Autoimmune Thyroid Diseases in 65 Japanese Women with Turner Syndrome

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Abstract. Turner syndrome (TS) is associated with a number of complications including thyroid disease. In this study, the prevalence of thyroid disease was evaluated in Japanese women with TS. The medical charts of 65 TS women (age 30±9 years old, range: 15-61), treated with estrogen replacement therapy or with antosteoporotic pharmaceuticals at our outpatient clinic, were reviewed. History of thyroid disease, titer of thyroid autoantibodies and thyroid function were recorded. Thyroid autoantibodies were undetectable in 28 of 65 women (43%), and thyroid function was normal in all these women. of the 37 women with thyroid autoantibodies (57%), 3 had Graves’ disease and 20 women were hypothyroidism and diagnosed as Hashimoto’s thyroiditis. The remaining 14 women with euthyroidism were also considered to be so-called probable cases of Hashimoto’s thyroiditis. In 20 women with hypothyroidism, 14 (70%) received replacement therapy with levothyroxine. The replacement with levothyroxine started between age 17 and 60 (median: 31 years old). These data showed that more than half of Japanese women with TS in adulthood had thyroid autoantibodies. In women with TS, monitoring of thyroid hormone is important to detect hypothyroidism earlier and start adequate replacement therapy.

Key words: Hashimoto’s thyroiditis, Turner syndrome

TURNER SYNDROME is a relatively common chromosomal disorder, caused by complete or partial X monosomy in some or all cells. This syndrome is characterized by short stature and estrogen deficiency secondary to ovarian dysgenesis. Affected women are susceptible to a number of medical problems including cardiovascular disease, osteoporosis, and other endocrine disorders [1]. A high prevalence of autoimmune thyroid disease has been also described in Turner syndrome [1], but only a few surveys of Japanese women with Turner syndrome have been reported. Several previous studies showed that hypothyroidism began to appear even before adolescence in Turner syndrome [1]. In the general population, the prevalence of overt and subclinical hypothyroidism increases with age [2]. The incidence of thyroid disease apparently increases with age as well in women with Turner syndrome. In this study, we analyzed the prevalence of thyroid disease associated with Turner syndrome in adulthood in Japan.

Subjects and Methods

The medical charts of 65 women with Turner syndrome (age mean±SD: 30±9 yr. range; 15-61) who were attending our outpatient clinic to receive estrogen replacement therapy or antosteoporotic pharmaceuticals, were retrospectively reviewed. The diagnosis of Turner syndrome was based on clinical findings such as short stature, primary amenorrhea and cubitus valgus etc., and peripheral blood leukocyte karyotype analysis. The karyotype was 45X in 20 women, isochromosome-X (including 46XiX(q), 45X/46XiX(q), 45X/46Xi(X)(q10)) in 20, structural disturbances in 13, mosaic without structural disturbances (including 45X/46XX, 45X/ 47XXX) in 4. In 8 women, the
The findings of Hashimoto’s thyroiditis were revealed in 5 women. In other 9 women including two with normal findings by thyroid ultrasonography, the presence of goiter was not recorded in the medical chart. These women were considered to be so-called probable cases of Hashimoto’s thyroiditis.

Figure 2 shows the numbers of women who had thyroid autoantibodies in each decade. Fifty-two percent of women under 40 years old (29/56) had thyroid autoantibodies. On the other hand, 89% of the women above 40 years old (8/9) had thyroid autoantibodies.

**Discussion**

In this study, more than half of the adults with Turner syndrome had thyroid autoantibodies, and the prevalence of hypothyroidism was 31% (20/65). Seventy percent of the women with hypothyroidism were receiving L-thyroxine. In 1993, Konno et al. investigated the prevalence of thyroid dysfunction and its relation to thyroid autoantibodies in apparently healthy people residing in Japan. They reported that the thyroid autoantibodies were positive in 13.8% of females. There was an age-related increase in the prevalence of positive thyroid autoantibodies and the frequency was about 7% below 30 years old, and 14% above 30 years of age. The overall preva-
Elsheikh et al. reported that autoimmune thyroid disease has been found to be particularly prevalent in women with the isochromosome karyotype compared with other karyotypes [1]. In this study, 9 of 20 women with 45X (45%) and 13 of 20 women with the isochromosome (65%) had thyroid autoantibodies and the prevalence was not statistically significant by chi-square test for independence.

Recently, Hamano et al. assessed the arterial stiffness of patients with hypothyroidism by the brachial-ankle pulse wave velocity and found that an early atherosclerosis process was observed not only in overt but also in subclinical hypothyroidism, and this process could be reversed by an appropriate hormone replacement therapy [10]. Taking these findings into consideration, thyroid function in women with Turner syndrome should be monitored repeatedly to avoid unrecognized hypothyroidism.

In this study, 3 of 65 women had Graves’ disease. Our findings were consistent with the earlier findings that Graves’ disease may occur in women with Turner syndrome, although the incidence was low [5]. In line with earlier studies, we observed that Hashimoto’s thyroiditis was the most frequent type of thyroid dysfunction in women with Turner syndrome.

In this series, all women negative for thyroid autoantibody had normal thyroid function. However, Massa et al. indicated that some women without detectable antithyroid antibody had goiter or biochemical hypothyroidism and that the absence of thyroid autoantibody did not exclude a disturbance of thyroid function [11]. Therefore, it would be important to survey the thyroid function of these women with Turner syndrome regularly even if they were negative for thyroid autoantibody.

In conclusion, more than half of women with Turner syndrome in adulthood had thyroid autoantibodies. Thyroid function in women with Turner syndrome should be monitored repeatedly to avoid unrecognized hypothyroidism and start adequate replacement therapy with levothyroxine promptly.

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References


