

Riedel's Thyroiditis in a Patient with Recurrent Subacute Thyroiditis: A Case Report and Review of the Literature

MIN HO CHO, CHUL SIK KIM, JONG SUK PARK, EUN SEOK KANG, CHUL WOO AHN, BONG SOO CHA, SUNG KIL LIM, KYUNG RAE KIM AND HYUN CHUL LEE

Division of Endocrinology and Metabolism, Department of Internal Medicine, Yonsei University College of Medicine, Seoul, Korea

Abstract. Riedel's thyroiditis is a rare form of chronic thyroiditis, characterised by a fibroinflammatory process that partially destroys the thyroid and often involves surrounding tissues. The relationship of Riedel's thyroiditis to other forms of thyroiditis is not clear. A case of Riedel's thyroiditis in a 51-year-old woman presenting with symptoms of subacute thyroiditis, is reported. She was diagnosed with subacute thyroiditis based on clinical manifestation and laboratory results. She was treated with glucocorticoids for six weeks, and then followed-up for 12 months. Three years later, she visited with tenderness and enlargement of thyroid mass, and laboratory and radiology findings suggested that she had a malignant thyroid tumor as well as subacute thyroiditis. After thyroidectomy, histopathologic findings showed that she had Riedel's thyroiditis in the presence of subacute thyroiditis. Until now, few cases of Riedel's thyroiditis in patients with a history of subacute thyroiditis have been reported in the literature. Although the etiology of Riedel's thyroiditis is unknown, it may develop in the course of subacute thyroiditis.

Key words: Riedel's thyroiditis, Invasive fibrosing thyroiditis, Subacute thyroiditis, Thyrotoxicosis

(Endocrine Journal 54: 559–562, 2007)

RIEDEL'S thyroiditis is a rare and chronic inflammatory disease of the thyroid gland, and was first documented by Bernhard Riedel in 1896 [1]. The relationship between subacute thyroiditis and Riedel's thyroiditis is unclear; there have been no reports of recurrent subacute thyroiditis concomitant with Riedel's thyroiditis in the literature. A patient with a history of subacute thyroiditis presented in our clinic with enlargement of the left thyroid nodule and pain on her anterior neck. She was diagnosed with Riedel's thyroiditis after investigative surgery to rule out thyroid malignancy.

Case Report

A 51-year-old woman presented in our clinic with

sudden onset of painful swelling in anterior neck. She had been treated for upper respiratory infection at another clinic 2 weeks prior to this visit. Her height was 156 cm, weight 61 kg, blood pressure 140/90 mmHg, pulse rate 106 per minute, respiration rate 18 per minute, and body temperature 37.1°C. Her thyroid was diffusely enlarged, quite firm, and tender. A fairly fixed nodule (2.0 × 1.0 cm) was palpated in the left thyroid gland. Abnormal laboratory findings included elevated erythrocyte sedimentation rate (ESR) of 61 mm/hr, and elevated serum T₃ of 209 ng/dL (normal, 84–157 ng/dL), elevated serum T₄ of 13.0 µg/dL (normal, 4.7–9.3 µg/dL), and a decreased TSH level of 0.1 µIU/mL (normal, 0.39–3.68 µIU/mL). Both anti-thyroperoxidase (anti-TPO) antibody and anti-thyroglobulin (anti-Tg) antibody were negative and anti-TSH receptor antibody was also negative. Thyroid radioiodine uptake ratio was 3.42% after 2 hours and 3.10% after 24 hours. Fine needle aspiration cytology (FNAC) of the suspect thyroid nodule was negative, however, there was infiltration of lymphocytes and leukocytes, and several giant cells were observed.

Received: October 25, 2006

Accepted: March 2, 2007

Correspondence to: Chul Woo AHN, M.D., Yongdong Severance Hospital, Yonsei University, College of Medicine, 146-92 Dogok-Dong, Kangnam-Ku, Seoul, Korea



Fig. 1. Technetium-99m pertechnetate thyroid scintigraphy after 2 hour ingestion showed generalized decreased uptake of the radiotracer by both lobes of the thyroid gland.

With these clinical and laboratory findings, we made the diagnosis of subacute thyroiditis and prescribed glucocorticoids. Six weeks later, the pain disappeared and her ESR returned to normal. Follow-up FNAC at three and six months showed colloid and follicular cell proliferation, but no malignant cells.

We recommended periodic follow-up visits, however, she did not return until three years later, when she presented with enlargement of the left thyroid nodule and pain in her anterior neck. There was a diffuse enlargement in the thyroid, accompanied by tenderness, and a firm, fixed nodule was palpated in the left thyroid. Abnormal laboratory findings included an elevated ESR of 54 mm/hr, elevated serum T_3 of 182.4 ng/dL (normal, 71–161 ng/dL), elevated free T_4 of 2.0 ng/dL (normal, 0.8–1.7 ng/dL), and a decreased TSH of 0.01 μ IU/mL (normal, 0.41–4.43 μ IU/mL). Both anti-TPO antibody and anti-Tg antibody were negative. Technetium-99m pertechnetate thyroid scintigraphy showed generalized decreased radio-uptake by both lobes of the thyroid gland (Fig. 1). Thyroid radioiodine uptake ratio was 2.12% at 2 hours and 0.66% at 24 hours after iodine ingestion. Ultrasonogram showed a 2.5 cm-sized infiltrating hypoechoic mass with internal calcification and daughter nodule at the left thyroid (Fig. 2). We performed a subtotal thyroidectomy due to suspicion of malignancy. During the operation, it was noted that the thyroid gland was bilaterally enlarged and very hard, and had to be separated from the adjacent muscles and trachea with great difficulty. We

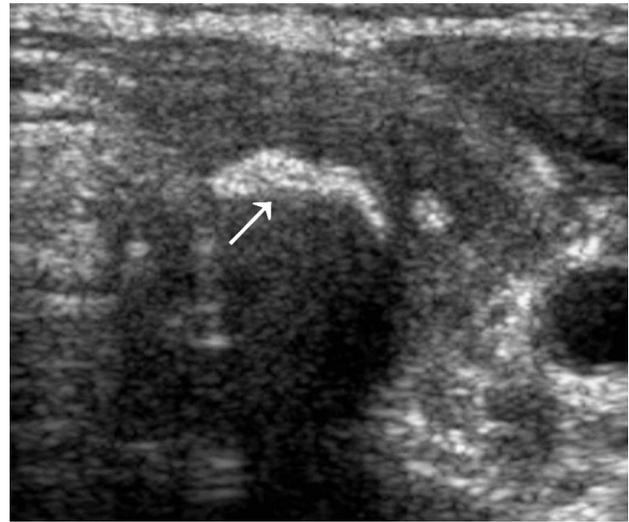


Fig. 2. Thyroid ultrasonography shows an approximately 2.5 cm-sized infiltrating lesion with internal calcification (white arrow) and daughter nodule at the left thyroid gland.

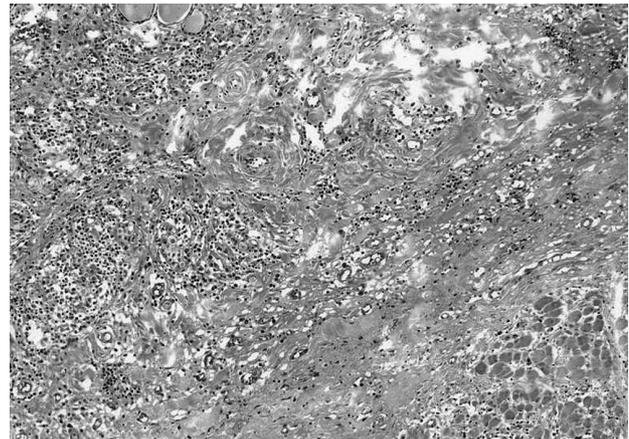


Fig. 3. In the histopathologic examination, diffuse fibrosis and inflammatory cell infiltration were observed (H&E stain \times 400).

also noticed severe inflammation and adhesion around the thyroid. Histopathologic examination revealed diffuse fibrosis and the infiltration of many inflammatory cells; however, no malignant cells were observed (Fig. 3). We made the diagnosis of Riedel's thyroiditis, and the patient made a full recovery.

Discussion

Riedel's thyroiditis is a rare form of thyroiditis. It is

a chronic inflammatory disease of unknown cause. In 1883, Bernhard Riedel observed patients who had chronic fibrosclerosis and inflammation of the thyroid, which resulted in destruction of the thyroid. The inflammation involved the peripheral neck tissues such as the esophagus or bronchus. Two such cases were reported by Riedel in 1896 [1]. In 1922, Ewing suggested that Riedel's thyroiditis was not an independent disease but an advanced form of Hashimoto's thyroiditis. This hypothesis was then accepted as the established theory [2]. After Graham [3] and Joll [4], however, reported that Riedel's thyroiditis was different from Hashimoto's thyroiditis or subacute thyroiditis in clinical and histopathologic patterns, Riedel's thyroiditis was considered an independent disease, but there was still controversy as to its cause. In 1957, Woolner *et al.* [5] reported that they found 20 cases of Riedel's thyroiditis from the results of re-examination of 42,000 thyroid resection specimens collected for 36 years (0.05% out of those who underwent surgery due to thyroid disease), and that Riedel's thyroiditis was characterized by the spread of fibrosis outside of the thyroid capsule and the total destruction of normal thyroid structure. Since then, Riedel's thyroiditis is considered a part of multifocal fibrosclerosis, and Bartholomew *et al.* [6] in 1963 and Rao *et al.* [7] in 1973 reported that the thyroiditis was accompanied with sclerosing cholangitis, retroorbital pseudotumor, mediastinal fibrosis, and retroperitoneal fibrosis. Drury *et al.* [8] in 1974 reported that Riedel's thyroiditis and pernicious anemia were observed in a female patient and that the findings indicated the relation between the thyroiditis and autoimmunity. In 1985, Hay [9] diagnosed 37 cases of Riedel's thyroiditis based on 56,700 thyroid resections performed between 1920 and 1984. He observed fibrosclerosis in the non-cervical areas such as the orbit, mediastinum, and retroperitoneum of 12 patients with thyroiditis after a 10-year follow-up. His research indicated that the mean age of the patients was 51 and that the disease was more frequent in women than in men (3.1:1).

Patients with Riedel's thyroiditis present with a sudden inflammation of the goiter, dyspnea or dysphagia due to a compressed bronchus or esophagus, and a firm mass in the anterior neck that does not move when palpated. The disease is bilateral in most cases, and the function of the thyroid can be normal (60% of cases) or decreased (34% of cases) but thyrotoxicosis is rare. A CT scan will show that the lesion infiltrates the thyroid,

peripheral organs, and vessels in the neck; the ESR will be increased. Thyroid autoantibody is positive in approximately 45–67% of cases, but the titer is lower when compared to that of Hashimoto's thyroiditis. Fibrosis characteristically invades and encloses the adjacent structures in the neck extending beyond the limits of thyroid capsules. It is important to distinguish between Riedel's thyroiditis and thyroid carcinoma; however, it is clinically impossible to make an absolute diagnosis [10, 11]. Therefore, surgical biopsy and treatment is commonly used.

Further, it is important to distinguish fibrous Hashimoto's thyroiditis and subacute thyroiditis from Riedel's thyroiditis. Histopathologically, Riedel's thyroiditis is considerably different from subacute thyroiditis, but may be similar to fibrous Hashimoto's thyroiditis [12]. Riedel's thyroiditis was once considered an advanced form of Hashimoto's thyroiditis [13], but recently has been classified as an independent disease [14]. Subacute thyroiditis is much more common than Riedel's thyroiditis, and the two diseases are classified separately [15]. Subacute thyroiditis is characterized by the diffuse enlargement and febricity of the thyroid that is solidified and accompanied with pain, an increase in ESR, and a decline in radioactive iodine uptake. These features are not observed in patients with Riedel's thyroiditis in general. Infiltration into peripheral tissues is common in Riedel's thyroiditis but is not typical in subacute thyroiditis. Fibrosis in the thyroid can be also seen in subacute thyroiditis, chronic thyroiditis, radiation-induced thyroiditis, nodular thyroiditis, pachydermatosis, trauma, and amyloidosis, but such diseases are evidently discriminated from Riedel's thyroiditis in clinical patterns [16].

There are only two previous cases reported in the literature of patients with Riedel's thyroiditis who also presented with a history of subacute thyroiditis [15, 17]. Although there are some differences, most of the features are similar to ours including the results of laboratory and radiologic examinations. The patients were all women and almost all the same age (47, 49, and 51 years old) and had no autoantibodies associated with the thyroid gland; however, an important difference is that our patient presented with recurrent subacute thyroiditis. However, the distinction between the two diseases may not always be as clear as once thought. Several earlier reports on Riedel's thyroiditis included many patients with subacute thyroiditis [5], indicating that competent observers could not always

make the distinction [15]. It seems reasonable to conclude that Riedel's thyroiditis may be a rare stage in subacute thyroiditis. We believe our patient represents such a case because she had both clinical subacute thyroiditis and histopathological Riedel's thyroiditis; however, it is also possible that the subacute thyroiditis was superimposed on an otherwise unrelated Riedel's thyroiditis, a coincidence that appears to be quite rare as well.

In most cases, Riedel's thyroiditis has a relatively benign course and mortality is extremely rare and is mainly caused by recurrent pneumonia due to bronchial compression and dyspnea. As for treatment, high-dose corticosteroid is advisable as the initial treatment method. This treatment brings about a rapid improvement in symptoms and softens the neck mass, but some patients may need long-term steroid therapy because the disease will recur when the dose is decreased. Surgery is appropriate when tissue samples are needed for diagnosis, drug treatment shows no effect, or compres-

sion symptoms are very severe. During surgery, wedge resection is conducted on the isthmus to separate the right from the left lobe in order to relieve compression and tissues can be obtained for diagnosis. Total or subtotal thyroidectomy is not appropriate because of the severe adhesion to peripheral tissues. Low-dose radiation therapy, effective in some cases, can be considered when other therapies are useless. Few *et al.* reported that using tamoxifen was effective in patients with Riedel's thyroiditis who showed no response to other existing treatments including glucocorticoids [18].

In the case reported here, the patient underwent surgery to rule out malignancy, and the left lobe of the thyroid was ablated because the features of thyroiditis were observed in the frozen section examined intraoperatively. After the surgery, the patient showed a favorable outcome without using glucocorticoids and no evidence of recurrence has been found. Further follow-up will be necessary to determine whether other clinical patterns of multifocal fibrosclerosis occur.

References

- Riedel BMCL (1896) Die chronische, zur Bildung eisenharter Tumoren fuhrende Entzündung der Schildduese. *Verh Dtsch Ges Chir* 25: 101–105.
- Ewing J (1922) Neoplastic Disease, 2nd ed. Saunders, Philadelphia.
- Graham A (1931) Riedel's struma in contrast to struma lymphomatosa (Hashimoto). *West J Surg* 39: 681–689.
- Joll CA (1939) The pathology, diagnosis and treatment of Hashimoto's disease (struma lymphomatosa). *Br J Surg* 27: 351–390.
- Woolner LB, McConahey WM, Beahrs OH (1957) Invasive fibrous thyroiditis (Riedel's struma). *J Clin Endocrinol Metab* 17: 201–220.
- Bartholomew LG, Cain JC, Woolner LB (1963) Sclerosing cholangitis. Its possible association with Riedel's struma and fibrous retroperitonitis. *New Engl J Med* 269: 8–12.
- Rao CR, Ferguson GC, Kyle VN (1973) Retroperitoneal fibrosis associated with Riedel's struma. *Can Med Assoc J* 108: 1019–1021.
- Drury MI, Sweeney EC, Heffernan SJ (1974) Invasive fibrous (Riedel's) thyroiditis. *Ir Med J* 61: 388–390.
- Hay ID (1985) Thyroiditis; A clinical update. *Mayo Clin Proc* 60: 836–843.
- Marin F, Araujo R, Paramo C, Lucas T, Salto L (1989) Riedel's thyroiditis associated with hypothyroidism and hypoparathyroidism. *Postgrad Med J* 65: 381–338.
- Marchesi M, Biffoni M, Mastropietro T, Millarelli M, Marzullo A, Campana FP (1997) Riedel's thyroiditis: a case report and review of the literature. *G Chir* 18: 820–822.
- Barsano CP (1996) Other forms of primary hypothyroidism. In: Braverman LE, Utiger RD (ed) Werner and Ingbar's The Thyroid, ed 7. Lippincott-Raven Publishers, Philadelphia, p. 768.
- Zelmanovitz F, Zelmanovitz T, Beck M, Cerski CT, Schmid H, Czepielewski MA (1994) Riedel's thyroiditis associated with high titers of antimicrosomal and antithyroglobulin antibodies and hypothyroidism. *Endocrinol Invest* 17: 733–737.
- LiVolsi VA (1996) Pathology. In: Braverman LE, Utiger RD (ed) Werner and Ingbar's The Thyroid, ed 7. Lippincott-Raven Publisher, Philadelphia, p. 501–502.
- Chopra D, Wool M, Crosson A, Sawin CT (1978) Riedel's struma associated with subacute thyroiditis, hypothyroidism and hypoparathyroidism. *J Clin Endocrinol Metab* 46: 869–871.
- LiVolsi VA (1990) Fibrosis in the thyroid. In: Li Volsi VA (ed) Surgical Pathology of the Thyroid. W.B. Saunders Company, Philadelphia, p. 98.
- Kabalak T, Ozgen AG, Günel O and Yetkin E (2000) Occurrence of Riedel's thyroiditis in the course of subacute thyroiditis. *J Endocrinol Invest* 23: 399–401.
- Few J, Thompson NW, Angelos P, Simeone D, Giordano T, Reeve T (1996) Riedel's thyroiditis: treatment with tamoxifen. *Surgery* 120: 993–999.