Splenic marginal zone lymphoma is a type of marginal zone lymphoma.

Marginal zone lymphoma is a rare type of non-Hodgkin lymphoma (NHL). The 3 main types of marginal zone lymphoma are: extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT); nodal; and splenic*. Splenic marginal zone lymphoma (SMZL) is a rare type of slow-growing lymphoma accounting for less than 2% of all NHL cases. It is sometimes referred to as ‘splenic lymphoma with villous lymphocytes’, which refers to the type of cells found in the blood stream of people with SMZL. Whilst it is unlikely to occur, SMZL does have the potential to transform into an aggressive lymphoma.

How does Splenic marginal zone lymphoma affect the body?
SMZL usually begins in the spleen, with symptoms often taking years to develop. The spleen becomes enlarged (splenomegaly), which puts pressure on surrounding organs and tissue, and can result in abdominal discomfort and swelling (generally in the upper left side of the abdomen). Splenomegaly can affect the usual function of the spleen and is associated with low numbers of blood cells. A diminished number of red blood cells (anaemia) leads to shortness of breath, lethargy and fatigue. Reduced numbers of platelets (thrombocytopenia) will result in an increased risk of bleeding and bruising. Unlike many other types of NHL, splenic marginal zone lymphoma rarely causes swollen lymph nodes, however it does often involve the bone marrow and blood. In SMZL there may be enlargement of the liver, known as hepatomegaly.

Who does Splenic marginal zone lymphoma commonly affect?
It typically affects people aged over 50 years and is slightly more common in women.

Do we know what causes Splenic marginal zone lymphoma?
Whilst the exact causes are not known, it has been associated with hepatitis C infection. SMZL is not infectious and cannot be passed from one person to another.

How is Splenic marginal zone lymphoma treated?
There are several treatment options for SMZL. If no symptoms exist, people are usually managed using the ‘watch and wait’ approach*, which involves regular monitoring of the condition by their doctor or specialist. Symptoms generally develop as a result of an enlarged spleen which are usually resolved once the spleen is surgically removed (splenectomy). A splenectomy is not always a viable option, in which case other therapies can be considered. Radiotherapy can be given to the spleen in an attempt to shrink it to a more regular size and function. Chemotherapy drugs may be used to treat SMZL either as single agents or in combination.

Commonly used regimens for SMZL include: CVP (cyclophosphamide, vincristine and prednisolone) or CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone). Fludarabine is another chemotherapy drug often used alone or in combination with cyclophosphamide. The monoclonal antibody rituximab is commonly given in combination with chemotherapy to remove the lymphoma cells.

*Please refer to factsheets: ‘MALT Lymphoma’, ‘Nodal Marginal Zone Lymphoma’ and ‘Understanding Watch & Wait’.


For more information, freecall 1800 620 420
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