Schnitzler’s Syndrome

What is it?
Schnitzler’s syndrome is an extremely rare disease which is characterised by a combination of chronic urticarial skin rashes and haematological abnormalities (monoclonal gammopathy, paraproteinaemia). Monoclonal gammopathy develops from the uncontrolled growth of a single clone of plasma cell precursors. The antibodies (immunoglobulins) produced are belonging to an immunoglobulin class, usually presenting as a monoclonal gammopathy of the IgM sub-type.

Inflammatory processes in various organ systems occur which present with joint pain, bone and muscle pain, fever bouts and fatigue.

How common is it?
Only some 150 cases of this non-hereditary disease have been reported worldwide so far; it usually starts in the middle-aged (age at onset of the disease approx. 50) and takes a chronic course. Here again the number of undetected cases is presumably clearly higher. The syndrome affects male and female patients equally and does not seem to depend on either ethnicity or climate.

What are the causes of the disease?
The causes underlying Schnitzler’s Syndrome are unknown. An inflammatory substance which plays a vital part, is “Interleukin-1”. This substance is, among others, responsible for the interaction of various inflammatory cells and is chronically elevated in patients with Schnitzler’s Syndrome.

Is it inherited?
Schnitzler’s Syndrome is not inherited.

Why do I get the disease? What can be done against it?
As far as we know today, anyone could get the disease. The disease is incurable. There is currently no approved therapy for Schnitzler’s Syndrome. Off-label treatment with selective Interleukin 1 inhibitors have shown a favourable effect on the clinical symptoms of Schnitzler’s Syndrome.

Is it contagious?
Schnitzler’s Syndrome is not contagious.

What are the characteristic symptoms?
The typical symptoms of Schnitzler’s Syndrome are: Chronic urticarial rash which does not respond to antihistamines, recurrent bouts of fever, bone pain, joint and muscle pain as well as fatigue. Some patients also suffer from inflammation of the eye and increased sensitivity to exposure to cold temperatures.

Are the symptoms of the disease always the same?
Schnitzler’s Syndrome may differ from patient to patient with respect to beginning, duration and severity of the symptoms.

What complications may be experienced?
after years or even decades haematologic diseases, (e.g. lymphomas) may develop In individual cases. Furthermore, the chronic inflammation harbours the risk of an amyloidosis.

How is it diagnosed?
Diagnostic criteria of Schnitzler’s Syndrome are the urticarial rash, a monoclonal IgM or IgG in the
immunofixation of the serum and at least 2 of the following symptoms: fever, joint pain, palpable lymph nodes, enlargement of the liver or spleen, increased sedimentation and bone morphology changes.

What are the diagnostic criteria?
If Schnitzler’s Syndrome is suspected immunofixation of the serum is essential. If there is evidence of monoclonal gammopathy diseases of the hematopoietic system, infectious and autoimmune disorders have to be excluded before the diagnosis Schnitzler’s Syndrome can be made. Consultation of a haematologist is highly recommended. A bone marrow biopsy is often necessary.

How is the disease treated?
There is no therapy yet to prevent an outbreak of the disorder or to cure it. Anti-inflammatory drugs such as Ibuprofen, Diclofenac etc. only have a weak effect in case of fever, joint and muscle complaints. High doses of cortisone are often efficient, whereby a continuous administration leads to severe side-effects. Recently, good responses to the treatment with Interleukin-1 blockers (e.g. Anakinra) were shown.

How long should treatment last?
According to current knowledge treatment should be maintained lifelong.

What regular control check-ups are necessary?
As a rule approx. quarterly checks of the clinical findings and of blood analyses are necessary.

How long will the disease last?
It is an incurable disease with a chronic course.

Can the disease be completely cured?
No, it is a lifelong incurable disorder.

What does the disease mean in matters of nutrition/diet?
Single patients report a deterioration of their symptoms after certain foods. Generally, however, foods do not seem to influence the course of the disease.

Can climate influence the disease?
In some patients the symptoms are more pronounced in winter and in cold climate zones, as compared to summer-times and warm climate zones.