

Marginal Zone Lymphoma

Your doctor has just diagnosed that you have a "marginal zone lymphoma".

This is a non-Hodgkin's type of lymphoma, an illness linked to malignant over-production of cells in the immunosystem. These cells are mainly found in the lymphatic ganglions, the lymph, the spleen and in the bone marrow.

The lymphatic ganglions are where pathogens are neutralised by the immune cells.

The occurrence of a follicular lymphoma is linked to uncontrolled reproduction of abnormal B lymphocytes. Lymphocytes are white cells whose special purpose is to produce antibodies. They circulate in the blood vessels and lymphatic vessels. When one of them becomes abnormal, it tends to proliferate as it reproduces more quickly and/or lives longer than normal cells. An accumulation of these malignant cells ends up forming one or several tumours.

The lymphocyte at the origin of these marginal zone lymphomas is a somewhat unusual cell, which is called a "memory cell". It is a lymphocyte which has kept in its memory a previous interaction with a foreign agent (antigen) and which, because of this, is able to produce a more rapid and specific immuno-response. These memory B Lymphocytes are stored in a ganglion zone that is called "the marginal zone". It is from this zone that the abnormal lymphocytes spread. Hence the name of the disease.

Marginal zone lymphomas are non-Hodgkin's Lymphomas (NHL) of which 30 or so types have been identified up to now. Amongst the marginal zone lymphomas, three quite distinct diseases are distinguished:

- ♦ **MALT Lymphomas.** Unlike most NHLs this type of lymphoma rarely affects the lymphatic ganglions. Abnormal cells infiltrate one or more organs. Most often it will be the stomach (gastric MALT lymphoma). But the disease can also affect the lungs, the skin or the eye etc. This type of Lymphoma may be caused by chronic infection by a bacteria. For instance, the *Helicobacter Pylori* bacteria may be at the root of gastric MALT Lymphomas.
- ♦ **Splenic lymphomas.** In these cases, the cells accumulate mainly in the spleen as well as the blood and bone marrow.
- ♦ **Lymphomas of the ganglions.** This kind of Lymphoma results in an attack on lymphatic ganglions by abnormal lymphocytes.

Marginal zone lymphomas are relatively widely-occurring NHLs. There are around 2800 new cases in France each year. The occurrence of this disease increases with age, the average age currently being 60. This disease is neither hereditary nor contagious.

The first symptoms

Marginal zone lymphomas are among the NHLs called "indolent", meaning that they develop slowly. Several years can pass between the onset of the disease and the appearance of the first symptoms. These are different according to the type of lymphoma.

Thus MALT lymphomas may result in burning sensations or heaviness around the stomach. In the case of splenic lymphoma, an increase in the volume of the spleen may produce a sensation of heaviness on the left flank. Ganglionic lymphomas, for their part, cause an increase in the size of one or more ganglions. Rarely painful, even when they become apparent, these are to be found in the neck or armpits, but they may be present in other parts of the body, particularly the chest and abdomen.

Other more general non-specific symptoms may also show, but they tend to be rare. There might be a temperature, an unexplained loss of weight, abundant night-time sweating or fatigue.

Doctor's notes :

Certain treatments may lead to sterility, especially where men are concerned. So sperm conservation is offered after the diagnosis.

Use antibiotics against a lymphoma

It is not often that a cancer can be treated using antibiotics or antiviral. Marginal zone lymphomas are one of the rare lymphomas that can be treated in such a way. When they are associated with a chronic bacterial infection treatment with antibiotics may in fact be enough to treat the lymphoma. The disappearance of the bacteria is accompanied by remission from the disease.

Any treatment is likely to produce unwanted side-effects and may present risks. Your doctor will inform you and will tell you which signs to look out for before you start the treatment suggested

Participating in a clinical trial

The best way to contribute to the improvement of disease management is to treat patients in the context of clinical trial. If your doctor suggests this could apply to you, he will explain its purpose, protocol, expected benefits, potential risks and will give you an information leaflet.

Participating in a trial of course means you will first have to give your written informed consent.

Diagnosis

These different symptoms are suggestive of a marginal zone lymphoma. The actual diagnosis of this is formally arrived at following a biopsy which involves removing a sample of tissue from one of the ganglions or organs presenting abnormalities. The cells contained in the samples are then examined under a microscope by an anatomo-pathologist, i.e. a doctor who specialises in tissue study. Other tests are generally carried out to establish the spread of the disease and its state of development. Several scans or x-rays are taken especially in search of "deeper" effects, i.e. those than cannot be detected in the course of a clinical examination. As a general rule, the doctor will prescribe a chest X-ray, and an ultrasound scan of the neck, chest, abdomen and pelvis. Blood tests are also carried out, especially to measure the elements that mark the progress of the disease. Depending on the state of health of the individual patient, other tests may be requested to complete the assessment.

Treatment

Treatment of marginal zone lymphomas is adapted according to the type of lymphoma, its stage of development, the organs affected as well as the general state of the patient's health.

Where the disease is linked to a bacteria or a virus, a treatment using antibiotics or antiviral will be adopted. Eradicating the bacteria will bring about a complete remission from the lymphoma in a large number of cases.

For the other types of marginal zone lymphoma, treatment is based either on a surgical intervention (aimed at removing, when this is possible) the zone or the organ affected, or by radiotherapy or by chemotherapy. These different therapeutic approaches may be offered individually or successively.

Radiotherapy consists of subjecting the zone or zones where the abnormal lymphocytes are to be found to X-rays in order to destroy them.

Chemotherapy is based on the administration of one or two medicines which will attack the abnormal lymphocytes. Chemotherapies used against marginal zone lymphomas are mild and only expose the patient to limited side effects.

In recent years, a new kind of treatment has been perfected: monoclonal antibodies. These are medicines that are specifically targeted on the diseased cells. They imitate natural antibodies and bring about the death of the cells they are aimed at. This treatment may be administered on its own or along with chemotherapy. In the great majority of cases these current treatments bring about a prolonged remission from the disease, i.e. the disappearance of all the clinical symptoms and biological signs of the lymphoma.

In case of a relapse of the disease, it is quite possible to undertake a new treatment and gain a new remission.

Monitoring

Monitoring of the disease is adapted according to the general state of health of the patient and the response to the treatment. After a remission, it is generally suggested that the patient visit a haematologist once or twice a year for a clinical assessment and complete blood tests.

Useful contacts:

- Secretarial / appointment:
- Nursing consultation:
- Consulting psychologist:
- Social worker:
- In an emergency: