

COMMUNICATIONS

ORBITAL INVOLVEMENT IN RIEDEL'S THYROIDITIS*

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RIEDEL'S thyroiditis (Riedel, 1896; 1897) is a rare condition. Many conditions previously described as invasive fibrous thyroiditis were probably in fact other forms of thyroiditis, which include struma lymphomatosa (Hashimoto's woody thyroiditis) and granulomatous thyroiditis (de Quervain's disease) (Woolner, McConahey, and Beahrs, 1957; 1959).

Hashimoto's thyroiditis may be either diffuse or focal. Usually occurring in elderly females of average age 55 years, it presents with a lump in the neck, of variable duration, and sometimes a previous history of goitre. The swelling of the thyroid gland is firm and, apart from causing a sense of fullness, is usually asymptomatic. Clinical findings include a low basal metabolic rate with actual myxoedema in 25 per cent. of cases. The paradoxical association of low normal or subnormal levels of serum protein-bound iodine with a normal or even increased thyroïdal uptake of ^{131}I is a diagnostic feature. Titres of antithyroid antibodies are high. The histological picture of the thyroid varies; there may be diffuse epithelial destruction, oxyphilic epithelium, varied epithelial changes, or lymphoid infiltration. Characteristically there is a diffuse interfollicular infiltration with plasma cells and lymphocytes and a variable amount of fibrosis. The lobular pattern of the gland remains; although the capsule of the gland may be involved and adhesions form to the surrounding tissues, invasion never occurs. Medical treatment with desiccated thyroid is commonly employed; failure to respond to such treatment or the presence of a suspicious nodule or nodules provides adequate reason for surgical exploration.

Granulomatous thyroiditis is three times more common in females than in males and the average age incidence is 50 years. While 10 per cent. of affected patients present with an asymptomatic lump in the neck, the rest have moderate to severe symptoms. There may be tenderness or pain of the thyroid gland, the pain often being referred to the ears or jaws; fever, malaise, and symptoms of recent pharyngitis or upper respiratory infection may be present. The gland is usually two or three times its normal size; the affected area is firm and resilient and one or both sides may be involved. The histological findings vary according to the stage of the disease; zones of activity frequently alternate with zones of healing and fibrosis. The active granulomatous area consists of a central mass of colloid with a margin of giant cells and mononuclear cells, which often contain ingested colloid particles. Aggregations of polymorphonuclears may be present. A somewhat later stage in the process is marked by a pronounced tuberculoid response with almost complete disappearance of colloid and somewhat more extensive inflammatory fibrosis. As the inflammatory

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process subsides still more, there remains a fibrous stroma. Although occasionally the condition follows a prolonged course, it is usually self-limiting, the patient becoming asymptomatic and the gland impalpable in the course of a few months.

Riedel's thyroiditis affects patients in a younger age group than those with Hashimoto's disease, and women preponderate. The patient presents with a goitre which may have been present for only a few months or for several years. The disease starts in one part of a lobe of the thyroid gland; it may spread to involve the whole of that lobe, and may later involve the other side. The thyroid enlargement is of extreme hardness and becomes fixed to the surrounding tissues; encirclement and invasion of the trachea and oesophagus may cause obstruction; dyspnoea and dysphagia may occur early in this condition and dominate the picture. Even in cases of bilateral involvement there are commonly large areas of normal thyroid tissue. Hence hypothyroidism, whilst it may occur, is not the rule. The titre of anti-thyroid antibodies in the serum is low or absent. Radioactive iodine uptake and plasma levels of protein-bound iodine may be normal. In some of the cases described in the literature there has been an incidental finding of a raised erythrocyte sedimentation rate and a hypochromic anaemia.

The characteristic feature of Riedel's thyroiditis is the fibrotic invasion of the strap muscles of the neck and other neighbouring structures, with no clear line of demarcation between what was originally thyroid tissue and contiguous muscle. Crile (1949) has remarked that the condition is really a diffuse fibrosis of the neck with the thyroid at its centre. Microscopy confirms the presence of a dense fibrosis with small collections of lymphocytes, plasma cells, or mononuclear cells. In the areas involved there is complete destruction of all thyroid tissue with no sign of thyroid lobulation or discernible capsule.

As a great rarity previous cases have been described in which patients with invasive fibrous thyroiditis have had fibrous involvement of other parts of the body. The occurrence of "combined" cases would appear to establish the point of view that the disease process is not uniquely thyroidal. In one case reported by Hache, Woolner, and Bernatz (1962), necropsy revealed a combined inflammatory sclerosing lesion in the thyroid gland and retroperitoneal region. The inflammatory fibrous mass in the retroperitoneal region extended from the duodenum to the promontory of the sacrum, both the aorta and the ureters being within the mass.

The following case illustrates this involvement of tissues additional to the thyroid and is of ophthalmological interest in view of the extensive orbital involvement.

Case Report

An unmarried woman aged 43 years attended the Outpatient's Clinic on September 20, 1960, with a 3 weeks' history of puffiness of the eyelids and epiphora. Apart from lassitude and a feeling of depression, accompanied by anorexia, there were no other symptoms. The patient did not complain of any excessive perspiration and was not particularly irritable.

Examination.—The lower eyelids were markedly, and the upper moderately, swollen and oedematous. There was no lid lag or retraction of the upper lids, nor at this time was there proptosis. Ocular movements were full. Visual acuity was 6/6 in each eye. The anterior and posterior ocular segments were also normal.

At this time a general examination revealed no abnormality apart from an enlargement of the thyroid gland which appeared as a moderately large, smooth, but diffuse discrete goitre. There were no associated palpable lymph glands, and no bruit was heard. The hands and feet were warm

but not hot and they did not show excessive perspiration. The hair of the scalp and eye-brows was normal.

The radioactive iodine uptake was 2 per cent. of the dose. A tentative diagnosis of struma lymphomatosa was made.

Treatment.—1-thyroxine 0.1 mg. daily was started. In spite of this treatment chemosis of both conjunctivae developed together with increased retro-orbital tension and exposure keratitis of both corneae.

On November 4, 1960, a lateral tarsorrhaphy was performed on each eye. Some 2 weeks later, as there was no amelioration in the ocular condition, a course of radiotherapy was commenced. Treatment was given to both orbits by anterior and lateral fields; 220 kV., 2.4 mm. Cu. H.V.L. For the anterior field the eye itself was protected by a 1.5 cm. lead shield and for the lateral eye field the anterior part did not include the lens. The field sizes were lateral 6–4 cm. and anterior 4–4 cm. The dose was 50 r. to each eye twice weekly, the total skin dose being 6,675 r. in 32 days, giving an estimated dose to the apex of the orbit of 900 r.

Progress.—There was an improvement in the ocular condition with some regression of the proptosis, clearing of the chemosis, and healing of the corneal ulcers.

In March, 1961, the right cornea again developed a limbal ulcer at 6 o'clock, which perforated in the course of a few days; this was treated with a bridge conjunctival flap which effected healing.

In May, 1961, there was complete absence of elevatory action of both eyes; other ocular movements were within normal limits. The visual acuity was 6/36 in the right eye and 6/9 in the left; both corneae had scarring involving the lower third but there was no active keratitis. The ocular tension and fundi were normal.

The patient remained symptom-free and did not attend again until April, 1962, when a marginal ulcer developed on the right cornea involving the lower temporal quadrant. At this time it was observed that the lower part of both orbits was filled with a hard mass which had a sharp margin. The following month a similar marginal ulcer in the lower temporal quadrant developed in the left eye. These ulcers became chronic and during the following months, despite treatment with antibiotics, mydriatics, bandaging, etc., spread over the corneae both circumferentially and centrally. The appearance was not unlike that of a Mooren's ulcer, spreading centrally, undermining the surface to produce an overhanging lip, with the peripheral area remaining as a thin ectatic vascularized zone. During this time the orbital mass gradually encompassed both eyes, which now appeared to be encased in a hard fibrotic mass with a sharp edge simulating the orbital margin.

In September the patient fell and hit the side of her head on the edge of a table, rupturing both corneae. The right cornea had an extensive laceration along the length of the thin ectatic temporal area through which vitreous was presenting and was considered to be beyond surgical repair. There was also a small perforation of the left cornea; this healed with conservative treatment, leaving an active surface ulcer.

By January, 1963, the keratitis of the left eye involved the whole temporal half of the cornea with a sharp lip passing centrally down the vertical meridian. As soon as the midline had been reached the keratitis became quiescent, leaving the temporal half of the cornea thin, ectatic, and vascularized, while the nasal half was clear and of normal thickness. Unfortunately no sooner had the keratitis become quiescent than the left eye developed secondary glaucoma, but this was successfully relieved by a drainage operation.

In July, 1963, the patient developed a necrotic ulcer of the skin lateral to the right outer canthus (Figs 1 and 2, overleaf), and was therefore re-admitted to hospital for further investigations.

Biopsy.—Both orbital and thyroid biopsies were obtained. At operation it was found that the enlargement of the thyroid was infiltrating the strap muscles of the neck, with encirclement and invasion of the larynx.

The biopsy report on the thyroid stated: "There were no thyroid vesicles seen in the sections or signs of thyroid tissue but an abundance of hyaline scar tissue with chronic inflammatory foci and adherent striated muscle." (Fig. 3, overleaf). The orbital biopsy showed: "Hyaline scar tissue with broad collagen bundles and chronic inflammatory foci." (Fig. 4, overleaf). The orbital and thyroid biopsies had a remarkably similar histological appearance.



FIG. 1.—Frontal view very shortly after biopsies had been taken, showing necrotic ulcer of skin above right lateral canthus.



FIG. 2.—Lateral view, showing the sharp ridge below the right eye-brow caused by the underlying hard mass.

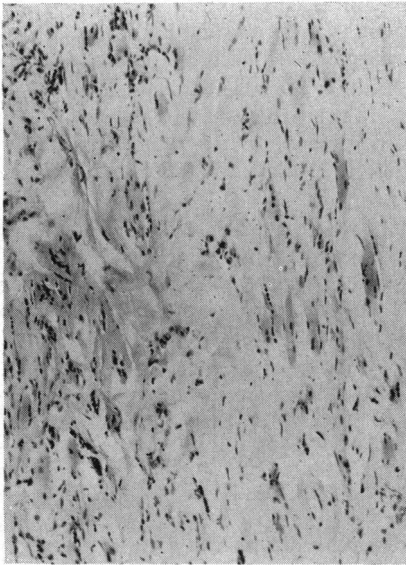


FIG. 3.—Histological section of thyroid, showing total absence of thyroid tissue which has been replaced by fibrous tissue with lymphocytes and plasma cells. $\times 90$.

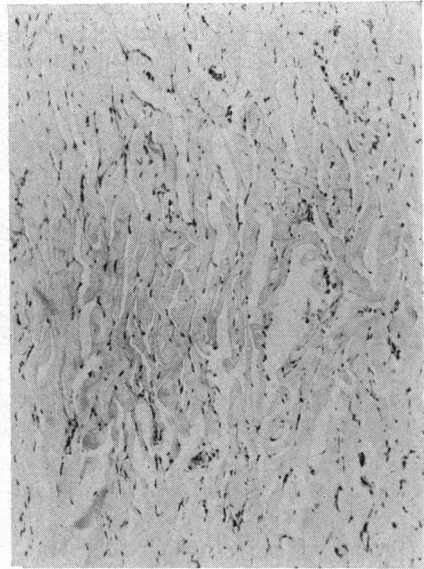


FIG. 4.—Histological section of orbital tissue, showing marked fibrotic reaction with scattered lymphocytes and plasma cells. $\times 90$.

Blood Analysis.—Hb 66 per cent. (9.8 g. 100 ml.); mean corpuscular diameter 7.3 μ ; red blood cells 3,700,000 per c.mm.; colour index 0.90; packed cell volume 35 per cent.; mean corpuscular volume 95 cu. μ ; mean corpuscular haemoglobin concentration 28 g. per cent.

The red blood cells showed marked hypochromic anaemia and slight anisocytosis.

White blood cells 10,500 per c.mm.; differential count neutrophils 58 per cent., lymphocytes 35 per cent., monocytes 5 per cent., eosinophils 2 per cent., basophils 0 per cent.

Erythrocyte sedimentation rate (Westergren) (100) 51 mm. at 1 hr.

The serum contained no antithyroid antibodies.

Diagnosis.—The above findings suggested a diagnosis of Riedel's thyroiditis. The patient has since been on a low dosage of systemic prednisolone, 5 mg. twice daily, and there has been a marked improvement in the red blood cell count and a reduction in the erythrocyte sedimentation rate.

Conclusions and Discussion

When this patient first presented with ocular signs the clinical appearance resembled to some extent that of thyrotropic exophthalmos. The subsequent course, however, differed in that the deeper tissues became increasingly hard, presenting to palpation the sensation of a broad bony orbital rim within the already existent orbit. The possibility of an unusual response of the orbital tissue to irradiation was considered unlikely in view of the relatively small dosage of *x* rays employed. The corneal changes were considered to be due to ischaemic necrosis secondary in the intense fibrosis; a similar change occurred later in the overlying skin of the right upper eyelid at its outer aspect.

It became evident, however, that the thyroid itself was undergoing a similar slow change and a biopsy was undertaken to establish whether the underlying changes were due to fibrosis or neoplasia. The resultant histological picture suggested that this was a case of Riedel's thyroiditis with orbital involvement.

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