Case history
A 62 year old female was referred because of persistent elevated inflammatory parameters. She had been treated for chronic lumbar pain with a morphine pump, leading to recurrent infections and finally removal of the pump. Furthermore her clinical history reveals peripheral vascular insufficiency treated with endovascular surgery and stent placement but complicated by a chronic wound on the right foot and recurrent Staphylococci infections (MSSA and MRSA), requiring long courses of antibiotics. She is also known with several allergies towards disinfectants and atopic dermatitis.

Pathogenesis
Antigen mediated
germinal center B2-cell expansion and isotype switching (IgA, IgE, IgG)
Polyreactive, non germinal center B1-cell secretion of “natural” IgM antibodies

Follow-up and management
➔ Screening for auto-immune disease, lymphoproliferative disease and allergic diathesis 1x/y
➔ DD between primary and secondary IgM deficiencies
➔ Treatment as suggested after splenectomy (because of the lack of direct opsonisation).
  • Low threshold to start antibiotics but no prophylaxis: on demand treatment is recommended in Belgium
  • No indication for IVIG because of low concentration of IgM and their short half life.

Diagnosis
A broad laboratory screening revealed increased inflammation parameters (sedimentation 44 mm/u, CRP 24.8mg/l), normal leucocytes count and formula, normal liver and kidney function but a remarkable dysglobulinemia characterized by an IgM of 0.179g/l (normal range 0.46-3), confirmed after repeating the test. IgG and IgA showed normal values and as expected the IgE values were also elevated (119kU/l) confirming the associated allergic diathesis. Further screening for associated auto-immune disease, acquired immunodeficiency syndrome (HIV), hematological disorders and lymphocyte analysis revealed no associated abnormalities.

Conclusion
Deficiency in serum IgM may explain a paradox of diminished responsiveness to foreign antigens (therefore susceptibility to infections) and increased inflammation (because of lack of homeostasis) resulting in auto-immunity (and possibly malignancy) in patients with Selective IgM deficiency.

References
- Louis AG et al. Primary selective IgM deficiency: An ignored immunodeficiency. Clinical reviews in Allergy and Immunology. 2014 Apr;46(2):104-11