised by a chronic inflammatory process which often extends beyond the thyroid gland to the surrounding tissues, resulting in hypoparathyroidism, hoarseness due to recurrent laryngeal nerve involvement, and tracheal compression with stridor. The fibrosis may extend down into the mediastinum and also involve the anterior chest wall.¹

The fibrotic process is, in fact, part of a systemic, multifocal fibrosclerotic disorder that may affect many distant sites including the retroperitoneal space, the retro-orbital area and the biliary tree.² Our patient

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had thyroid, retroperitoneal, mediastinal and lung involvement, with the retroperitoneal fibrosis being the likely cause of her chylous ascites.

Fibrous thyroiditis characteristically presents as a non-tender, hard anterior neck mass which is adherent to the adjacent soft tissues and consequently may fail to move on swallowing. Regional lymph nodes are sometimes enlarged so that the clinical picture may suggest a carcinoma. Depending on the extent of the perithyroidal soft-tissue involvement the patient may have stridor, hoarseness, dysphagia or hypoparathyroidism. Despite the extensive fibrosis, hypothyroidism occurs in only 30 - 40% of patients.

Radio-iodine thyroid scanning shows either low or heterogeneous uptake. Computed tomography and ultrasound scanning may be useful to delineate the extent of the fibrosis.

Fine-needle aspiration may be performed to exclude carcinoma, if suspected. However, in cases of fibrous thyroiditis, the cell yield is often very low. In such circumstances an open biopsy may therefore be necessary to make a definitive diagnosis. Macroscopically, the thyroid tissue is hard, pale and avascular. Histology shows dense hyalinised fibrous tissue with a sparse infiltrate of lymphocytes, plasma cells and eosinophils. Once the diagnosis has been established, the patient should be evaluated to exclude other sites that may be involved by the multifocal fibrosclerotic process. Fibrosis of the thyroid gland may also be seen in Hashimoto’s thyroiditis, but to a far lesser extent, and does not extend to surrounding tissues.

The aetiology is uncertain, but the chronic inflammatory infiltrate and vasculitis evident in the fibrous tissue, as well as the presence of antithyroid antibodies in the sera of two-thirds of these patients, suggest an underlying autoimmune mechanism. Alternatively, however, the multifocal nature of the disease is also compatible with a disorder of the fibroblast. No link between drugs and Riedel’s thyroiditis has been shown.

Treatment is directed at both the hypothyroidism and the fibrosclerotic manifestations. Limited surgery is performed to relieve any tracheal or oesophageal obstruction, if present, but more invasive surgery should not be performed as the tissue planes are often obscured by fibrosis, with a consequent high risk of trauma to neck structures. Therapy with corticosteroids and tamoxifen has been reported to be helpful in a small number of patients. Owing to the rarity of this condition, no clinical trials have been performed on the efficacy of these drugs. Without therapy, however, the condition is usually progressive, albeit slowly, and empiric medical therapy is therefore probably justified.

Fig. 1. A: Postmortem specimen of the patient’s thyroid gland showing complete replacement of the gland by avascular fibrous tissue. B: Haematoxylin and eosin (H&E) section through a normal thyroid gland. Note the follicles filled with colloid. C: H&E section through the patient’s thyroid gland. The normal architecture has been completely obliterated by dense hyaline fibrous tissue with a lymphocyte and plasma cell infiltrate. D: Postmortem specimen of the patient’s right kidney and surrounding tissue demonstrating the abundant retro-peritoneal fibrosis encasing the kidney.
Conclusion

This is the first reported case of a black patient in sub-Saharan Africa presenting with Riedel’s thyroiditis accompanied by extensive fibrosis.

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A pituitary macroadenoma presenting with hyponatraemia

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Clinical examination revealed a well-looking patient who was not confused, was apprehensive, and had normal hydration, a blood pressure of 167/92 mmHg with no postural drop, a pulse rate of 54 beats/min and a respiratory rate of 18 breaths/min. Apart from mild tenderness in the left lower abdomen, systemic examination was unremarkable. Since the patient was an elderly man with a change in bowel habits and loss of weight, the concern at this stage was that he might have a bowel malignancy and he was admitted to the emergency medical admission ward.

The findings on baseline biochemical investigation were as follows: sodium 108 mmol/l (normal 135 - 141 mmol/l), potassium 4.5 mmol/l (3.3 - 5.3 mmol/l), urea 1.5 mmol/l (2.6 - 7.0 mmol/l), creatinine 46 µmol/l (60 - 120 µmol/l), urinary sodium on a random urine sample 149 mmol/l, haemoglobin 10.2 g/dl (13.0 - 17.0 g/dl), mean cell volume 103.8 fl (79.1 - 98.9 fl), white cell count 4.42×10⁹/l (4.00 - 10.00×10⁹/l), platelets 191×10⁹/l (137 - 373×10⁹/l). A chest radiograph was normal. A working diagnosis of syndrome of inappropriate antidiuretic hormone secretion (SIADH) was made and the patient was placed on fluid restriction. Since his chest radiograph was normal a magnetic resonance image (MRI) scan of the brain was done and showed a well-defined pituitary lesion measuring 18 mm by 18 mm, compatible with a pituitary macroadenoma (Fig. 1). The patient was then referred to the endocrine