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THYROID CANCER

Medullary Thyroid Carcinoma: prevalence and the case for screening

BACKGROUND

Medullary cancer is a rare form of thyroid cancer that accounts for <10% of all thyroid cancers. Many of these cancers are associated with a genetic mutation and run in families, especially those that occur in younger patients. The prognosis of medullary thyroid carcinoma depends greatly on the completeness of the first surgical treatment. Calcitonin is a hormone that is secreted by medullary cancer cells and serves as a cancer marker. The utility of routine calcitonin level screening for patients with thyroid nodules, has been the subject of debate for years. Some studies support the notion that screening may detect medullary thyroid cancer at an earlier stage and therefore may improve overall survival. However, as with other thyroid cancers, the clinical consequence of small medullary thyroid cancers (<1 cm, micromedullary cancer) that may be identified by screening is unclear. Further, a pre-cancerous condition known as C-cell hyperplasia also secretes calcitonin. In the first study, the authors look at autopsy studies to try to identify how frequently micromedullary cancer is found. In the second study, the authors attempted to link preoperative calcitonin levels with the findings at surgery.

Article 1:
THE FULL ARTICLE TITLE:

Valle LA, Kloos RT. The prevalence of occult medullary thyroid carcinoma at autopsy. J Clin Endocrinol Metab. October 13, 2010. doi:10.1210/jc.2010-0959.

SUMMARY OF THE STUDY

The authors screened the autopsy reports on a total of 7900 subjects for the finding of micromedullary thyroid carcinoma. These subjects were without known thyroid disease at time of death. Based on this study, micromedullary thyroid carcinoma was found in 0.14% to 0.42 % of subjects at the time of autopsy. In comparison, the more common micropapillary cancer was found in 7.6% to 8.4% of subjects at autopsy.

Article 2:
THE FULL ARTICLE TITLE:

Chambon G et al. The use of preoperative routine measurement of basal serum thyrocalcitonin in candidates for thyroidectomy due to nodular thyroid disorders: results from 2733 consecutive patients. J Clin Endocrinol Metab. September 29, 2010. doi:10.1210/jc.2010-0162.

SUMMARY OF THE STUDY

A total of 2733 French patients with thyroid nodules who were scheduled for thyroid surgery had calcitonin levels measured before surgery. Only 43 patients had elevated calcitonin levels and all had a total thyroidectomy. A total of 7 patients had elevated calcitonin levels had a pre-operative diagnosis of medullary thyroid cancer by biopsy of a known thyroid nodule. A total of 5 patients had micromedullary thyroid cancer and 31 had C-cell hyperplasia. Patients with micromedullary thyroid cancer had no evidence of the cancer prior to surgery other than elevated calcitonin levels. One of these patients was found to have spread of the cancer to the lymph nodes at the time of surgery. Of the 2690 patients whose calcitonin level was not elevated before surgery, 2 had micromedullary thyroid cancer.

WHAT ARE THE IMPLICATIONS OF THESE STUDIES?

Medullary thyroid cancer is often diagnosed early in life, with most genetically-linked cases diagnosed in early adulthood and sporadic cases in fourth decade of life. It is clear that calcitonin screening should be performed in patients with a family history of medullary cancer. Further, surgery is indicated in patients with elevated calcitonin levels even if these turn out to be micromedullary cancers or C-cell hyperplasia. It is unclear what to do with micromedullary cancers that do not have a genetic mutation. The first study indicates that micromedullary cancer is found in <1% of individuals

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without known thyroid disease. Screening all patients with thyroid nodules would identify mostly C-cell hyperplasia. While C-cell hyperplasia is a precancerous condition, little is known about how frequently C-cell hyperplasia progresses to medullary thyroid cancer. Thus, calcitonin screening before surgery subjected 31 patients who had C-cell hyperplasia to surgery that may well be unnecessary. However, 5 patients were identified with micromedullary cancer and 20% of these patients had spread of the cancer to the lymph nodes, indicating that even micromedullary cancer can be aggressive. Thus, more studies are needed to define the behavior of C-cell hyperplasia and micromedullary cancer before recommending general calcitonin screening.

— Mona Sabra, MD

ATA THYROID BROCHURE LINKS

Thyroid cancer: http://thyroid.org/patients/patient_brochures/cancer_of_thyroid.html

ABBREVIATIONS & DEFINITIONS

Medullary thyroid cancer — a relatively rare type of thyroid cancer that also may be inherited through a genetic mutation. Medullary cancer arises from the C-cells in the thyroid.

Micro-medullary thyroid cancer — a medullary thyroid cancer <1 cm in size.

Thyroidectomy — surgery to remove the entire thyroid gland. When the entire thyroid is removed it is termed a total thyroidectomy. When less is removed, such as in removal of a lobe, it is termed a partial thyroidectomy.

C-cell hyperplasia — An abnormal growth of parafollicular (C-cells) cells that usually occurs before the development of familial forms of medullary thyroid cancer and is considered a pre-cancerous condition

Calcitonin — a hormone that is secreted by cells in the thyroid (C-cells) that has a minor effect on blood calcium levels. Calcitonin levels are increased in patients with medullary thyroid cancer.