Primary Thyroid Lymphoma

WHAT IS THE THYROID GLAND?

The thyroid gland is a butterfly-shaped endocrine gland that is normally located in the lower front of the neck. The thyroid's job is to make thyroid hormone, which are secreted into the blood and then carried to every tissue in the body. Thyroid hormone helps the body use energy, stay warm and keep the brain, heart, muscles, and other organs working normally.

PRIMARY THYROID LYMPHOMA

Lymphoma is a cancer that develops in the lymphatic system (the tissues and organs that produce, store, and carry white blood cells). Lymphoma usually occurs within lymph nodes, but in rare cases, it arises from lymphocytes that are present within the thyroid gland. This is called primary thyroid lymphoma, to distinguish it from lymphomatous involvement of the thyroid gland due to metastasis or direct extension.

WHAT CAUSES PRIMARY THYROID LYMPHOMA?

Primary thyroid lymphoma is more likely to occur in people who have Hashimoto's thyroiditis (see *Hashimoto's thyroiditis brochure*), which is an autoimmune condition where the thyroid gland is infiltrated by lymphocytes. However, it is important to understand that, while Hashimoto's thyroiditis is a common condition, thyroid lymphoma is rare, representing less than 5% of thyroid malignancies and less than2% of all lymphomas occurring outside of the lymph nodes. No other risk factors have been identified that cause thyroid lymphoma. Unlike other forms of thyroid cancer, exposure to radiation has not been clearly associated with thyroid lymphoma.

Thyroid lymphoma affects women more than men, and most commonly occurs between the ages of 65 to 75 years.

WHAT ARE THE SYMPTOMS OF PRIMARY Thyroid lymphoma?

In thyroid lymphoma, the thyroid gland becomes enlarged and firm, which often causes difficulty swallowing, shortness of breath, and/or a hoarse voice. Patients often notice a rapidly enlarging mass in their neck, which may be tender. If the enlarged gland presses on the veins through which blood drains from the head, this can cause puffiness or swelling of the face. A minority of patients may also experience more generalized symptoms associated with lymphoma, such as fever, night sweats, and unintentional weight loss.

Thyroid lymphoma can also cause hypothyroidism (see *Hypothyroidism brochure*), or decreased production of thyroid hormone, due to infiltration of the normal thyroid gland. The symptoms of hypothyroidism include feeling slow or tired, feeling cold, dry skin, and constipation.

HOW IS PRIMARY THYROID LYMPHOMA DIAGNOSED?

There are no blood tests to evaluate for thyroid lymphoma, although a blood test for Thyroid Stimulating Hormone (TSH) can be done to screen for hypothyroidism. An ultrasound of the neck is usually performed, which shows an enlarged thyroid gland and sometimes enlarged lymph nodes surrounding the gland. The diagnosis is often suggested by a *fine* needle aspiration (FNA) biopsy of the gland and/or lymph nodes. However, this test is often inconclusive for lymphoma and additional tissue may be needed to establish the diagnosis. This is usually obtained by using a larger needle (core needle biopsy) or surgically removing a small piece of tissue from the thyroid or an adjacent lymph node. Special immunostains (a general term in biochemistry that applies to any use of an antibody-based method to detect a specific protein in a sample) or other specialized studies of the cells are often needed to establish the diagnosis. Following diagnosis, additional studies will be obtained in order to determine if the lymphoma has spread beyond the thyroid and more accurately stage the cancer.

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HOW IS PRIMARY THYROID LYMPHOMA TREATED?

Unlike other forms of thyroid cancer, thyroid lymphoma is not treated by surgical removal of the thyroid gland. Instead, it is treated with chemotherapy, radiation treatment, or a combination of the two. Treatment is usually directed by a medical oncologist (a cancer specialist) or hematologist.

Patients who receive radiation for thyroid lymphoma are at high risk for developing hypothyroidism and need to have their thyroid function blood tests measured regularly.

WHAT IS THE PROGNOSIS FOR PRIMARY THYROID LYMPHOMA?

As with most cancers, the prognosis depends on the subtype of lymphoma and how far it has spread at the time of diagnosis. The prognosis for thyroid lymphoma is not as favorable as it is for the much more common primary thyroid cancers (such as papillary and follicular). However, many patients with localized tumors can be cured, and some studies suggest a combination of both radiation and chemotherapy results in better outcomes.

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FURTHER INFORMATION

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