What Is Medullary Thyroid Cancer (MTC)?

Medullary thyroid cancer, or MTC, is a rare form of cancer of the thyroid gland. This gland is part of the endocrine system as shown below. When there is no family history of the disease it is called sporadic MTC. 1 in 4 or 25% of cases of MTC occur as part of a rare disorder which can be passed down in families called multiple endocrine neoplasia Type 2 (MEN2). All apparently sporadic MTC patients must be offered DNA screening (a gene test) in case theirs is the first case of a gene disorder in the family. See our "What Is Multiple Endocrine Neoplasia Type 2A?" and "What Is Multiple Endocrine Neoplasia Type 2B" factsheets.

The thyroid gland is found at the front of the neck. This gland makes 3 hormones; thyroxine and tri-iodothyronine (essential for maintaining the body's metabolism and mental and physical development), and calcitonin.

MTC starts in the C-cells of the thyroid which make the hormone calcitonin. An increase in the number of C-cells (C-cell hyperplasia) occurs before they become cancerous. MTC can spread to nearby lymph nodes. Even so, there may be no physical symptoms of this. As MTC grows, calcitonin levels increase.

If the thyroid and nearby lymph nodes are removed by surgery while the cancer is still contained within the thyroid (total thyroidectomy and central lymph node dissection), a patient is usually cured. If calcitonin levels are still raised after surgery, this shows that the cancer has spread (metastatic) or has not been completely removed. In this case further surgery and other therapies may be used to control it. As yet there is no complete cure for MTC that has spread; however, it is often slow-growing and can exist without symptoms for many years. Symptoms that develop can often be controlled by the use of radiotherapy, new drug therapies and very rarely chemotherapy (see How is MTC treated when it has spread?).

How is MTC diagnosed?

In most cases, patients may notice a lump in the neck which was not there before, or this may be noticed by a partner or colleague. Sometimes MTC may cause diarrhea, although it is not immediately clear that this may be related to a problem in the neck. In some families there may be a history of MTC which warrants gene testing of other family members. A diagnosis of medullary thyroid cancer may be confirmed by a fine needle biopsy of the neck lump, and ultimately by surgery (see Testing for MTC). A blood test to measure calcitonin is sometimes used to make the diagnosis of MTC, but levels may be raised for a variety of reasons other than MTC.

Children and MTC

It is rare for children to develop MTC. However, those that do should be seen by a genetics clinic to be tested for genetic diseases that can be passed down in families, as they are likely to have this condition and will need testing for other potential health problems. For more information on related genetic diseases, see our "What Is Multiple Endocrine Neoplasia Type 2A?" and "What Is Multiple Endocrine Neoplasia Type 2B" factsheets

Testing for MTC

You may have the following tests to confirm a diagnosis of MTC:

Baseline calcitonin blood test
Calcitonin levels are usually raised when MTC is present (note that once drawn, the blood must be taken immediately and on ice to a chilled centrifuge in the lab).

Ultrasound scan and fine needle aspiration (FNA)
A painless scan of the neck provides pictures of the thyroid and any lumps or cysts. A tissue biopsy (sample) is obtained from the insertion of a very fine needle into the thyroid lump. The sample is looked at under a microscope and is a very reliable way of diagnosing MTC.

Treating MTC

MTC is different from other types of thyroid cancer. It is best treated in a hospital that is a center of expertise for MTC, and by an experienced endocrine or head-and-neck surgeon who regularly operates on such patients.

Once a diagnosis of MTC has been made, an ultrasound scan of the neck should be done to help map the extent of the disease (staging ultrasound). If there is no suspicion of enlarged lymph nodes, removal of the thyroid together with central lymph nodes is done (thyroidectomy with central node dissection). If enlarged nodes are detected, removal of other neck lymph nodes (lateral neck) should be undertaken at the same time as the thyroidectomy. If MTC is diagnosed, special blood and urine tests must be done (even if there is no family history of a genetic condition) to rule out a growth of the adrenal gland which could cause problems during or after surgery.

Surgery

Total thyroidectomy + central node dissection: A cut is made at the base of the front of the neck from which the thyroid and nearby lymph nodes can be removed. A larger cut is needed if the cervical neck lymph nodes need to be removed as well. Eating and drinking is possible almost straight away after waking up from the operation.

Hospital stay
You will probably stay in the hospital for around 2-3 days in total.

Risks
Thyroid surgery is generally safe but there are some possible risks that you need to be aware of:

- Injury to the nerves that control the vocal cords which may affect the voice (1-2%).
- Unavoidable removal of or injury to the parathyroid glands may occur which might result in a temporary drop in calcium levels in the blood (hypocalcemia). Sometimes this may be permanent. Symptoms of low blood calcium include tingling or ‘pins and needles’ in the lips, fingers and toes, and eventually cramping. All these symptoms can be corrected with medication.

What medication will I need to take?

Levothyroxine

Levothyroxine (thyroxine) must be taken life-long after a total thyroidectomy. Tablets are taken once a day and doses are based on the body weight of the individual (typically between 100-150mcg for adults, lower for children). You will need regular blood tests to make sure that you are on the right dose. A dose that is too big may cause symptoms such as rapid heartbeat, sweating, anxiety, shaking and weight loss. A dose that is too small may cause symptoms such as tiredness, a slow heartbeat, sensitivity to cold, and weight gain. Although the above symptoms may suggest a need to change the dose, the same symptoms can occur in other conditions. Only a blood test measuring the thyroid stimulating hormone (TSH) level can help doctors be sure when a change of dose is needed. Once your ideal dose is found, as judged by blood tests, repeat tests only need to be done once a year.

Calcium replacement medication (required if parathyroid glands are injured or unavoidably removed along with the thyroid)

Vitamin D

Vitamin D tablets or capsules aid absorption of calcium from the diet. Usually taken once a day, this may be the only life-long medication required after parathyroid injury or removal.

Calcium carbonate

This is a chalk-like tablet that should be chewed or sucked. This is often used as a temporary calcium "top-up" after surgery but is not necessarily required life-long. Too large a dose or an indication that this supplement is no longer needed may become apparent if the patient begins to suffer from headaches, nausea and vomiting.

Magnesium supplement

This may be in the form of an injection or tablet (e.g., magnesium glycerol-phosphate) but is rarely needed long-term.

How is MTC treated when it has spread?

Patients with MTC may have high levels of calcitonin in the blood even after surgery. However, although this shows that there are MTC cells left in the body, patients with calcitonin levels that are higher than normal, staying the same over a period of time, or slowly increasing, often do not need further tests or treatment. This is because scans are not always able to find a site of disease outside of the neck unless calcitonin levels are very high: calcitonin alone is not proof of a growing tumor. Even so, in some patients, the search for metastatic disease may involve various scans (including radioactive isotope scans), followed by treatment with more surgery or radiotherapy needed.

C-cells also produce a substance called carcinoembryonic antigen (CEA) which is measured alongside calcitonin to help gather more information about disease that has spread outside of the neck.

MIBG / octreotide therapy

Where surgery is no longer an option due to the extent or site of the disease, some expert medical centers may use radioactive treatment (octreotide or MIBG). These have very few side effects and

can help to reduce or control the spread of MTC. However, they are only used if tests show that they will be taken up by the tumor. The agent is given through a vein by slow injection. The patient remains radioactive for a few days and therefore must be nursed alone in a special room. The treatment may need to be repeated several times at 3 or 6-month intervals. Possible side effects of MIBG / octreotide therapy include feeling sick, and sometimes being sick too.

Until a complete cure is found, much of the current focus of treatment for extensive metastatic MTC is on the relief of the symptoms it causes:

Diarrhea: a change to the patient's diet may be required, together with anti-diarrheal medicine which can help to control it. In some cases, diarrhea can also be relieved by treatment with drugs called octreotide or lanreotide, although this is not the case for everyone. Some believe that in such cases it may also help slow down the growth of the tumor.

Flushing: medicines used to control stomach ulcers called H2 blockers (cimetidine or ranitidine) may sometimes be used to help ease flushing.

Painful bone metastases: external radiation therapy can sometimes provide rapid relief from pain when MTC has spread to the bone. In all cases, pain medications may be prescribed.

Other therapies for MTC that has spread

A class of drugs called tyrosine kinase inhibitors (TKIs) and multi-kinase inhibitors (MKIs) are becoming available for use in metastatic MTC where other treatments do not work. These drugs are not cures but may in some people help slow or stop the spread of MTC as well as relieve symptoms. You should discuss this option, the possible serious side effects and other issues about these therapies with your specialist. TKIs and MKIs are not always suitable for everyone.

Resources

Association for Multiple Endocrine Neoplasia Disorders

www.amend.org.uk

For the full list of INCA members: https://incalliance.org/members/