CCL2 in Schnitzler syndrome
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Summary

Schnitzler syndrome is a very rare chronic disease, which usually develops in the second half of life. It appears as skin rashes, muscle/skeletal pain and fever episodes. The trigger and molecular processes in the disease are not understood. However, drugs that inhibit the inflammatory molecule called interleukin-1β were found to relieve the disease. The authors of this German study wondered which further molecules are involved in Schnitzler syndrome. For this purpose, a range of molecules with inflammatory properties was quantified in the blood of the patients and, as a comparison, in healthy people and patients with other inflammatory diseases. Patients with Schnitzler syndrome were found to have strongly elevated levels of CCL2, a molecule known to attract inflammatory cells to affected tissues and to have a special role in bone alterations. CCL2 levels were particularly high in severely affected patients, especially those with intense bone pain. In the bone, CCL2 is known to be produced by certain bone-modifying cells. Laboratory experiments revealed that CCL2 was also highly produced by immune cells and connective tissue cells (fibroblasts). Interleukin-1β as well as the inflammatory molecule TNF-α triggered the production of CCL2 by these cells. When patients were treated with an interleukin-1β-blocking drug, health improvement was paralleled by a drop of CCL2 levels. The authors concluded that in Schnitzler syndrome CCL2 is an important player in the inflammation process in bones and other body sites and may be used in the clinic as an indicator of disease severity.